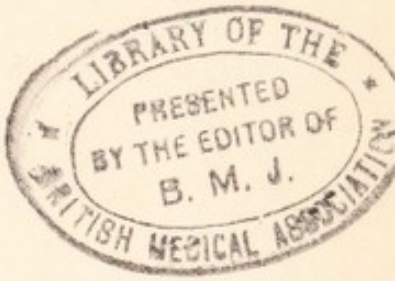
Orbital tumors : results following the transcranial operative attack / by Walter E. Dandy.

ORBITAL TUMORS

W. E. DANDY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ORBITAL TUMORS

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RESULTS FOLLOWING THE TRANSCRANIAL OPERATIVE ATTACK

By

WALTER E. DANDY

With One Hundred Illustrations

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PREFACE

The surgery of orbital tumors has for many years presented a technical problem to the ophthalmic surgeon, the reason being the frequent difficulty of removing such tumors without injury to the eye or the optic nerve. To meet this problem, various operative approaches to different parts of the orbit have been devised. The conventional methods of entering the orbit are either through the conjunctiva, through the upper lid, or by resection of the lateral wall of the orbit. Both the conjunctival and the upper orbital approach have a well-recognized place in orbital surgery, and by these methods many tumors lying anteriorly in the orbit can be satisfactorily and completely removed with a minimum of operative shock or damage to the orbital contents. Many tumors, however, lie deeper in the orbit and are inaccessible with either of these surgical procedures. For removal of such deep tumors the Krönlein approach, or resection of the lateral wall of the orbit, was devised. This operation, however, is open to these criticisms: first, it is applicable only to tumors lying in the temporal side of the orbit; second, it must of necessity leave a deforming scar. In the event that the orbital tumor has an intracranial extension, all of these methods are utterly valueless, since they permit the removal of only a portion of the tumor. Ophthalmologists have therefore long realized that the conventional methods of approach to the orbit are often totally inadequate, and so have turned to the neurosurgeon for assistance.

To meet this well-recognized need, Dr. Dandy has devised his transfrontal technique, which permits removal not only of the orbital tumor but also of any intracranial extension.

Since 1934, in the Johns Hopkins Hospital, it has been the custom for patients with orbital tumors to be operated only after consultation between the ophthalmological and the neurosurgical services. Tumors lying in the anterior portion of the orbit, when there is no reason to suspect an intracranial extension, are operated either by a conjunctival or upper orbital route. All tumors lying deep in the orbit, and invariably any tumor in which there is any reason to suspect an intracranial extension, are routinely referred from the ophthalmological to the neurosurgical service for operation. That this method of arbitrarily separating orbital tumors into two groups, one in the domain of the ophthalmic surgeon, and one in the domain of the neurosurgeon, is thoroughly satisfactory and entirely justified, is fully substantiated by this monograph of Dr. Dandy.

Dr. Dandy has brought to the study of orbital tumors his wide experience in surgical technique and his extensive knowledge of the pathology of intracranial tumors. This monograph, therefore, not only exemplifies what may be accomplished by the co-operation of the ophthalmologist and the neurosurgeon in the solution of a difficult problem, but also sets a definite milestone in the study of the pathology and surgical treatment of orbital tumors.

ALAN C. WOODS

Wilmer Ophthalmological Institute
Johns Hopkins Hospital
May 7, 1941

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ORBITAL TUMORS

INTRODUCTION

Two excellent papers on the pathological aspects of intraorbital tumors by Byers (1901) and Hudson (1912) emphasized the high percentage of these growths that passed into the cranial chamber. Removal through the usual orbital operations, therefore, meant that the patient usually succumbed later to the intracranial growth that remained. In addition, there was local recurrence in the orbit. In 1921 the writer encountered, by the cranial route, an intracranial tumor that had extended through the optic foramen into the orbital cavity. The orbital roof was removed in order to follow and extirpate this portion of the tumor. So simple was the operative attack, and so perfectly could the intraorbital contents be exposed, that this method suggested a very marked improvement in operative attack upon the great group of intraorbital tumors. No matter in which direction the intracranial portion of the tumor extended, or in which part of the orbit the tumor was situated, this approach appeared to be far superior to the usual frontal or lateral (Krönlein) routes by which these tumors had heretofore been attacked.

The present work describes the results since obtained with this operative approach. Even for tumors confined to the orbit alone, this approach has been found far superior to those hitherto used by ophthalmologists. The great advantage of this approach lies in the much fuller and safer exposure of the intraorbital contents. The optic nerve, the eyeball, three of the extraocular muscles, the ophthalmic veins and arteries can be well exposed and avoided during the dissection of the orbital tumor.

Since the original publication of this procedure, this

operative approach, which is identical with that used for hypophyseal tumors, has been greatly reduced and simplified, owing in large part to the introduction of avertin as the anesthetic. It has been found that swelling of the brain, so commonly associated with ether anesthesia, is avoided when avertin is used, and for that reason the size of the bone flap necessary for the exposure of the base of the brain can be greatly modified.

From the clinical findings on patients with exophthalmos it is not usually possible to tell whether or not there is an intracranial extension of the orbital tumor. And too often the orbital tumor is but a small fraction of the large but silent intracranial tumor which is usually the primary growth. Since intracranial tumors are present in approximately three-fourths of the orbital tumors in this series, it should be assumed on the law of probabilities that the tumor extends into the cranial chamber. And anything less than the combined intracranial and intraorbital approach will offer no surgical solution of the problem. All too frequently the orbital contents have been exenterated in a radical attempt to remove the tumor from the front of the orbit, with an infected granulating wound resulting. Such a result forever precludes the intracranial approach because of the certainty that infection will follow. In safe hands the intracranial approach involves very little risk and offers the maximum hope of cure, unless the frontal bone is involved, and is without cosmetic defects. Even when the character of the tumor prevents a permanent cure, the maximum period of relief and extension of life is afforded by subtotal removal of the extensive tumor.

Although the best available evidence of an intracranial component of the tumor is obtained from the roentgenograms, this proof is often lacking. The optic foramen is

occasionally enlarged or reduced in size, but the vast majority of tumors are not continuous through this opening. The dural tumors, which comprise almost one-half of the series, usually but not always show a diffuse hyperostosis of the walls of the skull and orbit. In these cases there is usually no gross defect in the orbital roof, but the extension of the tumor is due to diffuse invasion of the bone. At times calcification may be detected in the intracranial part of the tumor, entirely unsuspected from the clinical data; or a bony prominence may indicate an underlying growth. In none of the tumors has enlargement of the sphenoidal fissure indicated intracranial participation in the tumor.

From symptoms alone, intracranial extension of the tumor would have been suspected in perhaps two cases. In Case 12 there was headache but no papilledema, and in Case 14 there were uncinate attacks.

The following case, in which operation was performed nearly twenty-two years ago, is presented because it first suggested the procedure now in use. We were then deterred by theoretical possibilities, since found to be of no practical concern. It was feared that a pulsating exophthalmos would follow removal of the orbital roof, but this has never resulted in any of the subsequent cases operated upon by this method.

CASE 25

D. D., History numbers 45901, 46579: A twenty-eight-year-old white woman consulted me on July 1, 1918, because of severe headaches, presumably related to an osteoma on the right side of the head producing exophthalmos and marked depression of the orbital contents. She had been operated upon by Dr. W. W. Keen, of Philadelphia, thirteen years earlier, at the age of fifteen. He had explored and removed part of the

orbital roof, the approach being beneath the supraorbital ridge. He had continued his incision over the temporal region and removed some of the outer part of the skull, which was also greatly thickened. Since that time her deformity had gradually progressed and her headaches had become quite severe. The exact date of onset of the bony growth is not known, but she thought it was first noticed at about the age of six.

There was then a marked proptosis and downward displacement of the eyeball. The X-rays showed a marked thickening of the frontal bone and of the roof of the orbit (Fig. 1).

An intracranial approach was made on August 17, 1918. The bone was exceedingly thick over the supraorbital ridge, so thick in fact that it was necessary to groove the bone with the rongeurs before the DeVilbiss blade could be used to cut it. The bone was soft and spongy. When the fluid was evacuated from the cisterna chiasmatis, a finger-like osteophyte projected backward from the lesser wing of the sphenoid for a distance of perhaps 2½ cm. It was a hard, bony growth, arising with a broad thick base which extended around the optic foramen. This tongue of bone was chiseled away at its base and completely removed. The hard growth was in marked contrast to the soft bone encountered elsewhere. The roof of the orbit did not appear to be abnormal; this could only mean that all of the thickening was from the orbital surface of the orbital roof.

On September 16, 1918, a downward growth from the orbital roof was chiseled away from beneath, the incision being along the supraorbital ridge. The roof of the orbit remained intact. In reviewing the operation this comment was made: "Removal of the roof of the orbit from above was not considered seriously because of the deformity which

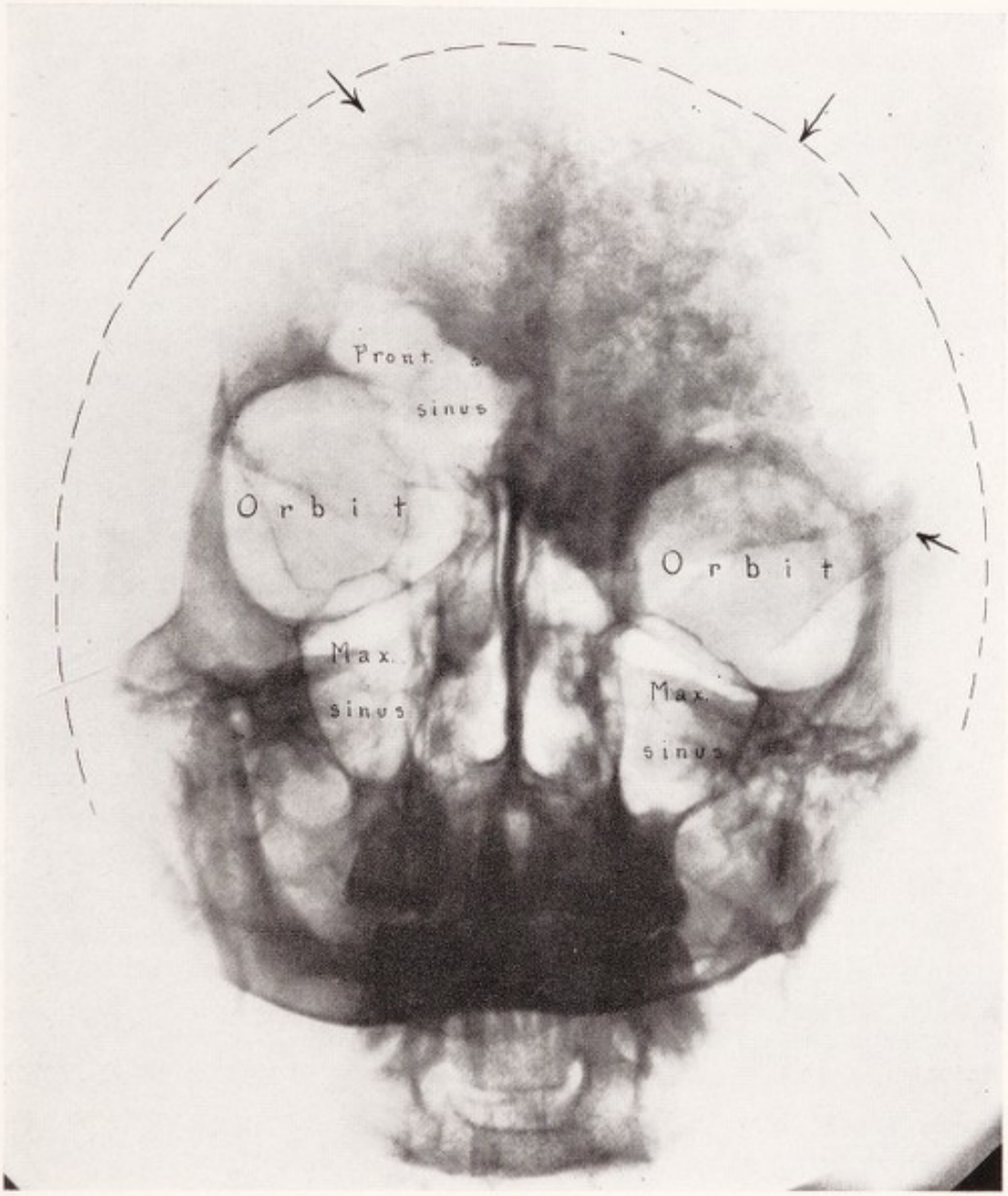


FIG. 1. Case 25. X-ray of diffuse osteoma (cf. Figs. 2 and 3), which has obliterated right frontal sinus. Entire right orbit is placed lower than the left. The antrum is also lower, almost suggesting a congenital displacement of these cavities.

would result by the gravity of the unsupported brain, and because all of the orbital growth was in the orbit. For these reasons, and also because of the very slow growth of the tumor and its diffuse nature, it seemed wiser to attack the orbital roof from below" (Fig. 2). A concavity resulted

FIG. 2

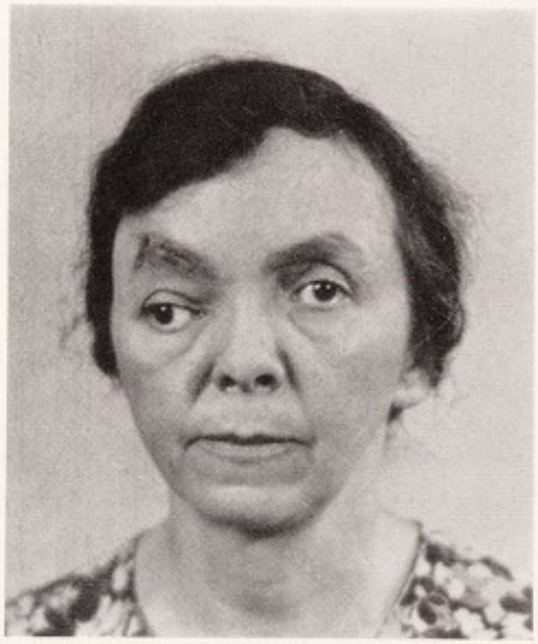


FIG. 3

FIG. 2. Case 25. Diffuse osteoma of the skull and orbit with exophthalmos and downward displacement of the eye (cf. Figs. 1 and 3). This picture was taken after removal of an osteophyte from the lesser wing of the sphenoid and chiseling of undersurface of orbital roof. (Photograph 21 years prior to operation for osteoma.)

FIG. 3. Same patient after operation. The time interval demonstrates the very slow growth of the benign diffuse osteoma.

in the orbital roof after chiseling away a large mass of this bone.

Eight years later the patient had no headache. The proptosis of the right eye persisted, though to a lesser degree than observed before the operation. The patient was alive and well in January, 1940 (Fig. 3). She has been free of headaches since the operation and has carried on her activities as a

nurse without restriction. Microscopic sections showed pure osteoma.

Comment: This would have been an ideal case for the resection of the roof of the orbit by the frontal approach. The cosmetic result would undoubtedly have been far better. Fortunately the bony growth was not associated with a tumor of the soft parts, and there has been no evident progression of the deformity in the succeeding twenty-two years. It is interesting that the removal of the little osteophyte projecting from the wing of the sphenoid abolished the headaches.

CHAPTER I

MISCELLANEOUS TUMORS OF THE ORBIT

In the following cases of orbital tumors, the operations have been by the cranial route (Fig. 4), occasionally with modifications dependent upon the size and position of the growth. They have been grouped according to the character of the lesions and not chronologically.

CASE 1

B. M., History number U-73917. White, female, age 39. Admitted: October 26, 1936. Discharged: November 7, 1936.

Diagnosis: Chronic inflammatory nodule.

Complaint: Protrusion of the right eye.

Family History: Negative.

Past History: Negative, except that patient had gallstones.

Present Illness: The onset of her present trouble was rather difficult to determine. There had been steady pains over both eyes for nearly a year and during this time increasing fatigue, general weakness, and pains over the entire body, particularly in the arms and shoulders.

Seven months ago (March 21, 1936) she had consulted an ophthalmologist because of the pains in the eyes and he is said to have found edema of the right upper lid. A month later (April 26) he palpated a small nodule along the orbital ridge; by May this was about one inch in diameter. There was edema of both eyelids, which were greatly swollen, red, and tender; the eye was completely closed. The diagnosis at that time was an infection of the lachrymal glands. By

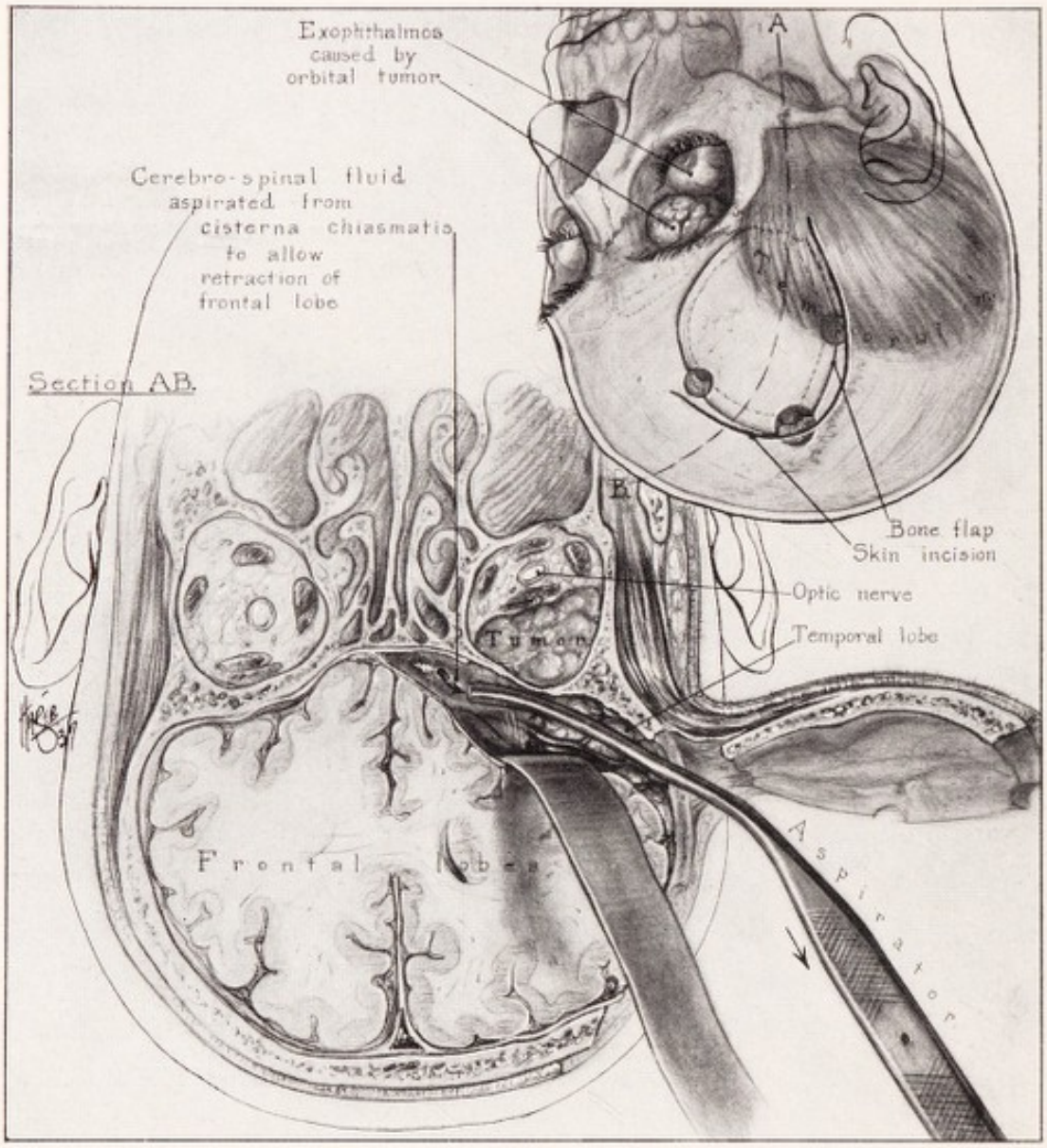


FIG. 4. Sketch showing approach to, and exposure of, roof of orbit for operation by intracranial route upon tumors within the orbital chamber. Upper drawing shows site of cutaneous incision beneath hair line, and relative size and position of bone flap. Lower drawing shows method of depressing frontal lobe and opening cisterna chiasmatis. Release of its fluid provides ample room for attacking roof of orbit, which is then chiseled away.

June 23, the nodule was scarcely palpable, but thereafter a slowly progressive protrusion of the eyeball, but no diplopia, developed. There remained a sharp darting pain over the right eye and the right frontal region; sometimes it extended over the entire side of the head.

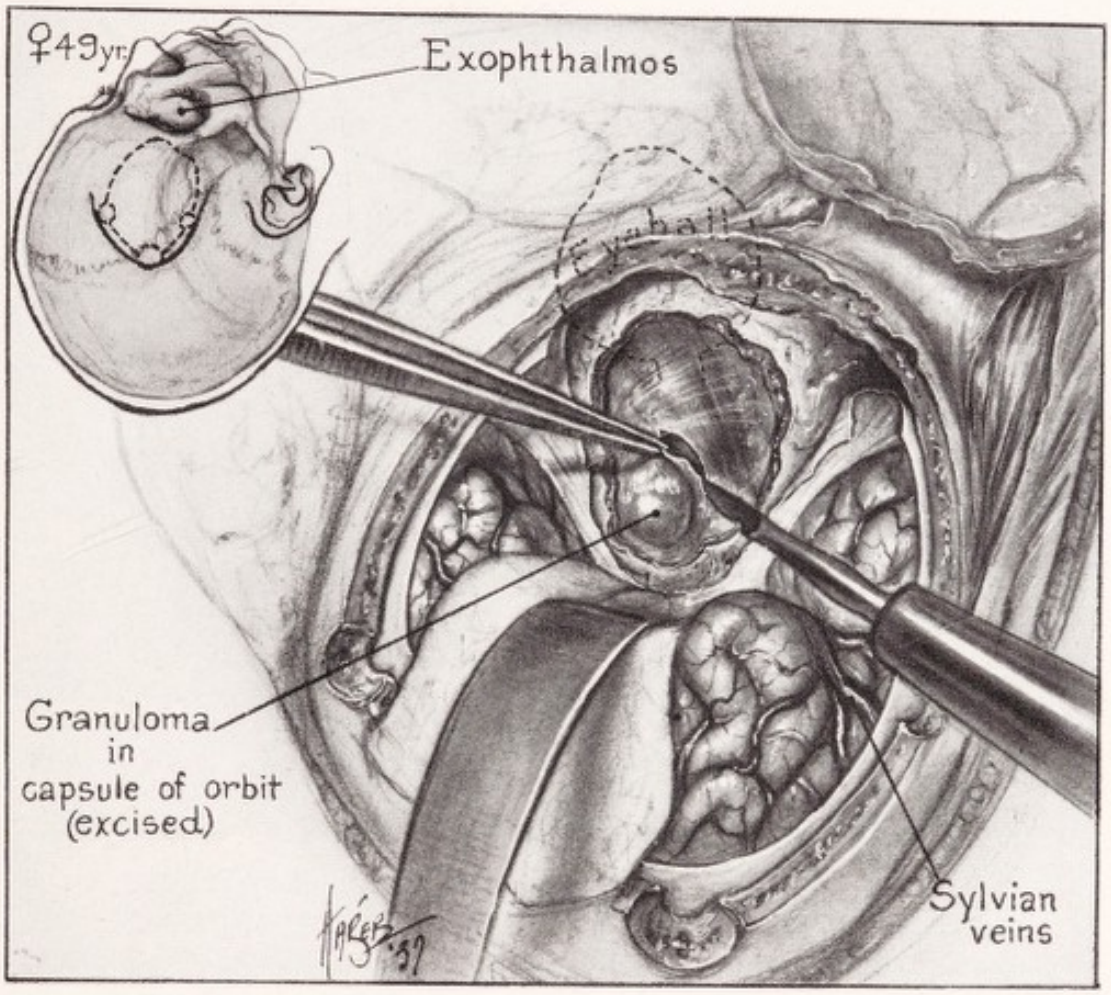


FIG. 5. Case 1. Operative exposure of inflammatory nodule in orbital capsule being removed by excision.

Examination: Negative, except for the local condition in the eye. The exophthalmometer showed 21 mm. in the right eye and 17 mm. in the left. There was no limitation of the extraocular movements. The cornea and conjunctiva were clear. The vision was 20/30 in each eye; the fundi, discs, and retinal vessels were normal. X-rays of the head and of the optic foramen were normal.

Probable Diagnosis: There were reasons for suspecting an inflammatory mass; orbital tumor was a second diagnosis.

Operation, October 28, 1936: A hypophyseal approach was made on the right side, the concealed incision being used. After evacuation of the cisterna chiasmatis, the dura was stripped from the roof of the orbit and a large part of the orbital roof removed with the rongeurs. One could then

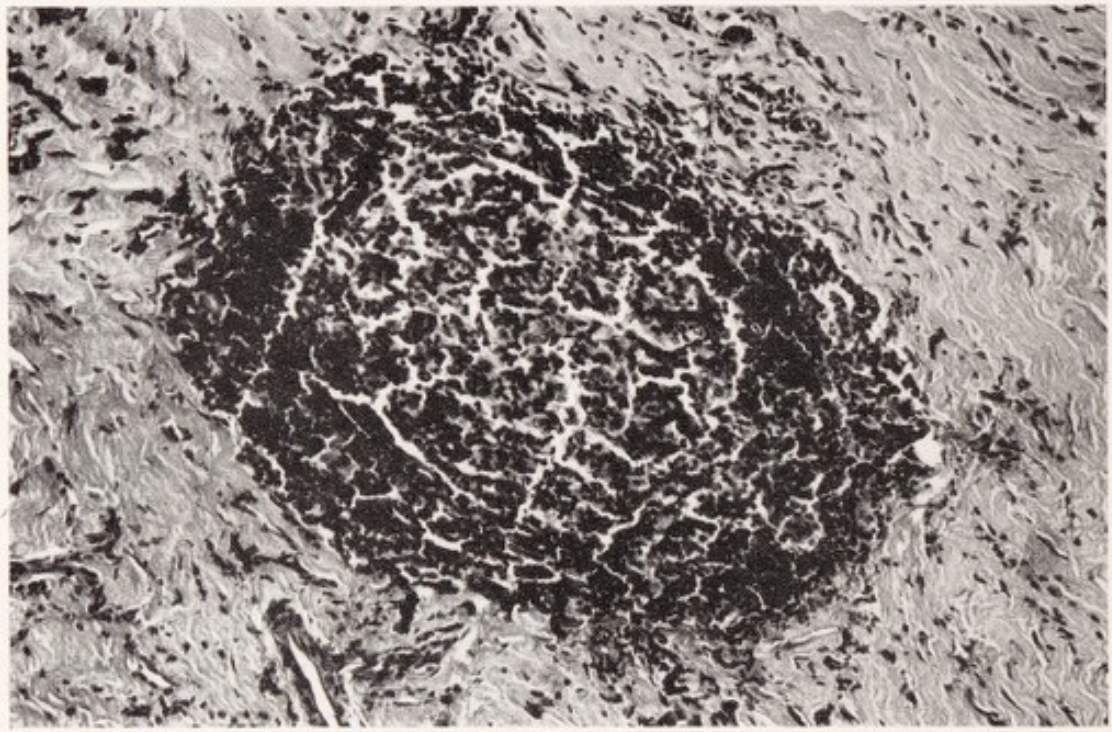


FIG. 6. Case 1. Photomicrograph of inflammatory nodule, showing cluster of small round cells of chronic inflammatory type.

feel a hard mass, not larger than a pea, in the capsule of the eyeball and just in front of the optic foramen. Its position was not far from the ethmoidal sinus, but this sinus was not seen. The orbital capsule was then opened and the hard and well-circumscribed mass excised with a good margin of normal tissue (Fig. 5). At the time we were not sure that it was not a small metastatic carcinoma, but the frozen section showed it to be inflammatory tissue. The superior rectus muscle was exposed when the tumor was excised, but

the orbital fat did not bulge. This assured us that there could be no additional tumor within the orbit.

Subsequent permanent sections confirmed the diagnosis of chronic inflammatory tissue (Fig. 6). Doubtless the mass was part of the generalized inflammation eight months earlier, when the eyelid became swollen and edematous and the mass was palpable along the supraorbital ridge.

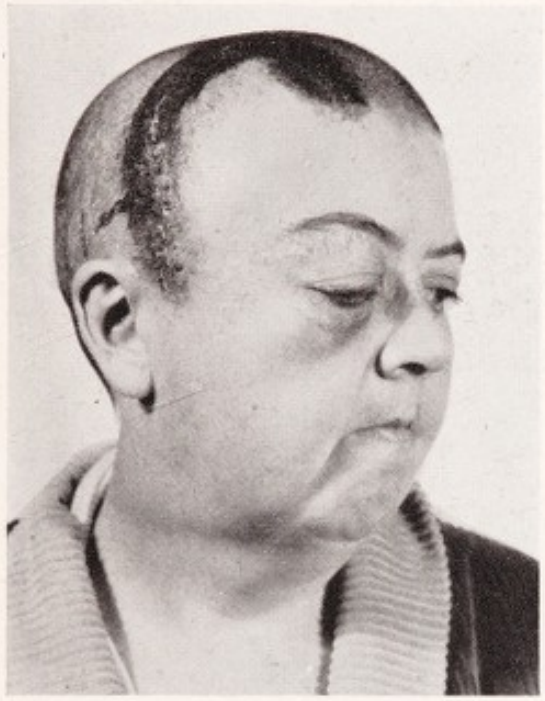


FIG. 7

FIG. 8

FIG. 7. Case 1. View of scar following operation, also showing low grade of exophthalmos of right eye.

FIG. 8. One and one-half years later, showing complete correction of exophthalmos.

Recovery was uneventful. The patient left the hospital twelve days later. At this time there was very slight protrusion of the eye (Fig. 7). The vision in each eye was 20/20.

Microscopic Report: "Chronic inflammatory tissue. In much of the section the fibrous tissue is solid, but in places there are areas of closely packed small round cells, either in irregu-

lar masses or in circular formation. Although there is a suggestion of tubercle formation, no giant cells can be seen."

Subsequent Course: The patient was last seen January 7, 1938 (Fig. 8). Dr. Angus MacLean found no exophthalmos. The vision and visual fields were normal. One could not tell which had been the affected eye. The patient had no headaches and could now see the movies without fatigue and headache.

CASE 2

F. D., History number U-66836. White, female, age 33. Admitted: December 23, 1935. Discharged: January 6, 1936.

Diagnosis: Pure fibroma.

Complaint: Protrusion of the left eye.

Family and Past Histories: Negative.

Present Illness: Eight months before, beginning protrusion of the left eye was observed. Six weeks after the onset the patient was seen by Dr. Alan Woods in the Department of Ophthalmology. She said the protrusion had been steadily increasing; that it was greater in the morning and receded during the day. There had been no pain or discomfort. At that time her vision was 20/20 in each eye; the pupils were active and reacted to light and accommodation. The exophthalmometer registered 23 mm. in the left eye and 15 mm. in the right.

Five months later she returned to Dr. Woods, and at this time the exophthalmos had greatly increased. The eye was pushed downward and outward and a solid tumor could be palpated behind the eye. For the past six months headaches had been almost constant. The exophthalmometer now registered 29 mm. in the left eye, an increase of 6 mm. in six months. The eyelids could not cover the globe. The palpebral conjunctiva was injected. Extraocular move-

ments were well performed and the tension of the eyeball was normal to the palpating finger. The disc was not elevated, but the nasal margin was blurred. The veins were somewhat engorged and tortuous. Vision was 20/200.

The patient was given deep X-ray therapy over a period of one month, and at the end of that time the vision in the good eye (right) was found to be badly affected. The visual acuity was then 20/300. On the left the visual fields had

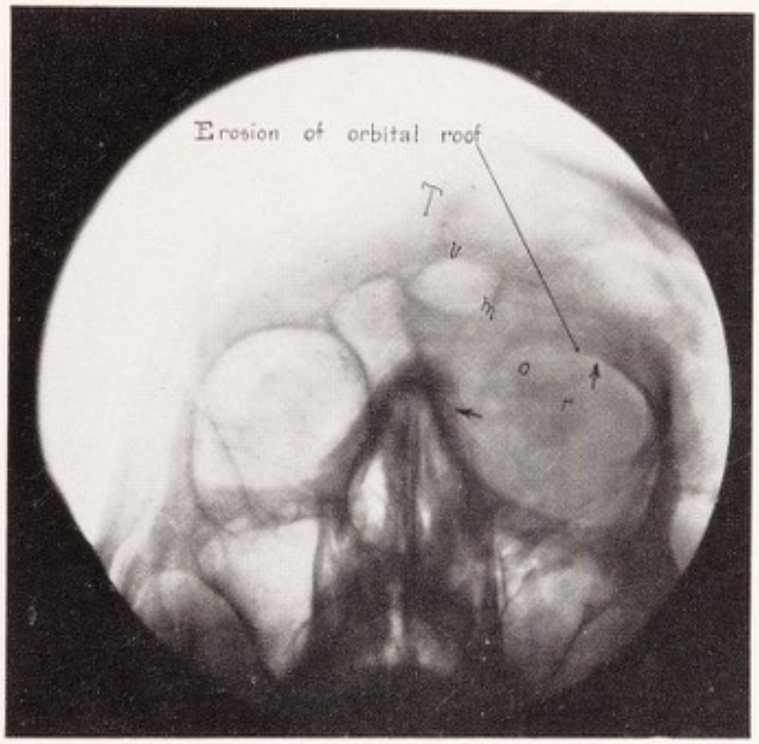


FIG. 9. Case 2. Retouched X-ray showing destruction of roof of orbit and anterior wall of skull by tumor.

decreased until only light perception remained; a small field of vision was left above the midline and both to the right and left of the vertical line. One week later the vision in the right eye was nil and in the left 4/200, with a still further reduction in the visual fields and complete loss of color perception.

I saw the patient toward the end of December, when there was but little remaining vision, and felt that because of the

tremendous exophthalmos and the rapid loss of vision the tumor was probably a malignant sarcoma, and advised against operation. Only through the insistence of Doctor Woods was the operation finally performed. The X-rays then showed marked destruction of the left orbital roof,

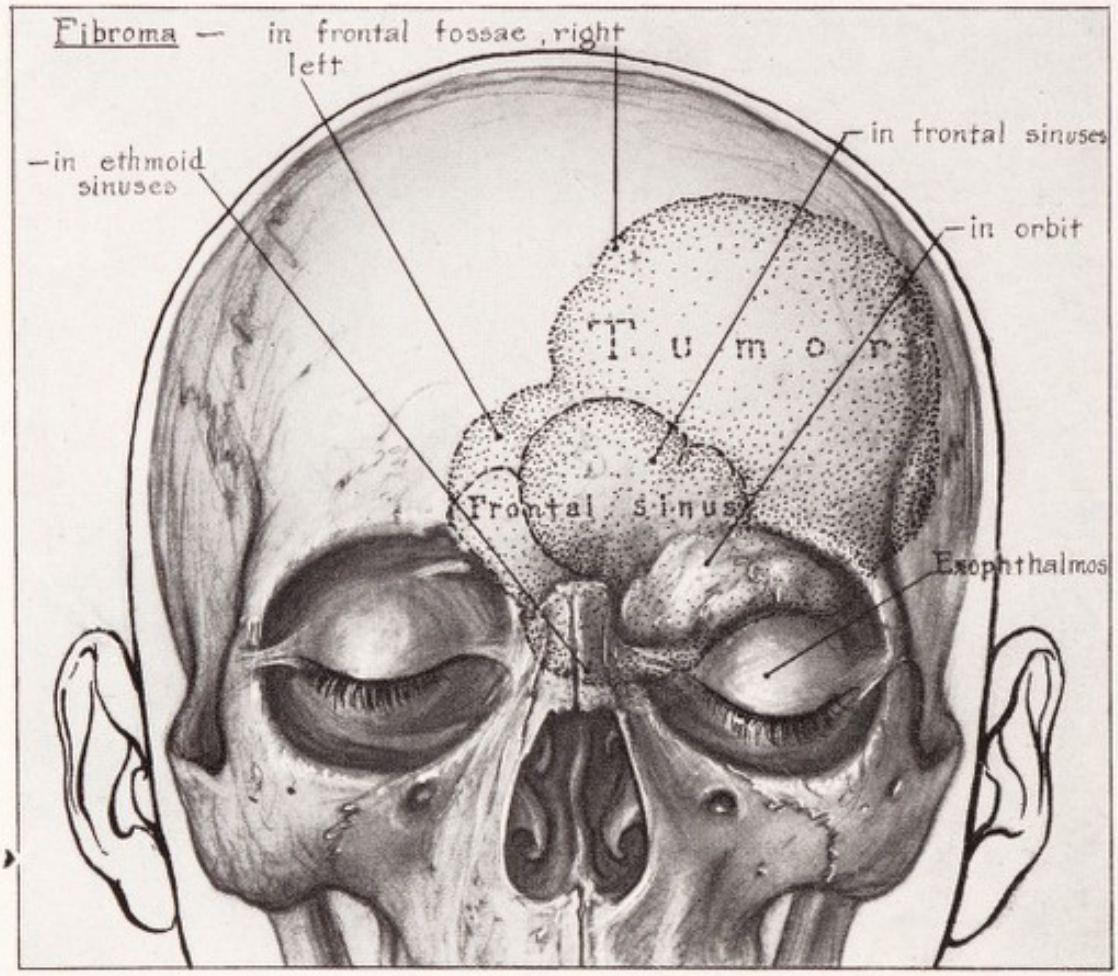


FIG. 10. Case 2. Sketch showing position of tumor, which had filled both frontal sinuses, destroyed roof of orbit, and invaded a large part of orbital chamber.

some on the right also, and massive destruction of the frontal bone (Fig. 9). The X-rays on the first admission six months earlier had been negative.

Operation, December 24, 1935: The usual hypophyseal approach with concealed incision was made on the left side. When the bone flap was elevated one could see an enormous

hard fibrous tumor pushing back the dura covering the frontal lobe. It extended over about two-thirds of the area covered by the bone flap and it was all extradural (Fig. 10). Everywhere over the tumor surface there were patches of thin bone, probably representing remains of the roof of the orbit. The tumor was one of the hardest fibrous growths I have ever seen. It was so big, so snugly bound to the dura and bone, and the convexity of its surface such that in efforts to enucleate the mass, the finger could scarcely make any headway. It was necessary to split the tumor near the middle and first remove the convex posterior portion, then separate the anterior part from the frontal bone (Fig. 11). The growth was so dense that it was difficult to cut with the knife and scissors; its section was quite bloodless. The tumor had grown across the midline, destroyed the cribriform plates and the roofs of both orbits, and filled both the right and left orbital cavities. A heavy toothed clamp grasped the border of the tumor, and by traction it was drawn forward and stripped from its bed with the periosteal elevator. The tumor was stripped from the eyeball, but it pushed deeply into the left orbit and into all the paranasal sinuses. The surface of this part of the tumor was quite nodular; the nodules pushed ahead in different directions, each apparently seeking its path of least resistance. Separate nodules were deeply wedged in the bony crevices of this region. Gradually the tumor was lifted away with the periosteal elevator and without breaking, because it was firm throughout. At one place an area of the dura which was firmly bound to the growth had to be removed with it. One could look directly into the ethmoidal cells. The danger of infection with an open dura was, of course, evident, and to obviate this risk a piece of fascia lata was painstakingly sutured to the margins of the dural defect. We were not conscious of an injury to the optic nerves.

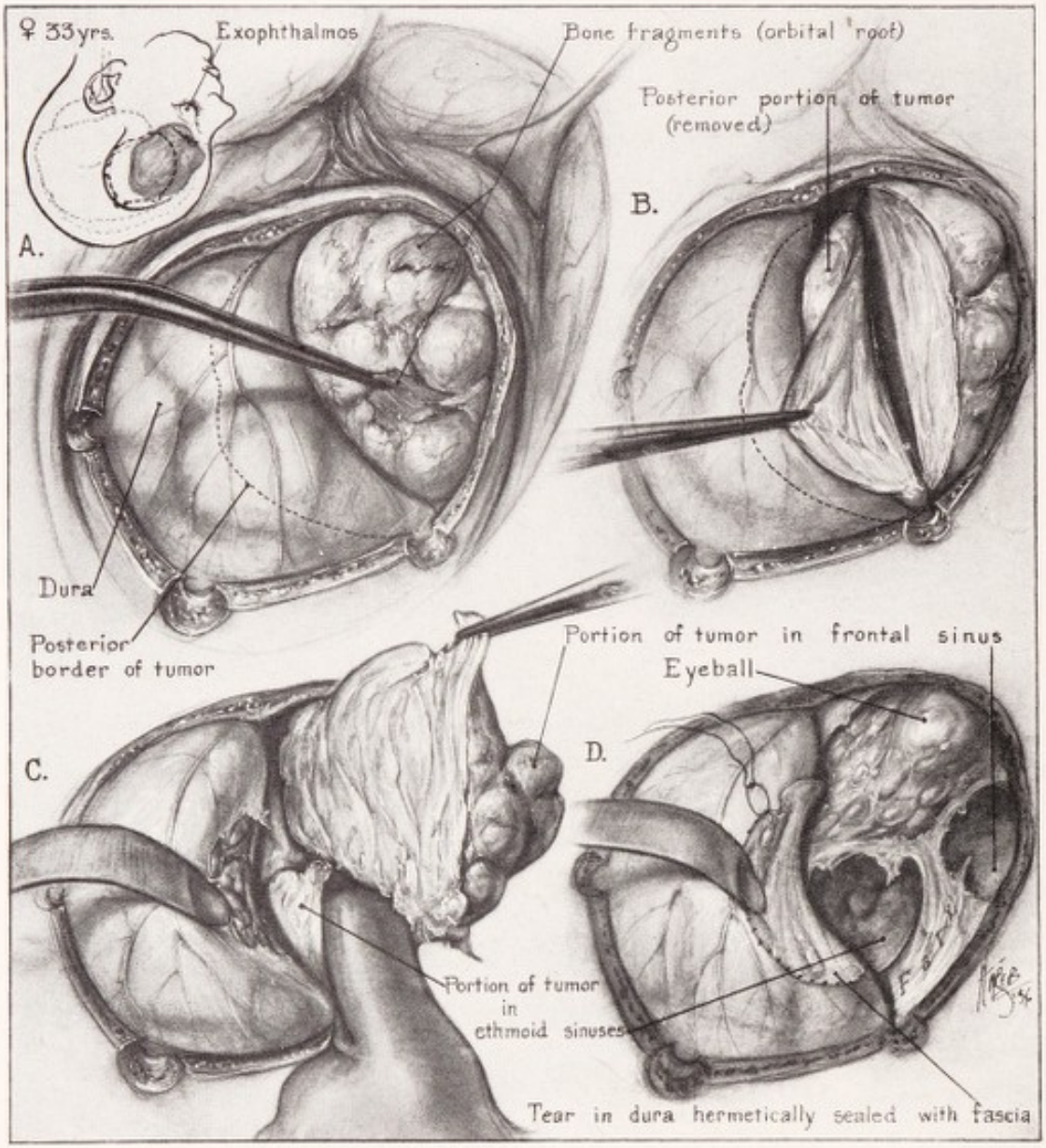


FIG. 11. Case 2. Operative sketch showing position of tumor, which was entirely outside the dura. It had projected across midline into opposite (right) anterior fossa destroying falx enroute.

On the following day serosanguineous fluid escaped through the nose. This was assumed to be the fluid that had accumulated in the large, wide-open epidural space. On the following day this had stopped and there was no subsequent rhinorrhea.

The weight of the tumor was 131.2 grams (Fig. 12).

The patient made an uneventful recovery from the operation, and without infection. She left the hospital on Jan-

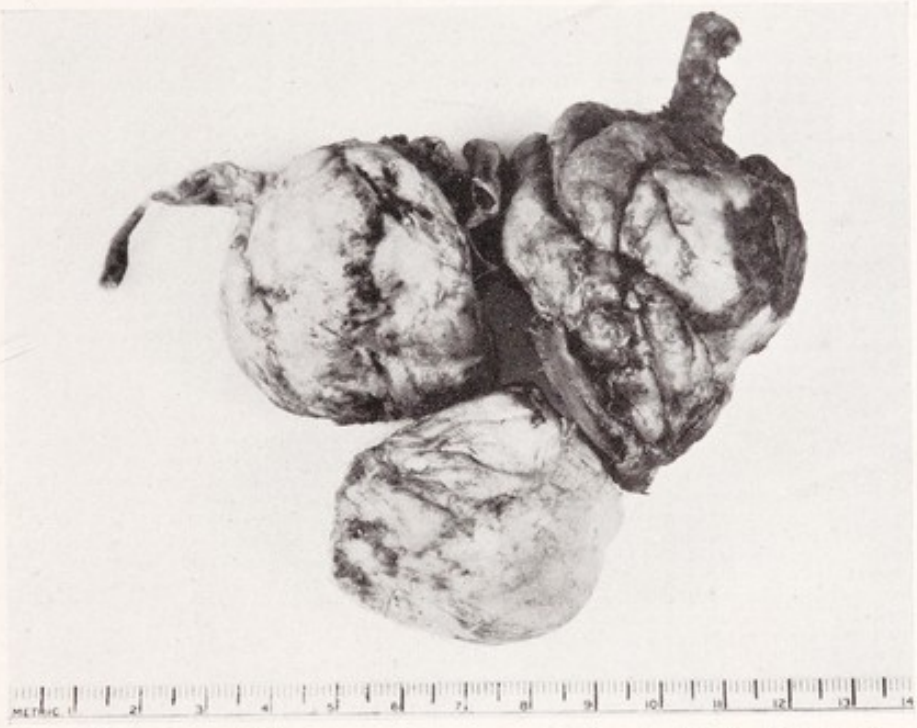


FIG. 12. Case 2. Tumor after removal: a pure fibroma, possibly arising from outer sheath of dura.

uary 6, 1936, two weeks following the operation. At that time there was a good return of vision in the right eye, but none in the left. There was a fairly normal though somewhat restricted field of vision in the right, with color perception in the central region.

Microscopic Report: "Pure fibroma, with few nuclei" (Fig. 13).

Subsequent Course: A letter from this patient on January

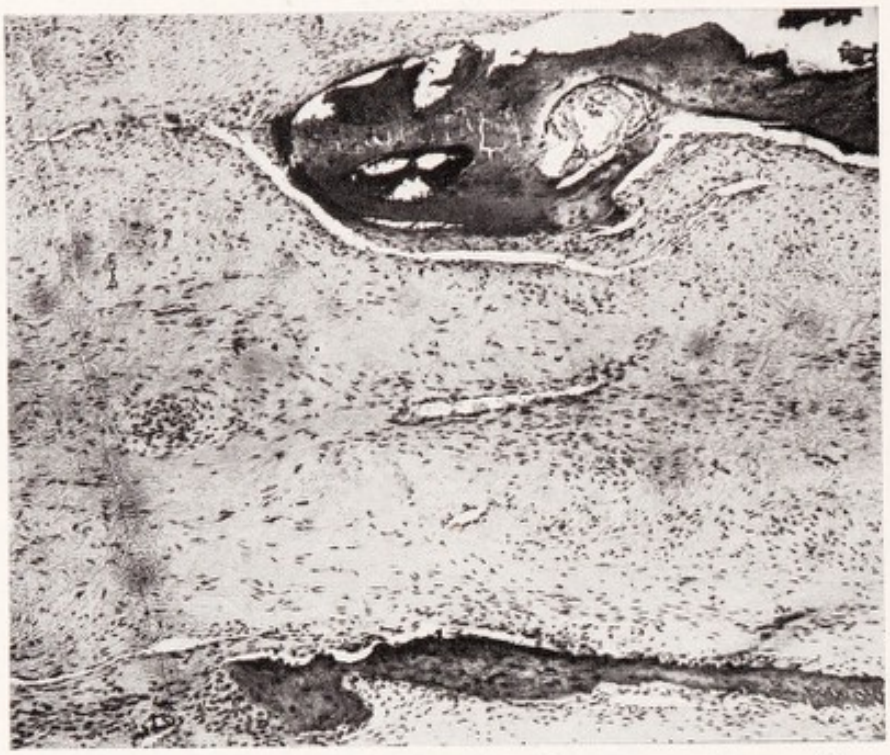


FIG. 13. Case 2. Photomicrograph of tumor of pure fibrous tissue.

FIG. 14. Case 2. Four years after operation. Eyeball completely restored to normal.

15, 1940, stated that she was well, but no note was made concerning her vision, which I think, however, must be normal (Fig. 14).

CASE 3

E. N., History number U-127718. White, female, age 13. Admitted: December 23, 1937. Discharged: January 7, 1938.

Diagnosis: Diffuse osteoma. (Referred by Dr. Wm. F. C. Steinbugler, of Brooklyn, New York, with the diagnosis of an orbital tumor.)

Family and Past Histories: Negative. The mother is in a mental institution where a diagnosis of dementia praecox has been made.

Present Illness: Four and one-half years ago, headaches had appeared. They came only now and then, were not localized, and were not very severe at that time. The patient's eyes were examined and it was then noted that the left eye was protruding slightly and that there was fulness in the left temporal region. There was also a little fulness in the supraorbital region. Everywhere the increased swelling was smooth and diffuse. The vision at that time was better in the left eye than in the right.

One year ago, prisms had been used to exercise the ocular movements. Since then there had been complete relief of headaches. Five months ago (July, 1937) the patient had noticed that the vision in the left eye was markedly impaired, and one month ago vision in the left eye had ceased. Since there was no pain or discomfort she did not mention the loss of vision to her family until a week before admission, and even then it was only casually brought to their attention.

Six months before, when the use of prisms was begun, the vision in the affected eye was 20/30. Despite the displace-

ment of the eyeball, the patient insisted that there had been no diplopia.

Physical and Neurological Examinations: The patient was a normal-appearing girl, except for the local deformity (Fig. 15). The examination of the eyes was made by Dr. Woods, whose note follows:

"There is downward displacement of the eyeball of 6 mm., the exophthalmometer showing 14 mm. right and 20 mm.

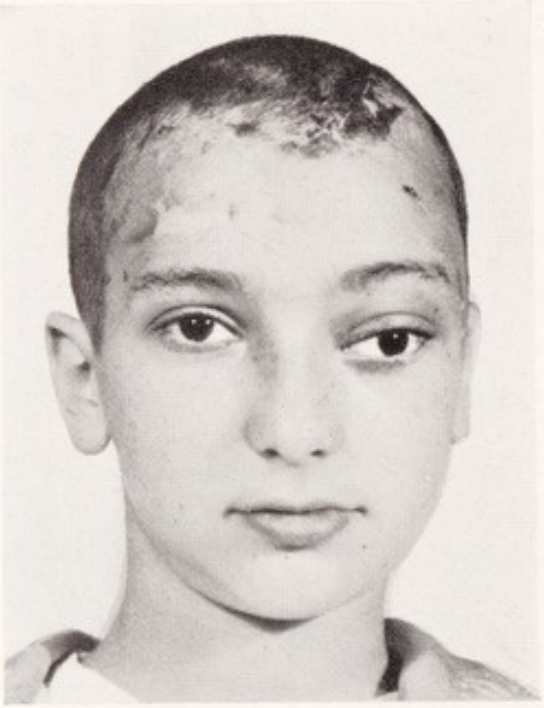


FIG. 15. Case 3. Preoperative photograph, showing downward and outward displacement of eyeball by diffuse benign osteoma of skull and orbit.

left; proptosis of 44 mm. The pupils are equal, the right reacting promptly to light and the left very slowly and sluggishly. The extraocular movements are practically normal. There appears to be a slight limitation of the upward movements. The intraocular tension is normal. The ophthalmoscopic examination reveals slight blurring of the margins of the disc on the right side, with obliteration of the physiological cup. There is a slight overfulness of

the veins. The elevation of the disc is about 1 diopter. The left disc shows a marked generalized pallor, especially in the temporal side. There is some overfilling of the veins and slight haziness of the upper and lower poles of the disc. Vision in the right eye is 20/15. Bare hand movements are visible with the left. The visual fields are normal in the right eye and nil in the left."

The X-rays showed a very marked, diffuse bony thickening of the entire roof of the orbit; it extended to the posterior limit of the orbit and involved the ethmoidal and sphenoidal sinuses mesially. Laterally it involved the side of the skull (Figs. 16A and 17A). The optic foramen was distinctly smaller on the left than on the right (Fig. 18). The diagnosis of orbital osteoma was made from the X-rays.

Operation, December 24, 1937: A left hypophyseal approach with concealed incision was made. The brain was quite full and the dependent position of the head gave very little room. The cisterna was approached with some difficulty because of the upward curvature of the orbital tumor. It was finally reached and a fair amount of fluid was evacuated. The volume of the brain was still much greater than we should have liked for an attack upon the orbit. The frontal lobe was elevated by the spatula protected by pledgets of cotton, and the roof of the orbit was then in full view. There was a pronounced hillock of bone in the posterior two-thirds of the orbit, the anterior one-third looking fairly normal. The dura was much redder posteriorly, where the bone was thicker. The dura was incised at the lateral side of the orbit and along the wing of the sphenoid and separated from the orbital roof by the periosteal elevator. It was rolled mesially toward the cribriform plate. An attempt was made to chisel away the roof of the orbit anteriorly, where the bone appeared to be fairly normal, but it was too thick to be pene-

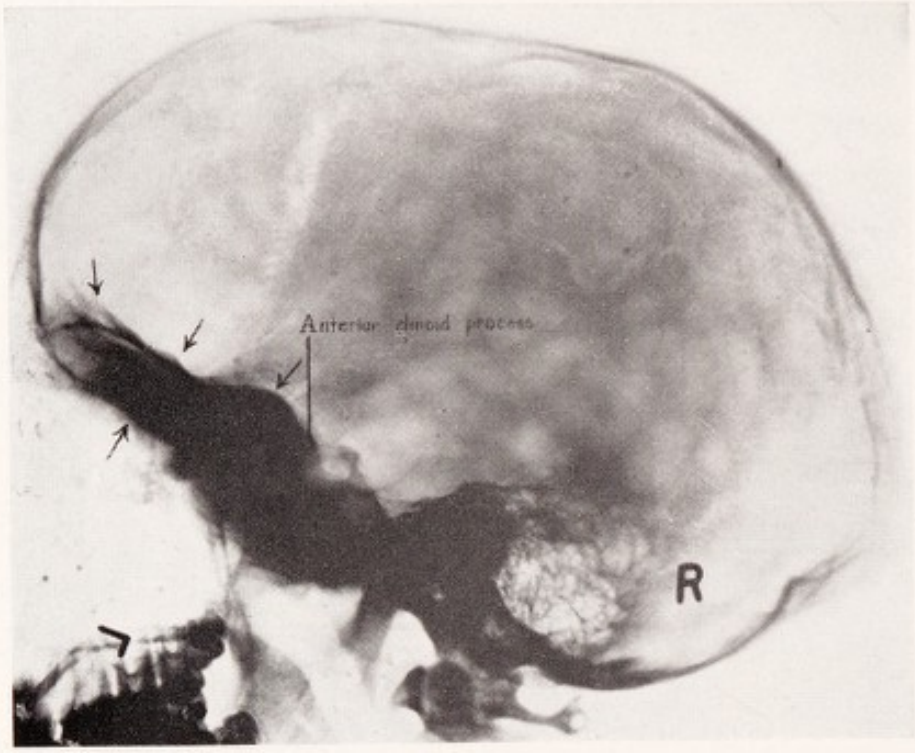


FIG. 16A

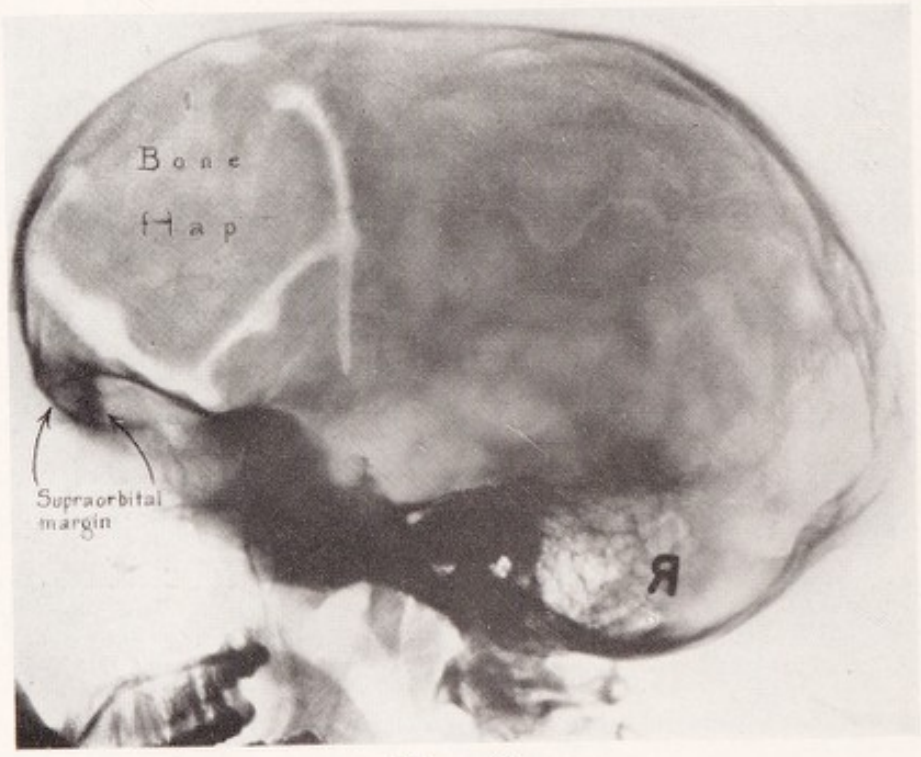


FIG. 16B

FIG. 16. Case 3. Lateral view of skull: (A) showing diffuse hypertrophy of the bone in region of roof of orbit; (B) showing removal of osteoma, also size of operative bony defect.

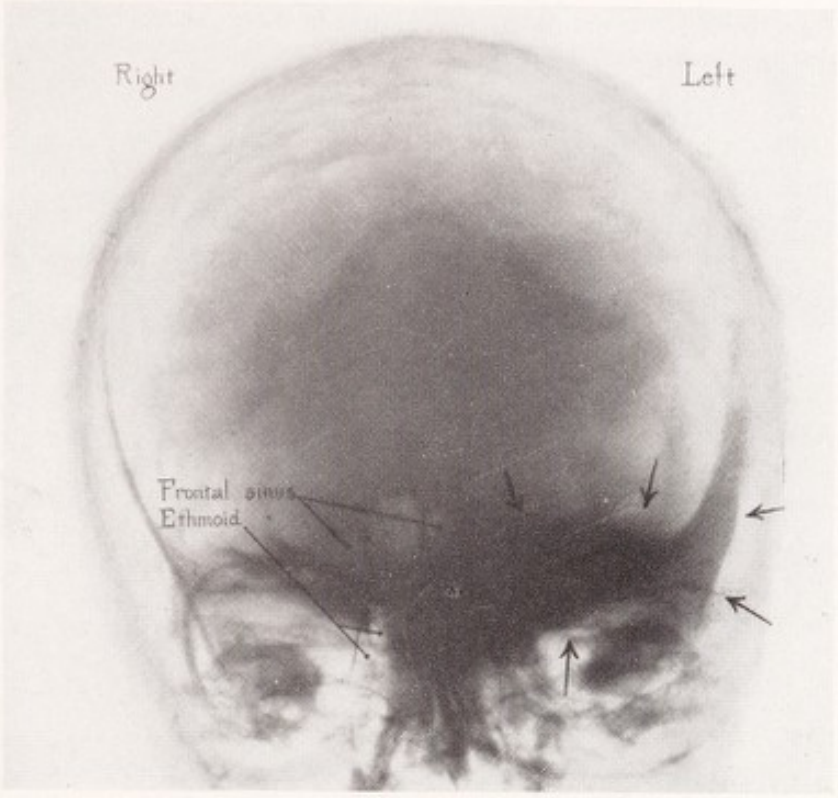


FIG. 17A

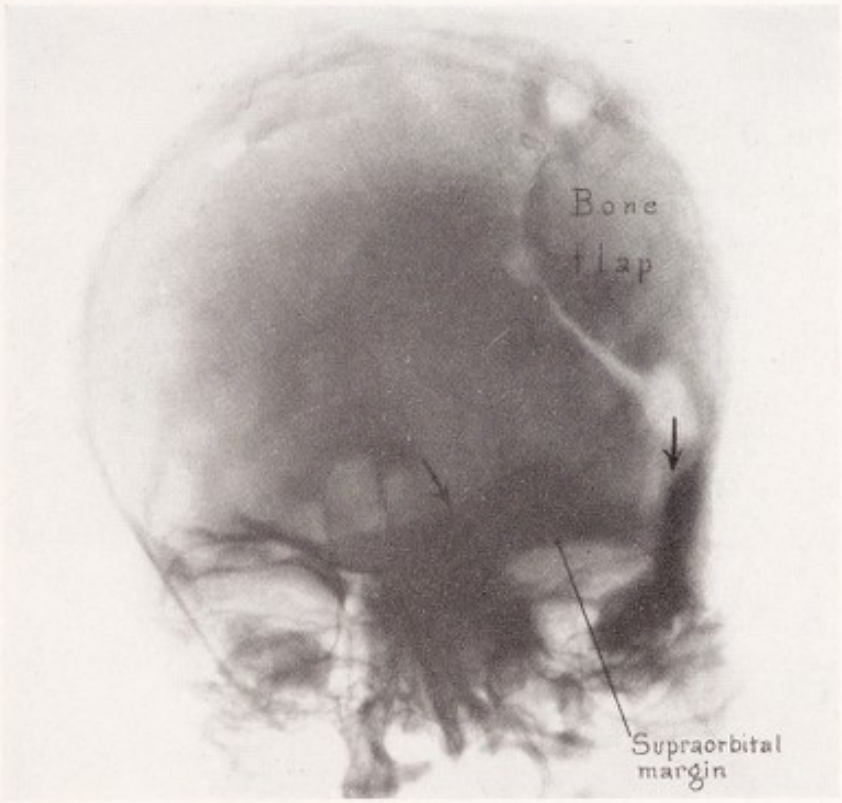


FIG. 17B

FIG. 17. Case 3. Anteroposterior view showing extent of hyperostosis: (A) before operation; (B) after operation, arrow indicating inner limit of operative extirpation.

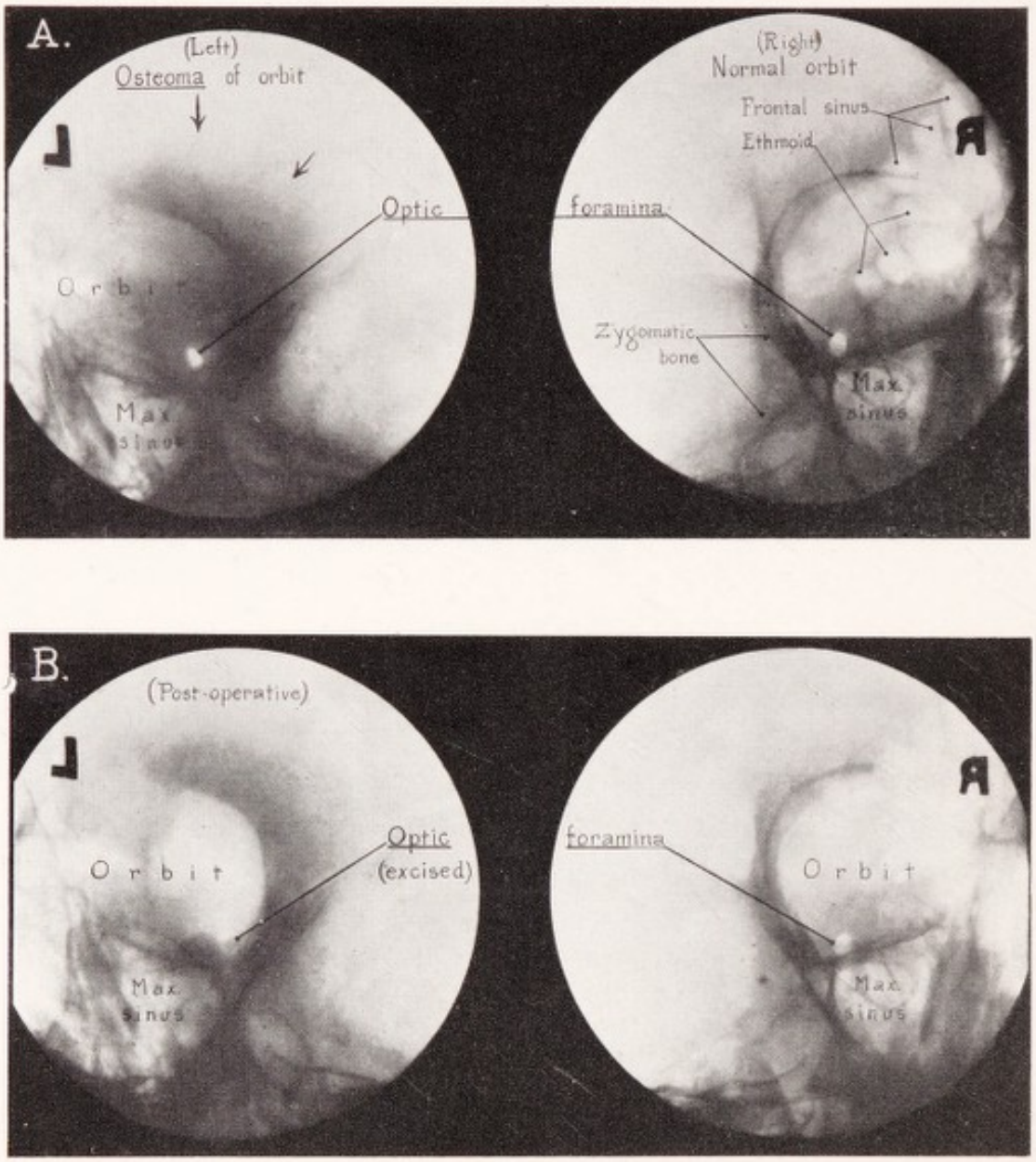


FIG. 18. Case 3. Retouched X-rays of optic foramen on tumor side and on normal side: (A) before operation; (B) after operation. Lateral wall of the foramen on the side involved by tumor was removed.

trated safely. It was evident that the best point of attack was laterally where the bony thickening was confluent with the skull. The inner table of the skull was then chiseled away laterally, until the orbit was entered and, by progressively biting away additional pieces with the chisel, the opening was gradually extended. The entire roof of the orbit was removed piecemeal in this fashion (Figs. 16B and 17B). Anteriorly the bone measured about 2 cm., posteriorly about 4 cm. A thick convex border of the lesser wing of the sphenoid bulged backward into the middle fossa. The anterior clinoid process was a rounded dense mass instead of the usual pointed process. The entire lesser wing of the sphenoid was removed piecemeal (Fig. 21). The projection of the dura containing the third, fourth, and sixth nerves and the ophthalmic vein was isolated when the covering bone was removed. There then remained a mass around the optic nerve. With as little trauma as possible this bone was split over the optic nerve in its longitudinal direction (Fig. 18), and then pried away both to the right and left of the nerve. We had the impression that there was not necessarily any injury to the optic nerve incident to the chiseling process (Fig. 20).

The lesser wing of the sphenoid was removed reluctantly because of the fear of pulsating exophthalmos later, but as the major part of the osteoma was in this region, there was no alternative. Inspection of the tumor removed from the orbital foramen disclosed a deep groove caused by the optic nerve. There was no sign of soft tissue tumor. The bone was chiseled laterally to the innermost part of the orbit, but, since it continued over to the opposite side, its complete removal was not possible (Fig. 19). One of the little ethmoid cells was entered and was at once plugged with wax, and the dura which had been rolled mesially was spread over this region.

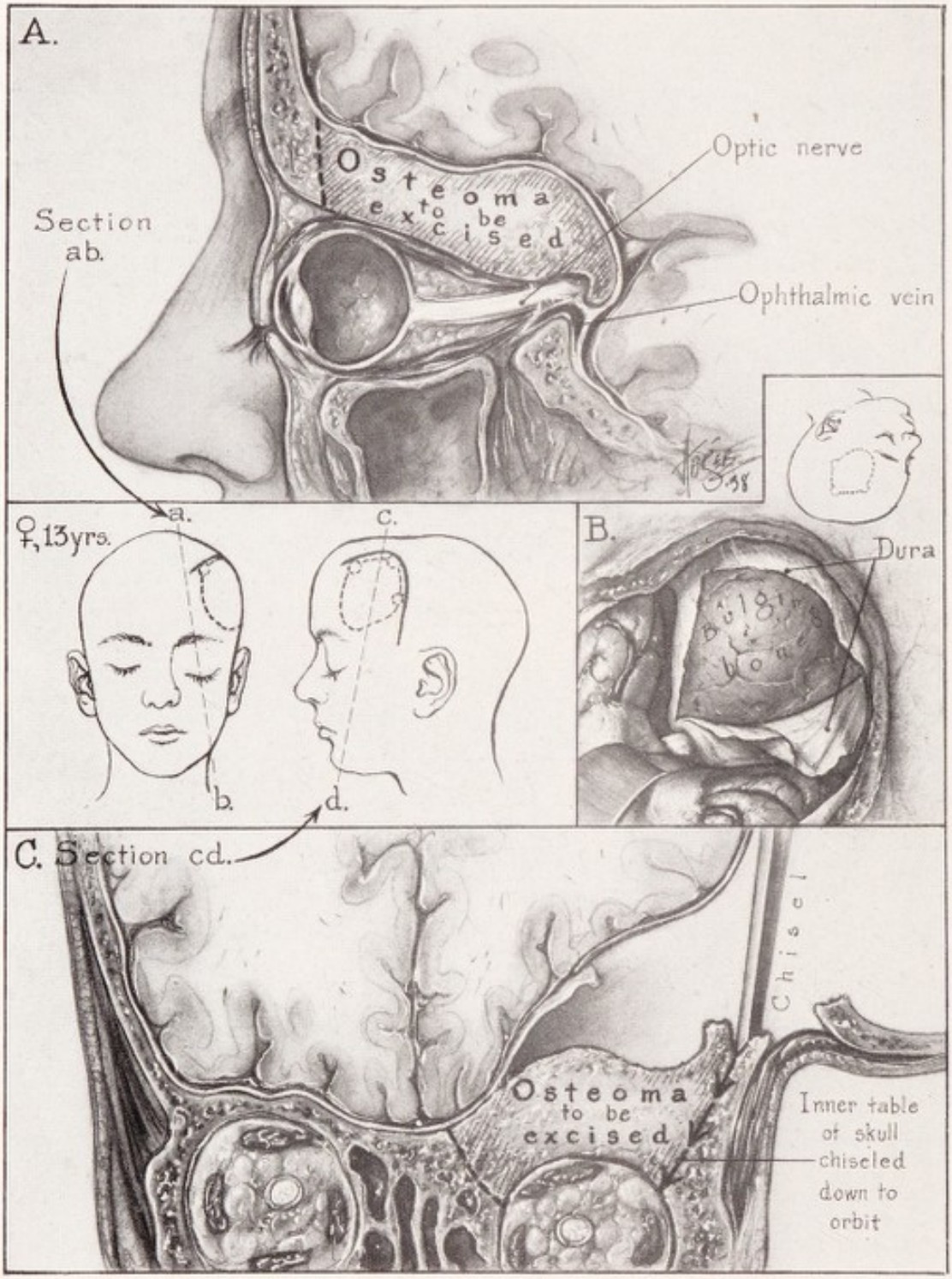


FIG. 19. Case 3. Sketch showing position of osteoma removed at operation. The removal included outer wall of optic foramen and anterior clinoid process.

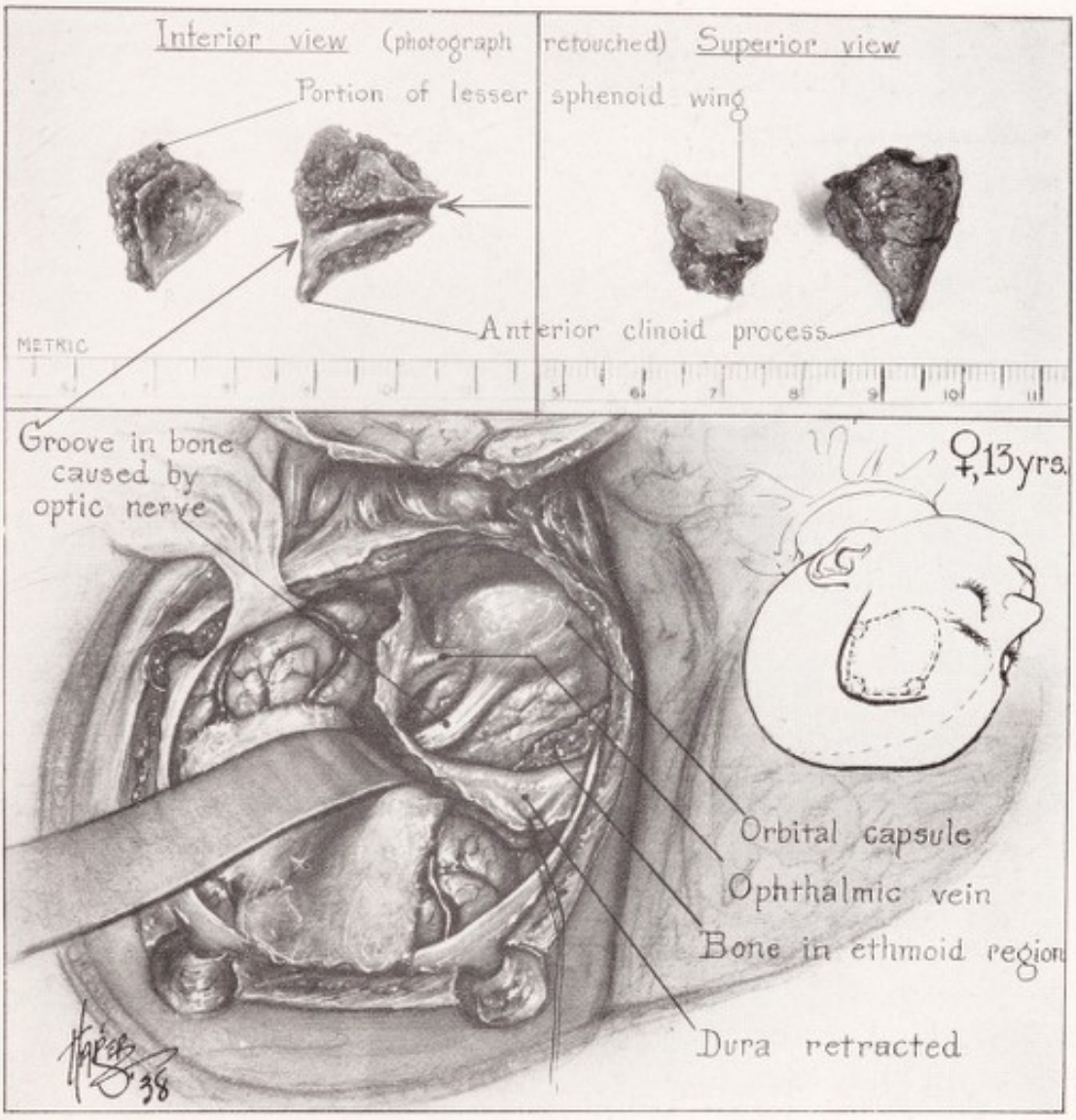


FIG. 20. Case 3. Operative sketch showing defect remaining after removal of tumor from roof of orbit. The optic nerve and eyeball are in full view. Upper inset shows fragments of bone excised with groove of optic nerve and anterior clinoid process.

The orbital capsule was everywhere smooth and glistening and disclosed no suggestion of a secondary soft tissue tumor. The wound was perfectly dry when closure was begun. The bone, being hard and white, was relatively avascular. A few applications of wax controlled the bleeding. There was a little contusion of the orbital surface of the frontal lobe and

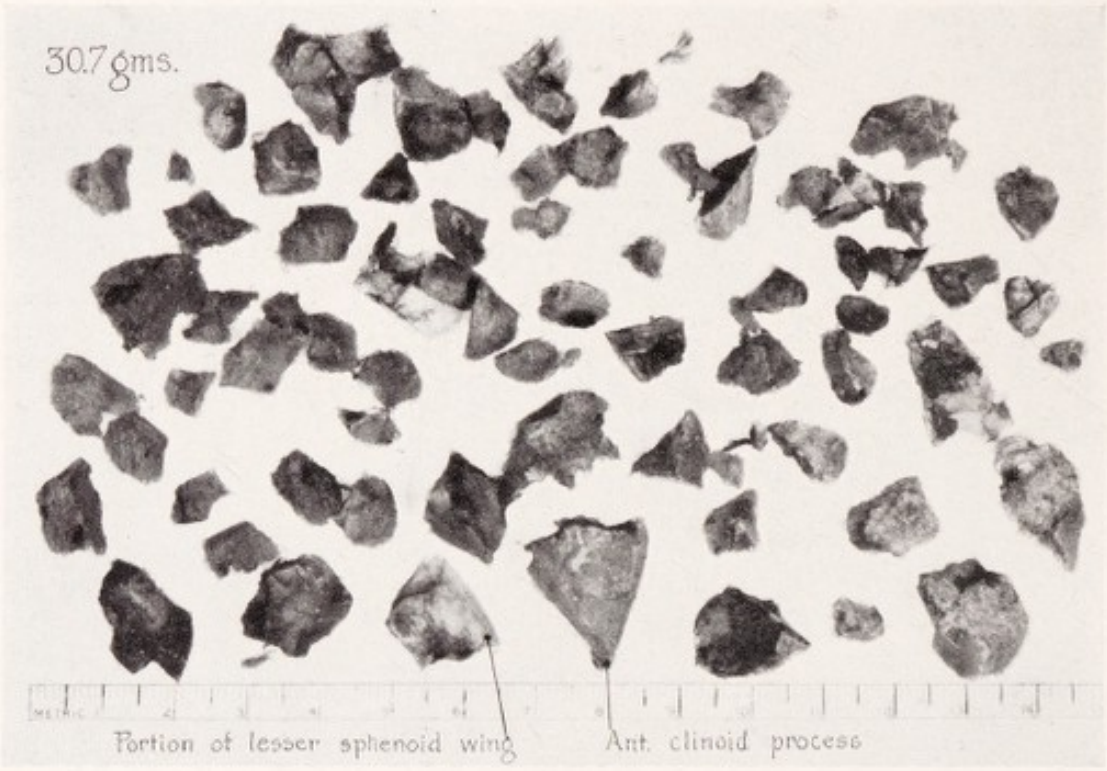


FIG. 21. Case 3. Assembly of bone fragments chiseled away in removing orbital osteoma.

this was removed; this area, about as large as a silver dollar, weighed 9.7 grams.

The postoperative course was uneventful. A little blood stain appeared in the nostril on the left side as a result of opening the ethmoid cell, but at no time was there any indication of infection and at no time a suggestion of rhinorrhea.

Five days after operation the patient could count fingers with the left eye; at least it could be said that the optic nerve

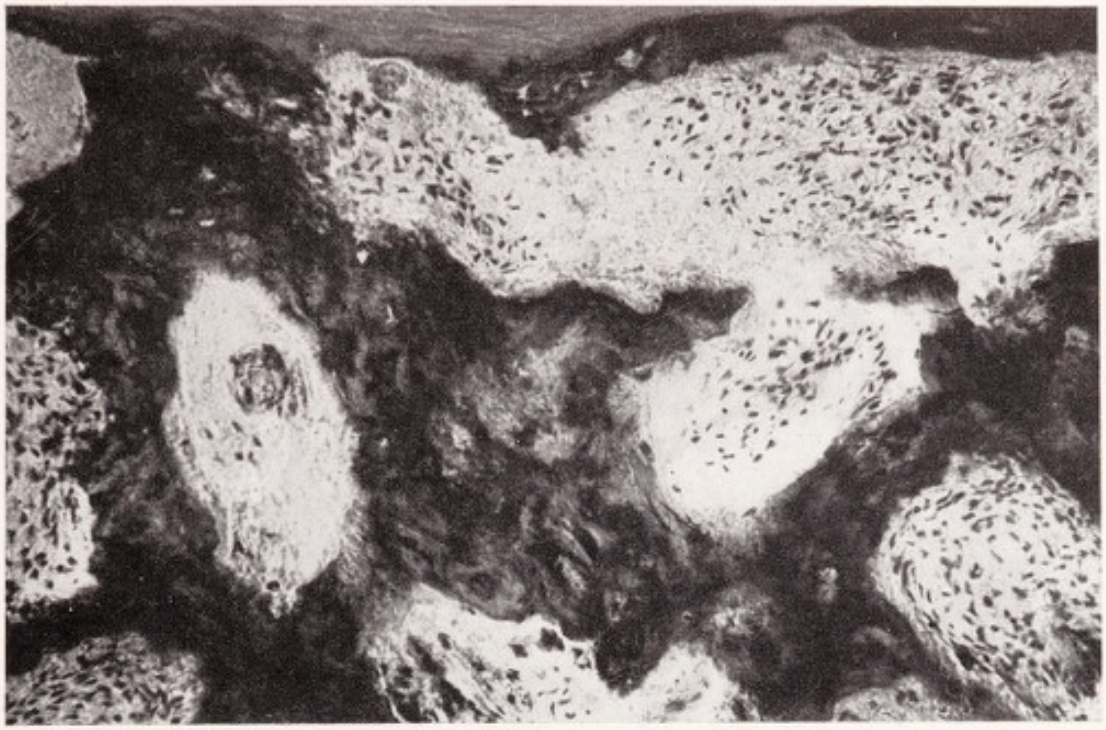


FIG. 22. Case 3. Photomicrograph of tumor: pure fibroma.

FIG. 23. Case 3. Eighteen months after operation.

had not suffered additional injury. The condition remained essentially the same at the time of her discharge on January 7, 1938. We were, of course, eager to learn whether any additional vision could return to the left eye.

Dr. Woods's note at the time of the patient's discharge showed the local picture of exophthalmos and the extraocular movements to be essentially the same as on admission. The eyeball did not pulsate. The postoperative swelling of the eye had disappeared only shortly before, and improvement in the appearance of the eye was expected with time.

Microscopic Note: Pure osteoma (Fig. 22).

Subsequent Course: The patient was seen December 1, 1939 (Fig. 23). There was only a bare suggestion of the old deformity. The vision had not changed. She was well except for occasional headaches, and had been attending school regularly.

CASE 4

D. H., History number 176719. White, female, age 23. Admitted: July 27, 1939. Discharged: August 23, 1939.

Diagnosis: Osteomatous cyst.

Complaint: Something pushing eye out.

Family and Past Histories: Negative.

Present Illness: Five years ago a friend had noticed that the patient's left eye was prominent. During the next four and one-half years the protrusion slowly increased. Six months ago severe frontal headaches had appeared, but could be relieved by aspirin. Four months ago sharp pains had developed in the left temporal region. There was no diplopia, no loss of vision, or pain in the eye. Two different surgeons wanted to operate for thyroid disease but she refused.

Examinations: The patient was a well-developed young woman. Dr. Woods's examination of the eye indicated that the left eye protruded 6 mm. forward and slightly downward (she should have had diplopia). One prominent vein ran across the upper lid. All of the movements of the extraocular muscles were normal. The ophthalmoscopic examination was negative. Visual acuity was 20/40 + in each eye. The visual fields were normal.

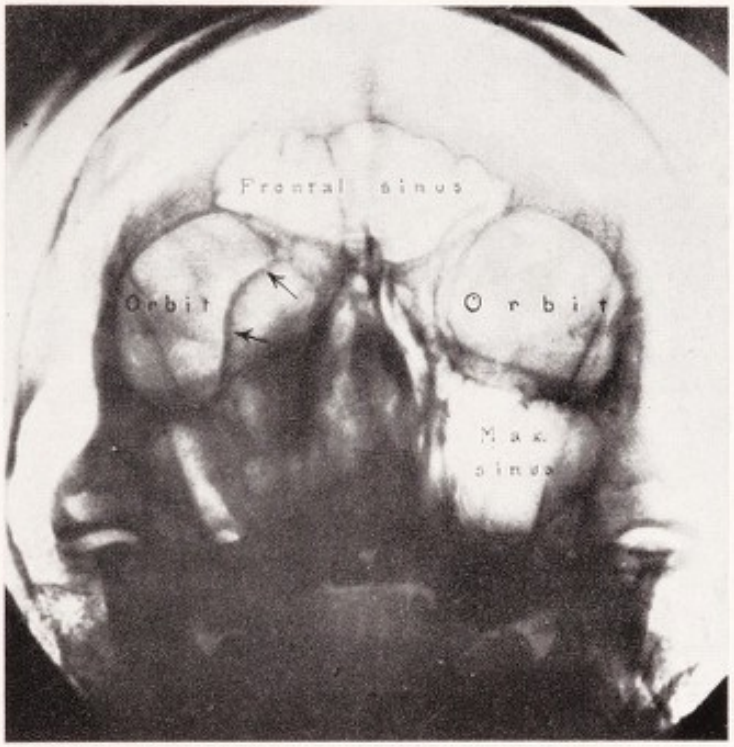


FIG. 24. Case 4. X-ray showing marked encroachment on inner side of orbit by osteomatous cyst pushed upward and outward.

X-rays showed (anteroposterior view) a sharp-curved convex bony line cutting across the inner side of the orbit (Fig. 24).

Wassermann reaction from the blood was negative.

Operation, July 28, 1939: A hypophyseal approach with concealed incision was made. The dura covering the posterior fourth of the orbit had lost its glossy appearance because of a capillary network, but there was no sign of

tumor. With a chisel the orbital roof was removed to the sphenoidal ridge. The orbital capsule, which did not look abnormal, was opened, but there was only a small amount of orbital fat anteriorly. The large superior rectus muscle bulged into the defect and was pulled aside with a traction suture. Immediately beneath it was a round, smooth,

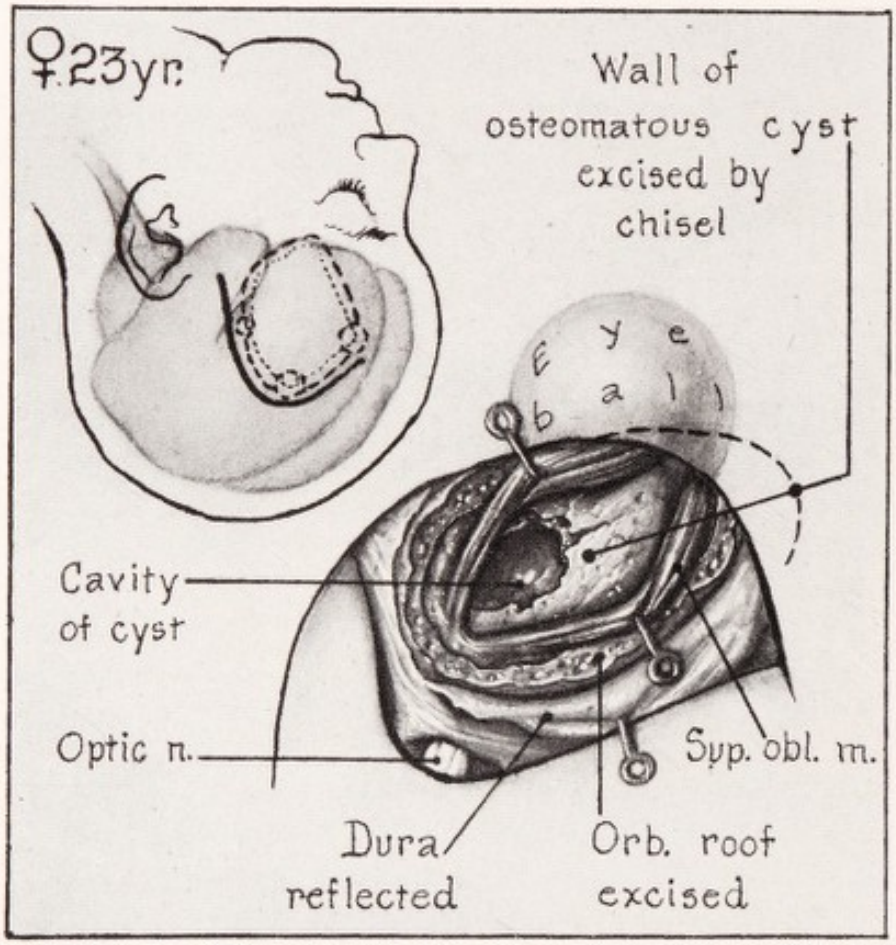


FIG. 25. Case 4. Operative sketch showing exposed wall of cyst which was chiseled away.

hard bony mass growing up from the floor of the orbit. With a small chisel it was opened and about an ounce of blood-tinged pink fluid escaped, filling the wound. No crystals were seen, and because the fluid was tinged with blood it was not examined. The forceps were passed through the opening into a big free cavity (about as large as

a bantam's egg) to a depth of 3 or 4 cm. I was fearful that this might be a big antrum and that cerebrospinal rhinorrhea might follow and that the removal of more of the bony skull might therefore end disastrously. There was no subsequent escape of cerebrospinal fluid. Reassured by this and the absence of any signs of infection, I reopened the wound two weeks later and chiseled away the entire bony skull (Fig. 25). It was fairly uniform in thickness, which was perhaps

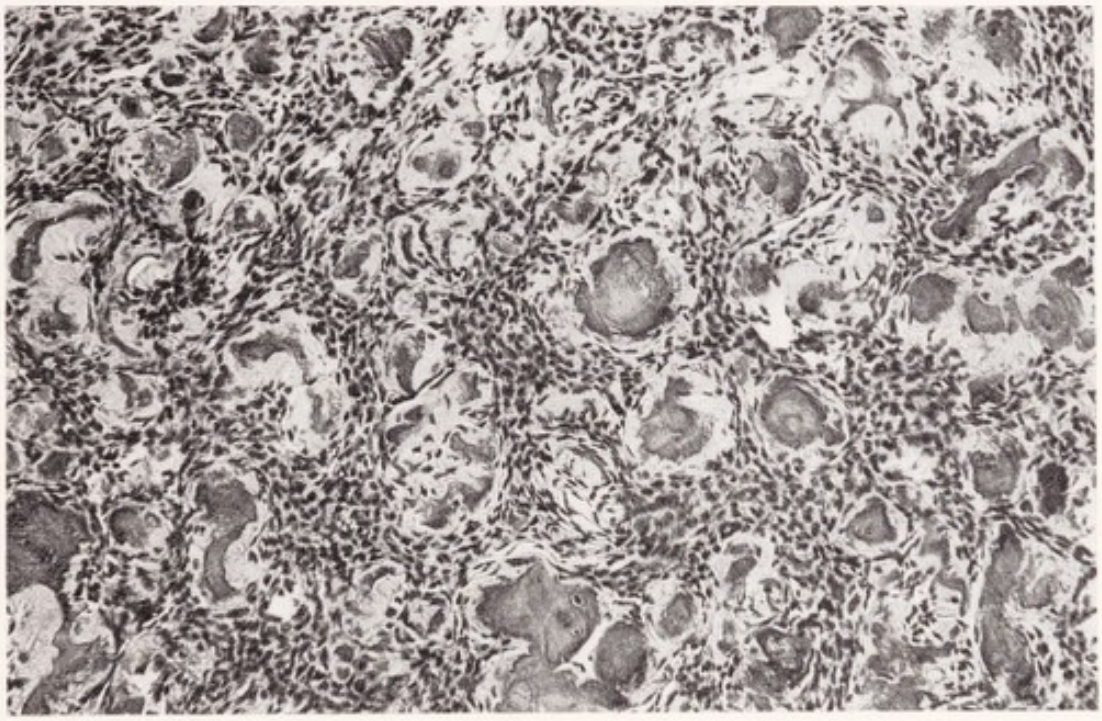


FIG. 26. Case 4. Photomicrograph showing osteoblasts in lining of cyst.

1 to 2 mm. The large cavity was now well exposed, but no sign of soft tumor could be found within. There was, however, a thin lining of soft tissue on the inferior border of the bony shell. Several bits of this tissue were removed for microscopic study and labeled "Is this tumor?" The weight of the bony roof of the cyst was 2.1 grams.

The wound healed per primam and the patient left the hospital thirteen days later—August 23. At the time of discharge there was no appreciable change in the exophthalmos

or vision. A letter dated November 18 (three months later) stated that the eye had receded almost to normal, that the vision in this eye was as good as in the other, and that there had been no double vision (Fig. 27).

Microscopic Note: "The shell of bone shows a normal histological structure. Bits of tissue removed at the second operation and labeled 'Is this tumor?' are made up of very

FIG. 27. Case 4. Three months after operation.

cellular fibrous tissue that is continuous with the marrow of the lining bone, which is normal. Throughout the fibrous tissue are very large but irregularly shaped hyalinized bone cells—osteoblasts [Fig. 26]. At first glance they look like psammoma bodies, but they have no concentric lamellation and do contain nuclei."

Diagnosis: Osteomatous cyst.

CASE 5

F. P., History number U-75001. White, male, age 61. Admitted: January 20, 1937. Discharged: February 7, 1937.

Diagnosis: Spindle-cell sarcoma.

Complaint: Protrusion of the right eye.

Family and Past Histories: Negative. The left eye had been removed several years before following an accident.

Present Illness: Thirteen months before admission protrusion of the right eye was first observed; it steadily increased. There was a dull pain in and behind the right eye. The vision had gradually decreased until on admission the patient could only count fingers. He had lost 20 lbs. in weight in the preceding four months. I had seen this patient four months before (August 4, 1936), when his vision had been much better. It was then clear that an operation should be performed, but I was reluctant to undertake it because the other eye had been removed. As long as his vision, which at that time was 20/100, was fairly good, it seemed better to wait until this was lost before suggesting an operative procedure with any possible hazard to vision. The patient thought his vision then had been failing for three or four months; nevertheless, it could be corrected to 20/20 with glasses. Dr. Woods made the following observations at that time: "The exophthalmometer registered 30 mm. as against a normal of 17. The eyeball was negative and the pupillary movements were normal. The eye was pushed forward and somewhat downward [Fig. 28]. The upward movements of the eyeball were markedly limited; lateral and downward movements quite well performed. The visual fields and blind spots were normal."

The patient returned on January 20, 1937. The vision had decreased to 5/200, but there was still a full visual field. The exophthalmometer read 33 mm., an increase of 3 mm. in five

months. No extraocular movements were possible above the horizontal plane. The eye could be rotated upward to 15° and downward to 20°. A mass could then be palpated outside and above the eyeball; it was tender on pressure. The fundus and disc were normal. There was a diffuse superficial keratitis, the entire cornea staining with fluorescein. The tension of the eye was 15 mm.(Schiotz).

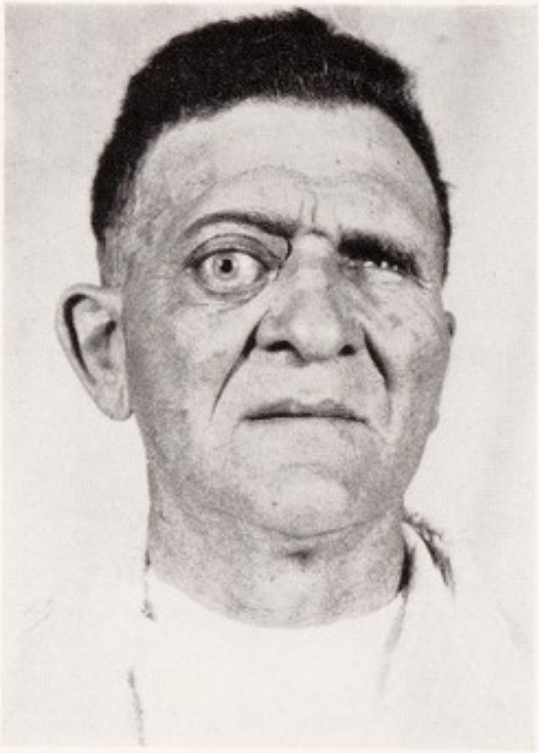


FIG. 28. Case 5. Preoperative photograph, showing marked grade of exophthalmos. Left eye had been removed following an accident years before; right eye was now almost sightless.

Operation, January 22, 1937: A right hypophyseal approach was made, using the concealed incision. The cisterna chiasmatis was opened, providing ample room for exposure of the orbital roof. The orbital dura was incised laterally and stripped from the bone. The roof of the orbit was so thin that it was transparent in one place; it was removed with rongeurs. The orbital contents, covered with the capsule, protruded through the defect. A longitudinal incision was

made on the orbital fascia and counter-incisions carried to both sides. The tumor bulged into the defect; it was semi-gelatinous and reddish-brown; it was fairly firm and well circumscribed. The tumor was so large and soft that it could not be shelled out intact. The interior of the growth was first curetted away, then the capsule was lifted with the forceps and completely stripped from its bed.

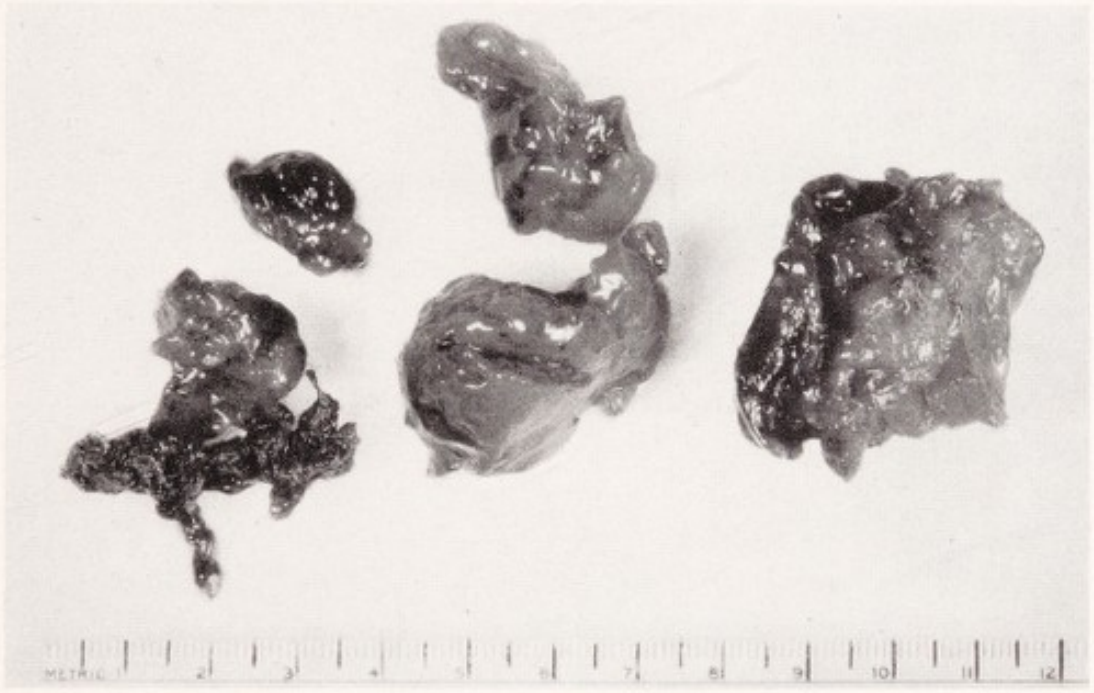


FIG. 29. Case 5. Fragments of tumor removed piecemeal.

After removal of the tumor one could see the optic nerve pushed mesially. There was practically no bleeding at any time during the dissection. The tumor was not attached to the back of the eyeball, which was now in full view. The orbital fascia was closed with few silk sutures.

The tumor weighed 20.7 grams (Fig. 29). From its gross appearance I made the diagnosis of sarcoma.

Postoperative Course: At the time of discharge from the hospital, February 7, the patient's vision had returned to 20/50 and the visual field was entirely normal for form and color.

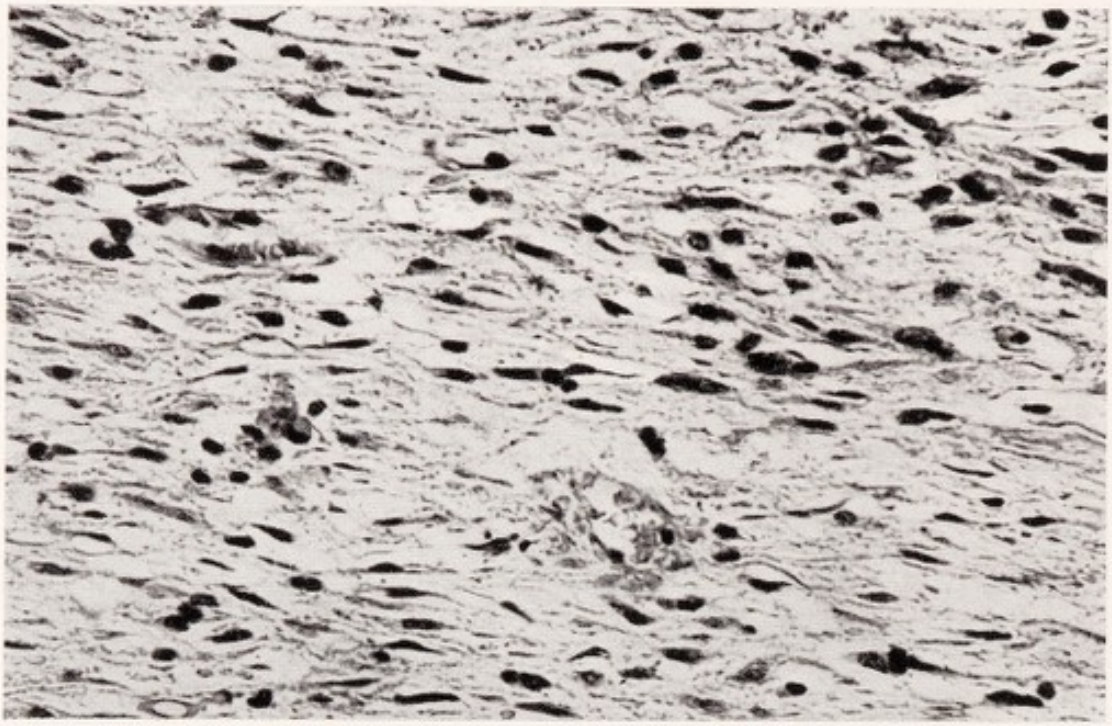


FIG. 30. Case 5. Photomicrograph of tumor: spindle-cell sarcoma.

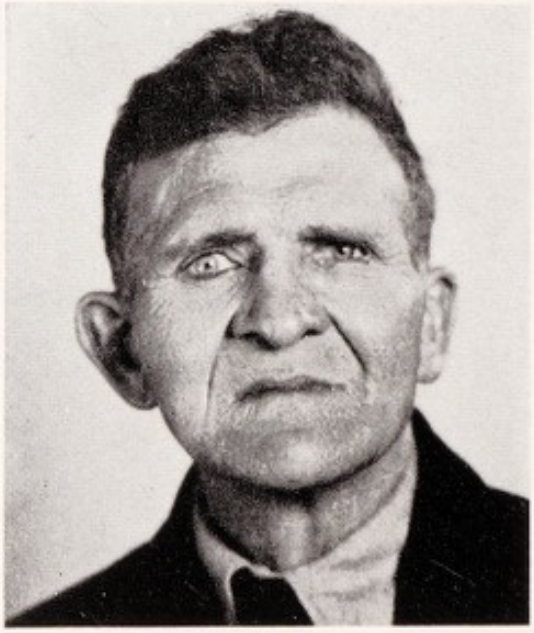


FIG. 31. Case 5. Two and one-half years after operation. Vision in remaining eye had returned to normal.

Microscopic Report: "In places the tumor has every appearance of a fibroma. In other places there is a myxomatous matrix with scattered cells, the nuclei of which are greatly elongated and of spindle shape, but many are bent at angles of varying degree. The tumor is probably a spindle-cell sarcoma, perhaps a rather cellular fibroma" (Fig. 30).

Subsequent Course: A letter of January 17, 1938, reported, "My eye seems to be getting along fine. It isn't any bigger as you can see by the snapshot. I can read the finest print without any difficulty. My head does not give me any pain whatever." Another received June 18, 1939, nearly two and one-half years after operation: "I can see as well as the day I left the hospital. I can read without the least strain" (Fig. 31).

CASE 6

A. B. S., History number U-72278. White, female, age 2½ years. Admitted: September 29, 1936. Discharged: October 9, 1936. Died: January 15, 1937.

Diagnosis: Round-cell sarcoma.

Complaint: Protrusion of the left eye.

Family and Past Histories: Negative.

Present Illness: Six weeks ago the mother had first observed a prominence of the left eye; this had steadily increased. The child had not complained of pain. The mother did not think that any vision remained in the affected eye. There had been no other symptoms.

Examination: The patient was a well-nourished, bright child and in no evident distress. The examinations were entirely negative, except for the exophthalmos in the left eye; this measured 11 mm. In the preceding month, under the observation of Dr. Woods, the orbital protrusion had increased 5 mm. There was no pulsation or bruit. The

extraocular movements were markedly limited, the upper movements being most affected. The left pupil was a little larger than the right. Ophthalmoscopic examination

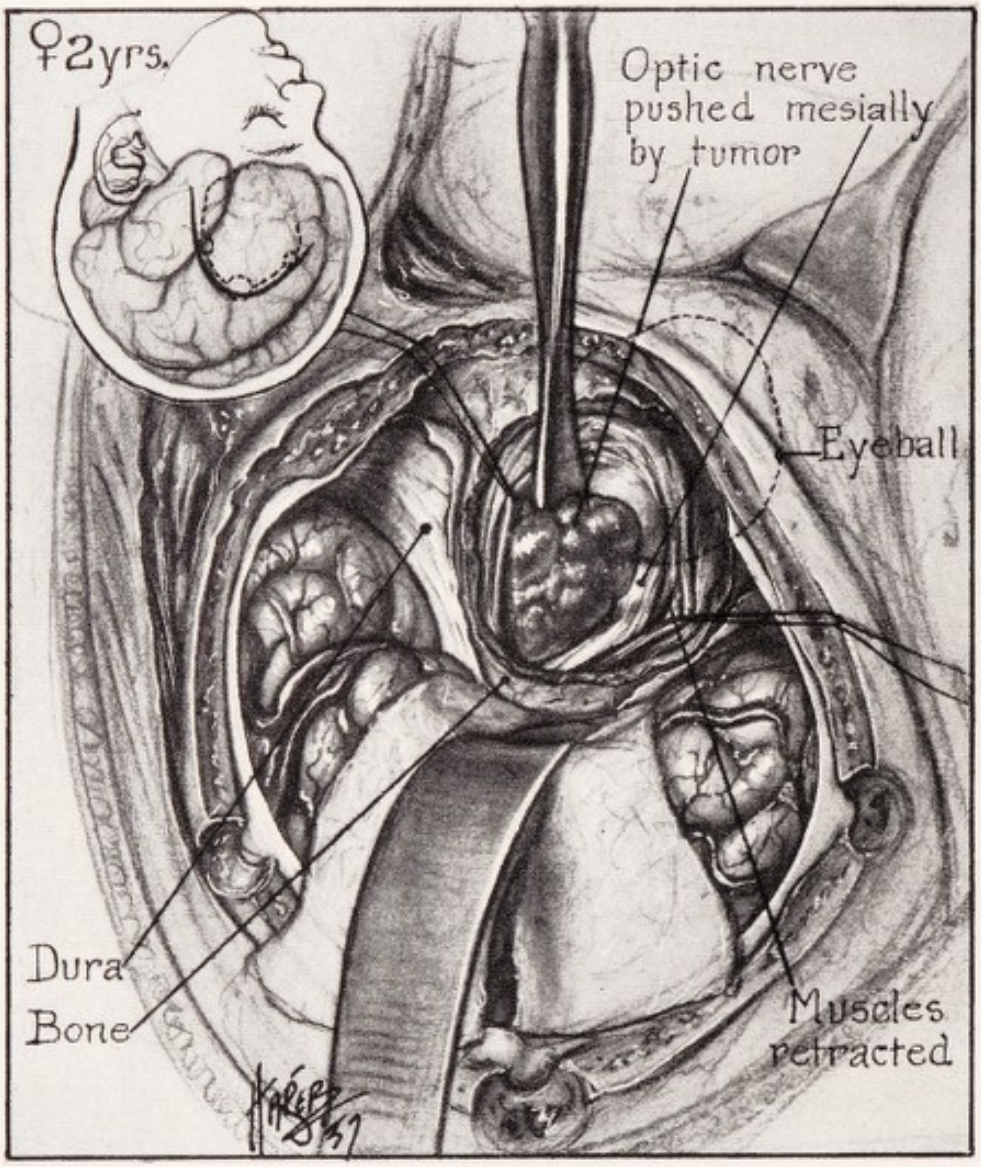


FIG. 32. Case 6. Operative sketch showing removal of tumor from superior and posterior parts of orbital chamber.

showed marked overfilling and considerable tortuosity of the retinal veins on the left.

X-rays of the orbital foramen were negative.

The age of onset and the rapidity of growth suggested a malignant tumor, probably a sarcoma.

Operation, September 30, 1936: Using the concealed incision, a left hypophyseal approach was made and the roof of the orbit removed. The orbital fat protruded markedly and was excised until the superior rectus muscle appeared; this was pulled aside by a ligature. The external rectus muscle was similarly identified and retracted. The tumor filled

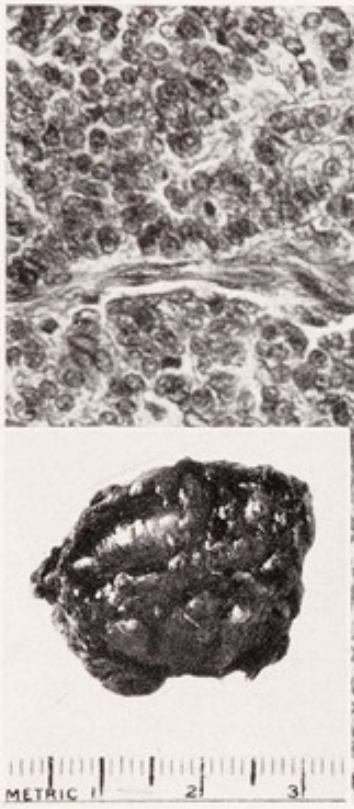


FIG. 33

FIG. 34

FIG. 33. Case 6. Tumor removed in toto, but subsequently recurring.
FIG. 34. Photomicrograph of tumor: round-cell sarcoma.

the back of the orbit and pushed the optic nerve mesially. It was very hard and well circumscribed; posteriorly, however, it became softer, and tore during the dissection. The anterior part of the tumor was dissected from its bed with instruments (Fig. 32) and finally the terminal posterior portion, which was firmly attached, was dislodged with the index finger; it was removed in one piece. Its weight was 7.5 grams (Fig. 33).

The postoperative course was uneventful; the patient left the hospital on October 9, ten days after the operation. The exophthalmos had disappeared; there were no extraocular palsies.

Microscopic Report: "The tumor is a round-cell sarcoma [Fig. 34]. The cells are larger than the typical small round-cell tumor. The nuclei are vesicular. There is alveolar formation of the cells in places; elsewhere the cells are closely packed, without arrangement. There is very little fibrous tissue in the tumor."

Subsequent Course: Steadily increasing protrusion of the eye, one month after discharge, and loss of weight and gradual physical decline were soon in evidence. Repeated deep X-ray treatments were without benefit. The child died on January 15, 1937, three and one-half months after the operation.

CHAPTER II

CASES OF SCHÜLLER-CHRISTIAN'S DISEASE

CASE 7

D. M. R., History number U-175119. White, male, age 46. Admitted: June 26, 1939. Discharged: July 14, 1939.

Diagnosis: Schüller-Christian's disease. (Referred by Dr. Robert S. Leach, Knoxville, Tennessee.)

Complaint: Orbital tumor.

Family and Past Histories: Negative.

Present Illness: Two and one-half months ago the patient had noticed a slight swelling of his right upper eyelid. The swelling was present only in the mornings and disappeared after he had been up for a while. The swelling extended over to the right side of the nose. It had been less pronounced in the mornings during the past two weeks and had been absent entirely the past two or three days. There had been no pain at any time, no nasal discharge, and no conjunctivitis.

Dr. Leach had made the diagnosis of an orbital tumor. He found a slight grade of exophthalmos and by X-rays a defect in the lateral wall of the skull; immediately beneath this was an opening in the roof of the orbit. There was no history of trauma.

Examination: There was a low grade of exophthalmos measuring 2 mm. It was barely detectable on observation (Fig. 35). There was a very slight swelling just above the supraorbital ridge and on the side of the skull. One could feel the opening in the skull at this point, but there was no pulsation. The region, however, was not tender. Dr.

MacLean was quite certain that the tumor could be palpated over the eyeball. There was no extraocular palsy and the patient had never had diplopia. The fundus was normal. The visual fields and visual acuity were normal in both eyes. The Wassermann reaction from the blood was negative.

FIG. 35. Case 7. Preoperative photograph. Low grade of exophthalmos on right side.

X-rays showed a small circular defect in the side of the skull (Fig. 36) just back of the orbital margin, and X-rays of the orbital roof showed the defect extending into it also (Fig. 37). The defect in the side of the skull was about as large as a five-cent piece, sharply defined and quite circular; that in the orbital roof was somewhat smaller.

Diagnosis: Orbital tumor with destruction of bone.

Operation, June 30, 1939: A hypophyseal approach was made. When the skin and galea were turned back no sign of tumor was seen, but when the bone flap was broken one could see the tumor coming through the bone at the lower

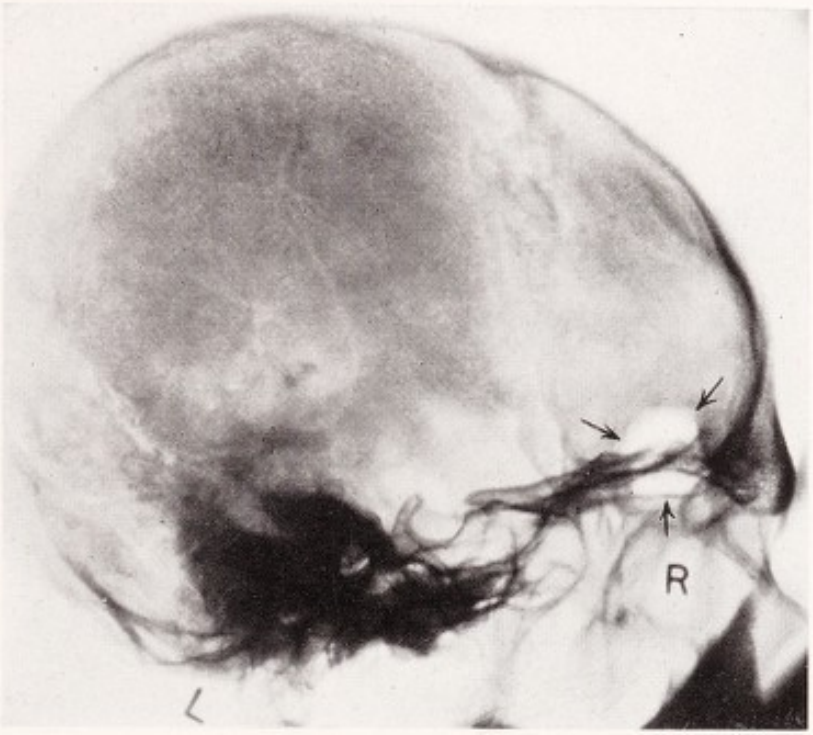


FIG. 36. Case 7. X-ray showing small circular area of destruction in lateral wall of skull and roof of orbit. Erosion indicated by arrows.

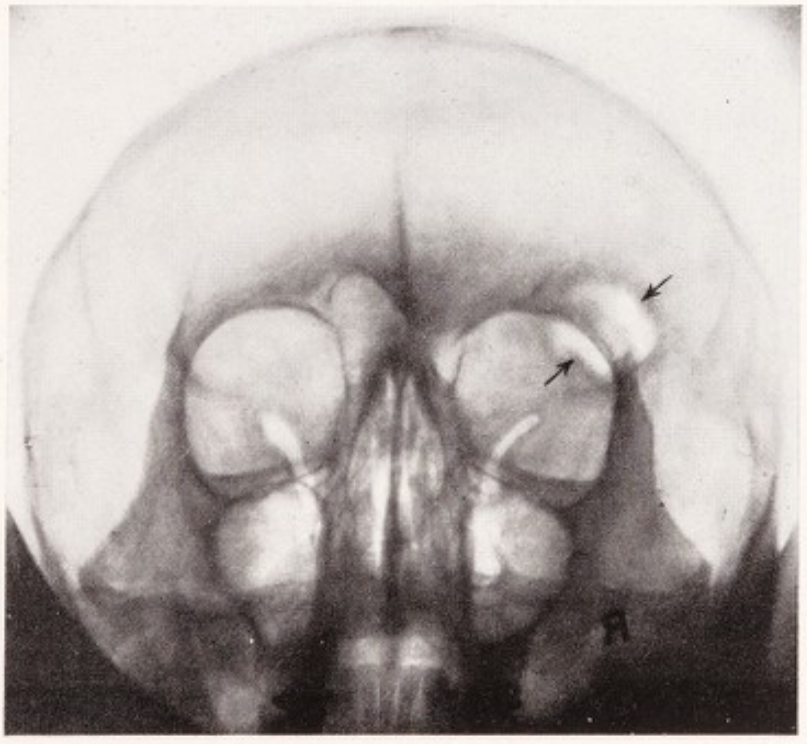


FIG. 37. Case 7. Anteroposterior X-ray showing area of destruction in lateral wall of skull and roof of orbit.

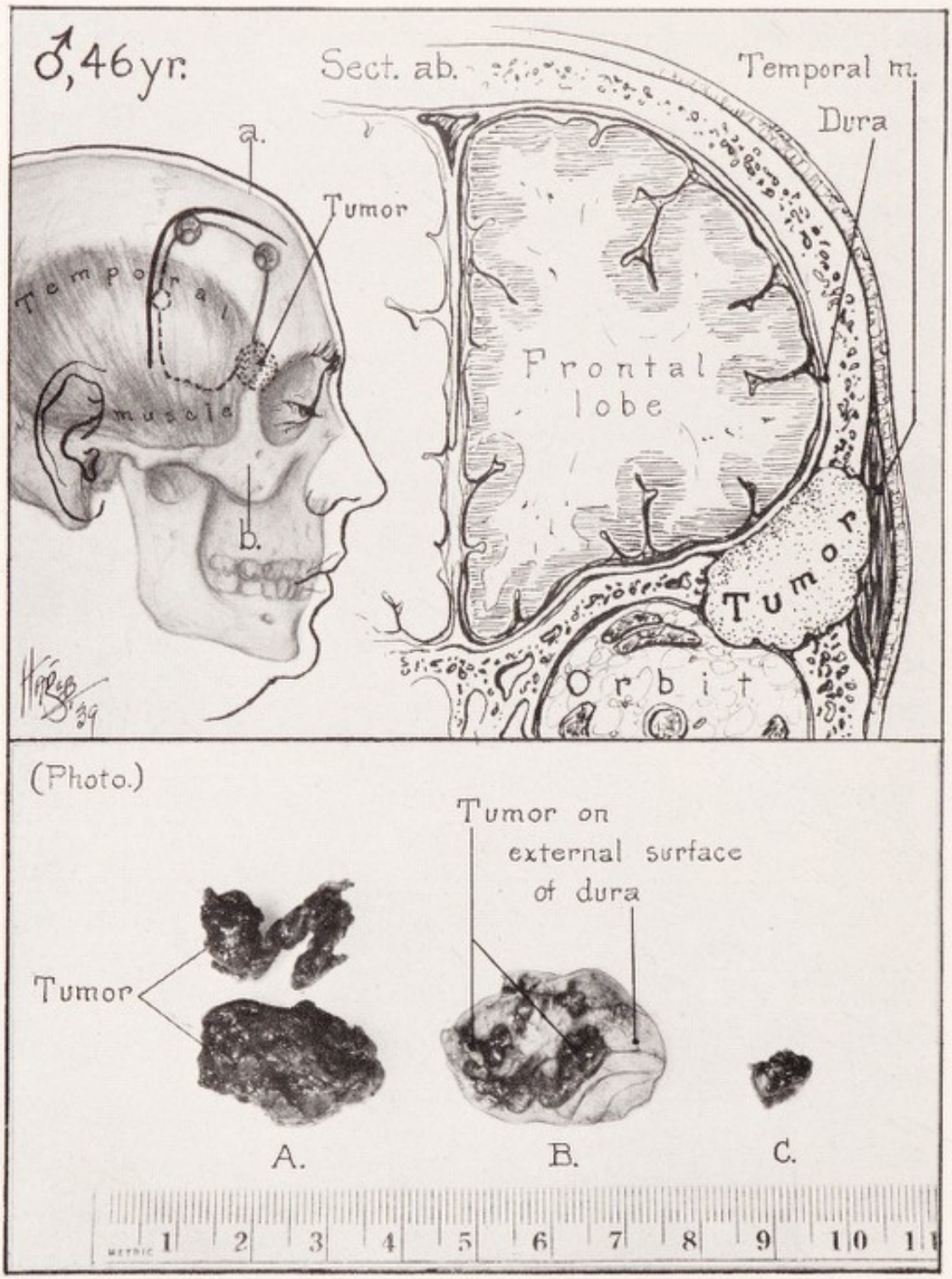


FIG. 38. Case 7. Operative sketch showing site of tumor, and relation to orbital roof and lateral wall of skull. Below: (A) tumor removed; (B) tumor attached to excised dura; (C) portion removed from roof.

anterior angle; it was just under the anterior border of the temporal muscle at its origin. It was a grayish-white mass and very grossly cellular. The defect in the lateral wall of the skull was about as large as a five-cent piece. The overlying muscle was incised around the tumor, after which the growth was shelled from its bed. However, it was not soft and not encapsulated and was, therefore, removed in fragments. The tumor could now be followed through the orbital roof,

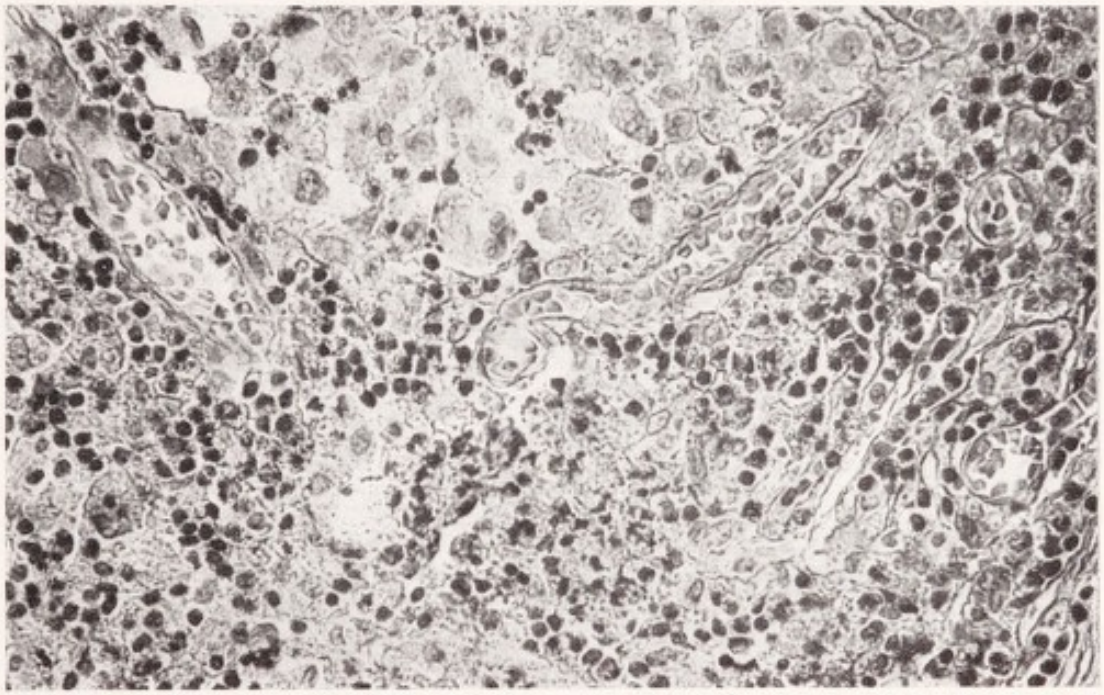


FIG. 39. Case 7. Photomicrograph of tumor: Schüller-Christian's disease.

the defect in which was about as large as a ten-cent piece (Fig. 38); the anterior margin of this defect was about 2 cm. posterior to the supraorbital ridge. This portion of the tumor was removed separately. It was quite adherent to the orbital capsule. An additional amount of bone was removed from the entire periphery of the defect, after which the portion of the tumor adherent to the orbital capsule was completely excised with a good margin of healthy

capsule. The intraorbital contents were not invaded. The defect in the lateral wall of the skull was also enlarged, in order to remove any possible remains of tumor in the bone. The remaining tumor, which was adherent to the dura, covered a circular area as large as a silver dollar and this was completely excised. Over the dural defect a transplant of fascia (removed from the temporal muscle) was snugly sutured to the dural margins.

Microscopic Pathological Report by Dr. Arnold R. Rich: "Section 1 shows spicules of bone, between which are masses of large mononuclear cells, apparently macrophages, and many eosinophils [Fig. 39]. The appearance is altogether like that found in the marrow in Schüller-Christian's disease. There are many foam cells.

"Section 2 shows dense hyalinized connective tissue with foci of small mononuclear cells, particularly about the blood vessels. This section is quite like most of the sections in Case 8."

Diagnosis: Schüller-Christian's disease.

Subsequent Course: The patient's postoperative course was uneventful. He left the hospital on July 14. On February 9, 1940 (seven months after operation), the patient wrote that he was "perfectly well and had never had a pain or ache, or, in fact, the least discomfort since the operation."

Comments: The question arose in the beginning whether a tumor located so far anteriorly could have been removed by a local approach, rather than by turning down a bone flap. However, the involvement of the dura and of the orbital capsule was much too extensive and too far back to have permitted its careful removal without a bone flap.

CASE 8

H. E., History number U-100252. White, male, age 43. Admitted: March 7, 1937. Discharged: April 19, 1937.

Diagnosis: Schüller-Christian's disease (probable).

Complaints: Pain in the left side of the face, loss of vision in the left eye, and protrusion of the left eye.

Family and Past Histories: Negative.

Present Illness: Four years ago the patient had begun to suffer from pains in the left upper lid and in the left eye. One year later he was operated upon in another hospital where the presumptive diagnosis of brain tumor had been made, but it was said that nothing was found. The pain in the face became steadily more severe and after several months facial numbness developed. Two years later he was operated upon again in another hospital and a tumor of the sphenoidal ridge was said to have been found and a small piece removed. Following this operation there was complete loss of vision in the left eye and complete ptosis of the left lid (Fig. 40). The incision had been carried along the orbital ridge and extended far backward, making a very large bone flap, only part of which had been replaced.

A dull, throbbing pain continued behind the left eye; in addition there were many attacks of lancinating pain. The pain occurred throughout the face, but more in the upper two branches of the trigeminal domain. There had been a great deal of nausea, vomiting, and headache.

Physical and Neurological Examinations: The patient was a sallow, ill-appearing man; he looked much older than forty-three. The positive findings were: (1) exophthalmos of 6 mm. in the left eye, (2) complete paralysis of the oculomotor nerve, (3) partial paralysis of the left abducens, slight remaining rotary function of the eyeball; (4) left pupil greatly dilated with no reaction to light, (5) a white glistening disk of primary atrophy in the left eye ground, (6) hypoesthesia of the first trigeminal branch and hyperesthesia over the second and third branches.

No tumor could be palpated above the eyeball. Vision

in the right eye was normal. X-rays showed a large operative defect in the parietal region. This was due to the partial removal of the bone flap in an earlier operation. The defect did not bulge. There were numerous clips in the middle fossa, where doubtless the tumor was encountered. The blood pressure was 130/85, but was said to have been elevated to 160 and even 180 at times in the past. Wassermann

FIG. 40. Case 8. Preoperative photograph. Tumor in middle fossa, incorporating Gasserian ganglion and carotid artery, and extending into orbit. Diagnosed as Schüller-Christian's disease.

reactions from the blood and spinal fluid were negative. There were no cells in the spinal fluid, and 150 mm. of pressure.

Operation, March 13, 1937: Only the posterior part of the old incision was reopened. This permitted exposure where the bone was absent over the temporal lobe. An additional small area of bone was removed anteriorly in order to expose the roof of the orbit. There was no tumor present over the orbital roof. The undersurface of the temporal lobe was

then exposed. The brain was quite tight, but when the dura, which was quite adherent, was stripped, a burst of fluid followed and provided ample room to expose and attack the tumor. Much of the temporal lobe had been destroyed at

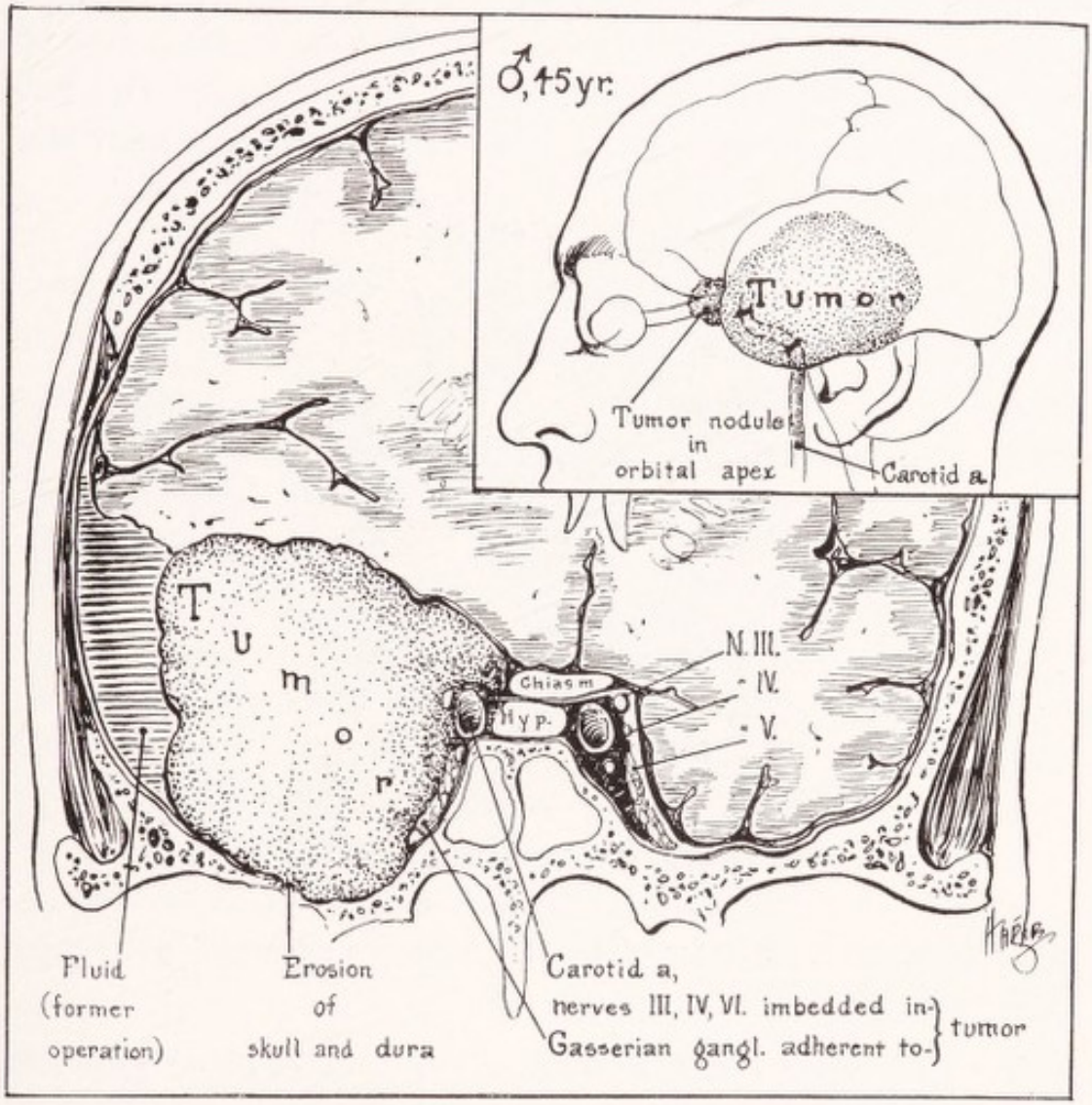


FIG. 41. Case 8. Sketch showing location and relative size of tumor. Inset shows the two points of ligation of internal carotid artery when tumor was removed.

the earlier operation and the space was filled with fluid. The growth filled much of the floor of the middle fossa and extended to the mesial border of the dura (Fig. 41). It was very hard and fibrous (Fig. 42) and was densely adherent to

the Gasserian ganglion and by its branches to the floor of the skull. With the periosteal elevator the dura was stripped from the middle fossa down to the tumor. It was evident that the tumor had grown into the Gasserian ganglion and there was no possibility of separating it from this. The dura itself was absent over the growth. It was hoped that the carotid artery might be avoided, but since most of the bone along the inner aspect of the middle fossa had been destroyed,

FIG. 42. Case 8. Cluster of tumor fragments of Schüller-Christian's disease removed piecemeal.

leaving this very largely exposed, it was evident that this hope could not be realized. The carotid was then sought anteriorly just before it entered the cranial chamber; for some distance the tumor was stripped from it. It was found increasingly more firmly imbedded in the growth and eventually was torn; its wall was greatly weakened by the tumor. A spurt of blood was immediately suppressed with a pack and the artery was occluded by cautery. The remainder

of the tumor was then dissected posteriorly toward the base of the skull, and as the tumor was being delivered the carotid artery was sought for a second occlusion. It was again torn where it entered the base of the skull, but the bleeding was easily controlled by cautery. The internal carotid artery was therefore cauterized as it entered the base of the skull and also where it entered the cranial chamber. At no time was there any bleeding from the cavernous sinus which had been entirely obliterated by the tumor. The third, fourth, and sixth nerves could be seen entering the mesial border of the dura, which was entirely surrounded by tumor. These nerves, which were without function, were divided. The tumor passed through the sphenoidal fissure into the orbit. A part of the roof of the orbit was removed and a small nodule of tumor was removed from the orbital apex.

Three days after the operation the patient had a convulsion involving the right side of the body; there was complete rigidity without loss of motor function.

On the eighth day there was another similar attack. There were no signs of intracranial pressure, however, and the temporal defect was not bulging. Although there were no signs of an intracranial hemorrhage, the fact that the internal carotid artery had been closed with the cautery made us apprehensive of this possibility. The internal carotid artery was ligated in the neck. The patient's subsequent postoperative course was uneventful.

Microscopic Pathological Report by Dr. Arnold Rich: "The sections consist for the most part of thick strands of hyalinized connective tissue. In places, numerous plasma cells and lymphocytes infiltrate between the hyalinized fibers. There are foci of densely packed lymphocytes, often perivascular, and some of these have germinal centers. Occasional multinuclear giant cells are found in association

with these lymphoid foci. In addition to these changes, there are found several areas in which there are necrotic masses of collagen surrounded, or partly surrounded, by elongated connective tissue cells arranged in palisade fashion (slide 65773-3). In the same section there are several little abscess-like accumulations of eosinophils, surrounded, or partly surrounded, by radially arranged connective tissue cells. Multinuclear giant cells are occasionally found in these foci. In one area there is an accumulation of typical foam cells, closely packed and occupying a third of a low power field. Smaller foci of foam cells are also found."

Examination by Dr. Alan Woods of tissue removed at the second operation showed: "In places the lymphocytes are present in round nodules suggesting lymph follicles. There are, however, no germinal centers. There are often multinuclear giant cells at the periphery. The section of the eye shows the lymphocytic and plasma cells infiltrating above the optic nerve. These extend into the sclera and in one place there is a mass of lymphoid cells in the choroid.

"I cannot make a certain diagnosis of this case. That it is an inflammatory lesion and not a tumor is clear. The foam cells and eosinophils suggest Schüller-Christian's disease, but the predominant change is the fibrosis and inflammatory infiltration. I have not seen the large areas of necrotic collagen surrounded by palisades of connective tissue cells in Schüller-Christian's disease. A precisely similar lesion does, however, occur in rheumatoid arthritis nodules and in subcutaneous nodules in syphilis. Such nodules are typical of granuloma annulare."

Subsequent Course: The patient slowly improved physically and was seen again in August, 1938, eighteen months after the operation. At this time a mass could be palpated beneath the supraorbital ridge; proptosis measured 6 mm. His

general health was much improved. The pain had not reappeared since the operation. Removal of the mass in the orbit was not then advised.

In March, 1939 (two years after the operation) he again returned and the orbital mass had become so much larger and so adherent to the conjunctiva that Dr. Woods operated for removal of the mass through the anterior orbital approach over the eyeball. The tumor filled the orbital cavity and extended into the ethmoidal cells and the antrum. It was subtotally removed. The patient returned home none the worse for the extensive removal of the growth in the sinuses and orbit.

CASE 9

W. B., History number U-106258. White, male, age 58. Admitted: May 25, 1937. Discharged: June 13, 1937. Died: January 19, 1938.

Diagnosis: Schüller-Christian's disease (probable).

Complaint: Protrusion of both eyes.

Family and Past Histories: Negative.

Present Illness: The patient was first seen by Dr. Woods, in the Department of Ophthalmology, May 5, 1937. Three months before that time the patient had noticed lachrimation of both eyes and consulted an oculist, who had dilated his tear ducts. Following the third dilatation there was some edema of the right lower lid and conjunctivitis; both spread rapidly to the left eye. Soon thereafter exophthalmos of both eyes was observed. At this time the patient was seen by Dr. C. C. Hansen-Pruss, of Duke University, who suspected multiple intracranial or intraorbital tumors, or perhaps abscesses. The exophthalmos had since continued to increase quite rapidly and at essentially the same pace in each eye.

Examination: The exophthalmometer then showed 26 mm. in the right eye and 28 mm. in the left (Fig. 43). A small nodule was felt under the upper lid of the right eye. The extraocular movements were normally performed. Vision was 20/30 in the right eye and 20/20 in the left. There was blurring of the nasal margins of both discs and elevation of 1 diopter on each side. A small piece of the nodule under the eyelid was removed by Dr. Woods. It was clearly

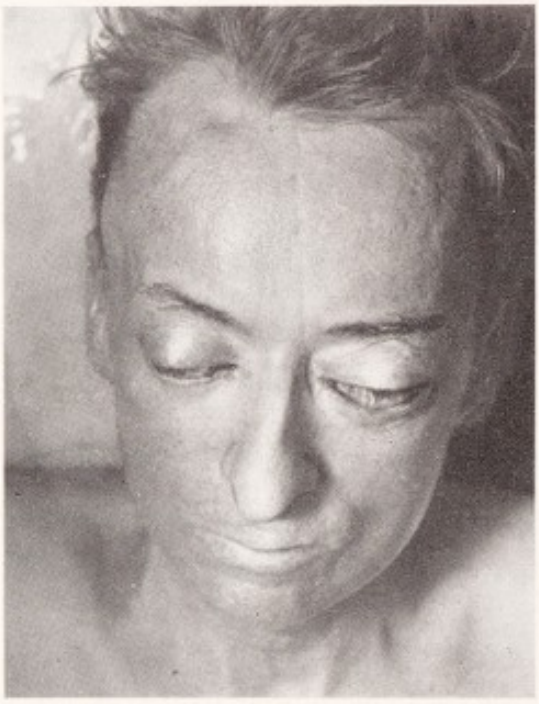


FIG. 43. Case 9. Marked bilateral exophthalmos resulting from Schüller-Christian's disease. There were masses throughout the body.

a tumor, but there was not sufficient tissue to make a positive diagnosis of its character. It was, of course, realized that it was a bilateral growth, but it was not certain whether it had spread laterally into both orbits from the paranasal sinus or cribriform plate, or whether it had arisen independently in each orbit. The only indication for an operation was to protect the patient's vision for a time: unless the growth was an extension from a central mass, there could be no possibility of a cure, or even prolongation of life.

Operation, May 26, 1937: A right hypophyseal approach was made, using the concealed incision. When the dura over the frontal lobes was thrown back, a thin plaque of deep-yellow tissue, scarcely as thick as tissue paper, was found to be attached to its undersurface, covering a roughly circular area perhaps an inch in diameter. It was not attached to the brain but solely to the dura, and was another of the multiple lesions which have subsequently been diagnosed as xanthomata. From the gross picture alone the thin dural lesion, although harmless-looking, almost certainly indicated a metastatic xanthoma and was probably ample reason for withholding further operative efforts. However, the operation was continued and the cisterna chiasmatis opened. The roof of the orbit was very thin; it was only necessary to touch the bone with the periosteal elevator to make the initial opening, after which the roof was easily removed with rongeurs. The tumor then presented; it was a salmon-colored, hard growth, filling the whole upper part of the orbit. For a time it looked as though it might be enucleated, but after its separation from the eyeball it was found for some distance posteriorly to be very densely attached to the apex of the orbit, and over a very wide base. There was no possibility of removing this mass intact.

The tumor weighed 4.2 grams.

The patient made an uneventful recovery and was discharged eighteen days later—June 13, 1937.

Microscopic Pathological Report by Dr. Arnold R. Rich: "One section of tissue from the orbit is composed almost entirely of foam cells. These are large cells with a small nucleus and many vacuoles in the cytoplasm. The vacuoles undoubtedly represent the sites of dissolved-out fat. Some of the cells are filled with minute vacuoles; in others, the vacuoles are larger, and in numerous ones, a pink-staining cytoplasm is present in which there are only a few vacuoles.

In places, the cells contain practically no vacuoles at all. Mixed with these cells there are lymphocytes and plasma cells and also numbers of eosinophils.

"A second section of tissue from the orbit shows a dense mass of connective tissue containing, here and there, islands of foam cells with eosinophils and mononuclear cells scattered among them. In places the foam cells are associated with what appears to be ordinary adipose tissue [Fig. 44].

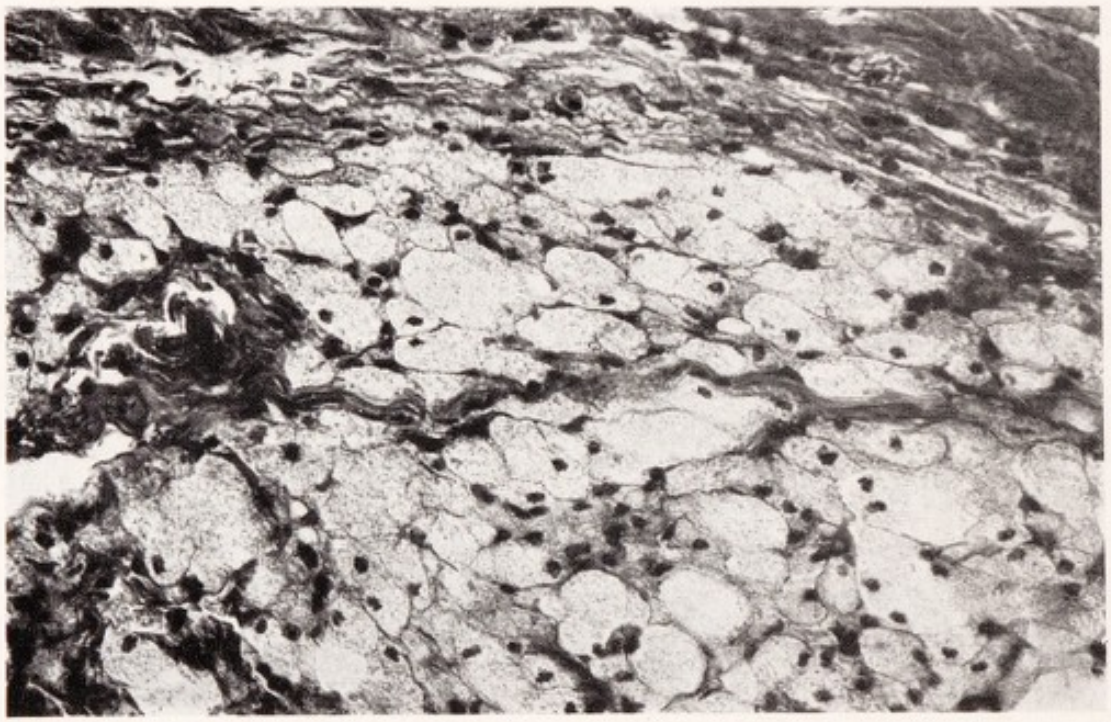


FIG. 44. Case 9. Photomicrograph of tumor, showing foam cells of Schüller-Christian's disease.

"The third section, clearly a piece of dura, shows a mass of foam cells on both sides. In one place there is a tiny accumulation of connective tissue-like cells embedded in the foam cells and associated with this is a nodule made up of cells with pink cytoplasm and oval nuclei. These appear to be of the same nature as the foam cells but contain no fat.

"*Impression:* The presence of the foam cells and eosinophils in this situation suggests Schüller-Christian's disease. How-

ever, in one of the sections, there is more dense connective tissue than is ordinarily found in Schüller-Christian's disease. Were it not for the eosinophils, one might regard it as a xanthofibroma, but taking the whole picture together, Schüller-Christian's disease has to be seriously considered as a possibility."

Subsequent Course: The patient was given deep X-ray treatments, and for a time there seemed to be some improvement, but the exophthalmos progressed. On November 9, his general condition was steadily declining, the orbital protrusion slowly increasing. At this time there was involvement of the left side of the chest, doubtless a metastatic involvement of the same tumor.

The patient died on January 19, 1938. Necropsy was performed by Dr. Roger Baker, of Duke University. The following is from his report concerning the lesion:

Anatomical Diagnosis: "Xanthomatosis involving especially the mesentery, the perinephric, retroperitoneal, mediastinal, and orbital connective tissues, the pericardium, endocardium, pleura, dura, and vertebrae."

CASE 10

L. I., History number 177903. White, female, age 42. Admitted: August 25, 1939. Discharged: September 26, 1939.

Diagnosis: Schüller-Christian's disease (probable).

Complaint: Blindness of right eye with bulging and pain.

Family and Past Histories: For the past seven or eight years the patient had had hypertension ranging up to 240.

Present Illness: The patient dated the onset of her present trouble to the sudden development of a hard painful swelling beneath the right zygoma ten months before. There were excruciating pains over the entire right side of the face and

both sides of the head, over a period of a month. These were so severe that frequent hypodermics were necessary. In two months the pains and the mass had disappeared. Seven months ago an "egg-sized mass" had developed suddenly in front of the right ear. A dull pain persisted over the mass for about two months, at the end of which time the mass had gradually disappeared. Two weeks ago during a sneezing spell an itching sensation had appeared in the right eye, and a dull ache in the right orbit. The latter pain was persisting and was worse at night. With the onset of the pain she was conscious of diminished vision in the right eye, the other being unaffected. There was also progressive ptosis of the right upper lid and protrusion of the eye at the onset of pain. The exophthalmos had progressed for two or three days, but since then had remained stationary. There had been no discoloration of the eyelids or eyeball.

Examination: The patient was quite obese with a blood pressure of 180/130 and 190/120, i.e., lower than her reputed higher pressure of 240. The right eye was proptosed 5 mm.; the upper lid drooped markedly. The external rectus had no function and all the other movements of the eyeball were impaired. The right eye was totally blind, the right pupil dilated, did not react to light, but reacted consensually. Vision in the left eye was normal. The eye grounds were negative except for a high degree of arteriosclerosis. The eyeball did not pulsate.

The Wassermann reaction from the blood was negative. X-rays of the orbit and orbital foramina were negative.

Impression: The sudden onset of the exophthalmos, blindness, and extraocular palsies, coupled with the hypertension, made us suspect an intracranial aneurysm. The history of the two recurring masses in the face, both of which had entirely disappeared without a trace remaining, suggested a

possible similar evanescent mass in the orbit. But how could such masses—if the patient's story was correct—be explained? She was advised to see what time would do. But sixteen days later she returned because the pain had become unendurable. An examination at that time showed no appreciable change.

Operation, August 29, 1939: A right hypophyseal approach with concealed incision was made. After evacuating the cisterna chiasmatis there was ample room to retract the frontal lobe and chisel away the orbital roof. The ethmoidal cells were not entered. There was no intracranial tumor. The orbital capsule was normal and abundant orbital fat protruded when the capsule was opened; much of this was excised. A small, hard, glistening-grey mass was located at the very apex of the orbit. It was densely fixed in place and was located just lateral to and beneath the optic nerve, which was well exposed and which it was necessary to retract in order to gain access to the growth (Fig. 45). The ophthalmic artery was seen alongside the nerve and retracted with it. With a sharp periosteal elevator the mass was shelled from its bed, considerable force being necessary; no bleeding followed. The tumor weighed 0.4 gram.

The postoperative course was uneventful. The patient left the hospital one month later. The pain in the eye had not returned, nor had the vision or ptosis changed; exophthalmos was less. A spinal Wassermann during the hospital stay was negative. A letter received November 1, 1939, stated that the patient had had no further pain, but no other comments were made.

Microscopic Pathological Report by Dr. Arnold R. Rich: "These sections are poorly preserved. They show infiltration of small mononuclear cells (apparently small lymphocytes) between the fibers of the orbital muscles [Fig. 46]. The

cells are uniform in size and appearance. Muscle fibers are atrophic where larger accumulations of these cells separate them. There is none of the dense connective tissue seen in the other cases. A section of lachrymal gland shows

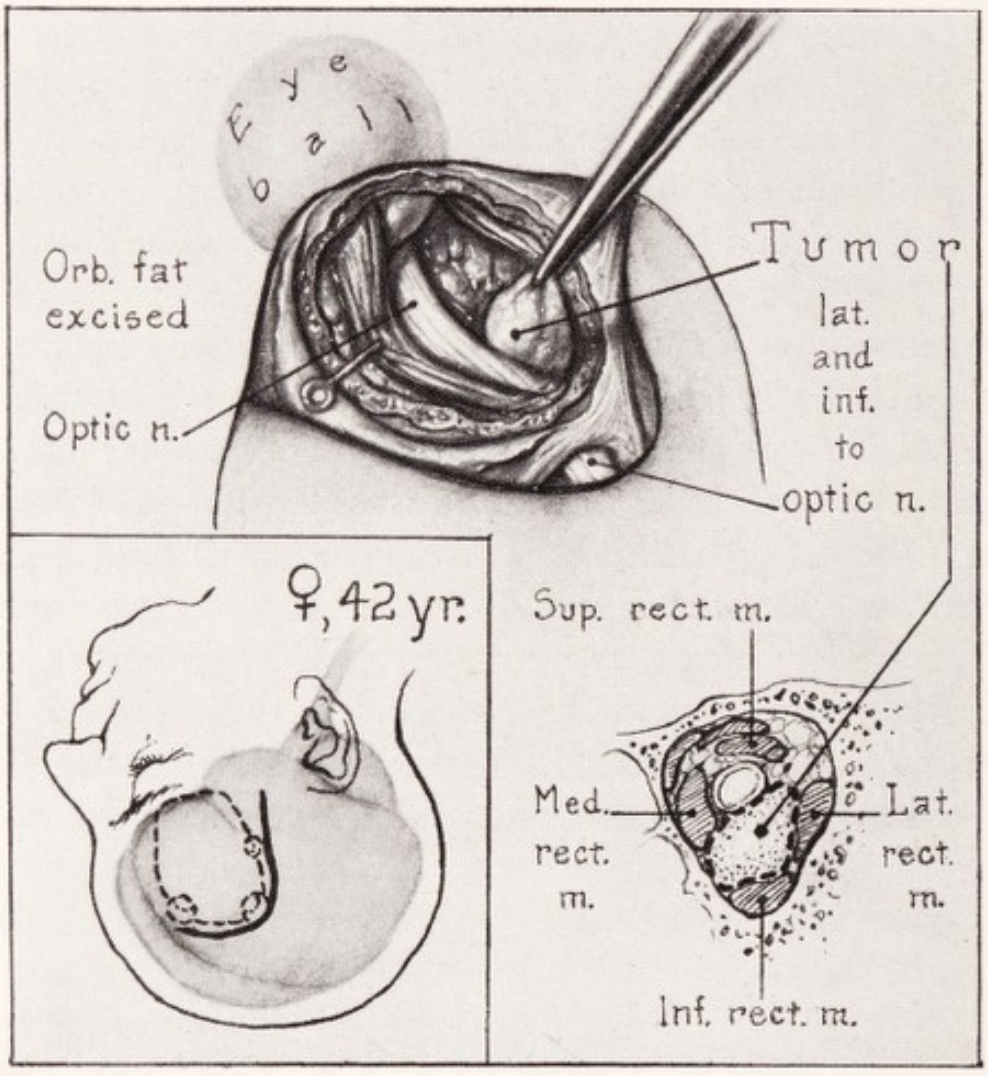


FIG. 45. Case 10. Operative exposure, showing tumor at apex of orbit. It was necessary to retract optic nerve to reach it.

several small foci of lymphoid cells. A section of bone (orbital roof) shows normal marrow in which there are several lymphoid foci."

Diagnosis: "Infiltration of orbital muscles with lymphoid cells. A leukemic infiltration would have this appearance,

but the marrow shows only slight lymphoid foci. Infiltration of the orbital muscles with lymphocytes occurs in hyperthyroidism. What the cause of the infiltration in the present case may be, I cannot say; perhaps Schüller-Christian's disease."

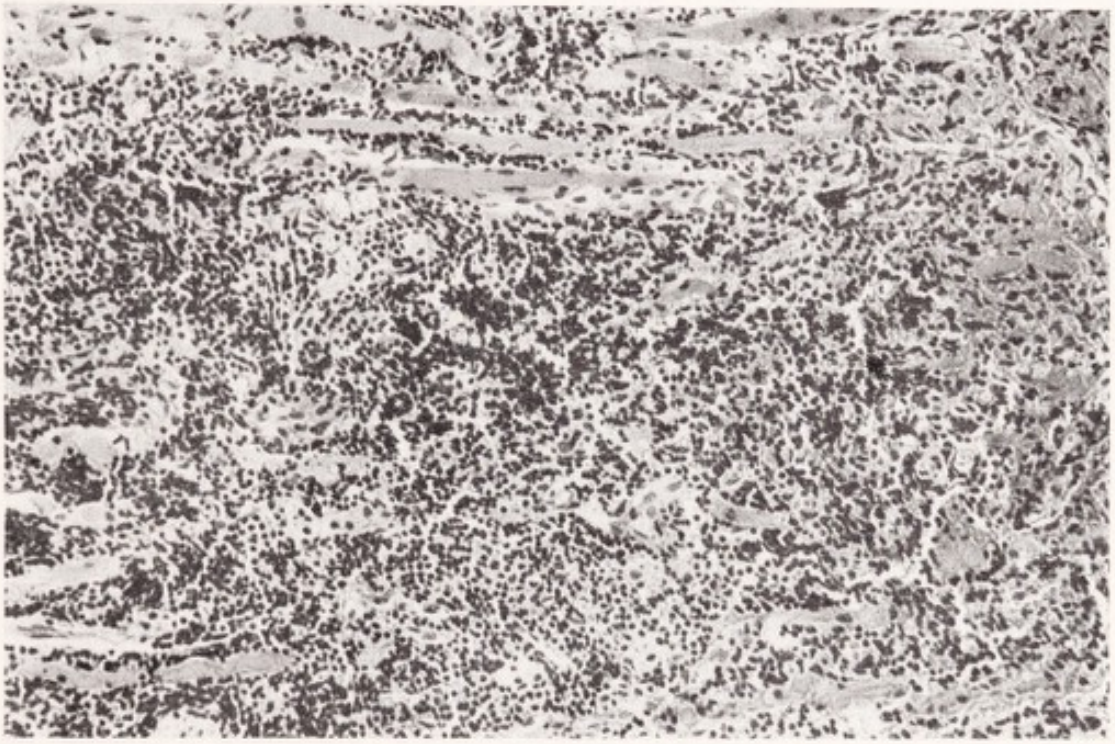


FIG. 46. Case 10. Photomicrograph showing orbital muscles in a mass of lymphocytes.

CASE 11

A. M., History number 180078. White, male, age 37. Admitted: August 24, 1939. Discharged: October 8, 1939.

Diagnosis: Schüller-Christian's disease.

Complaint: Painful bulging of the right eye.

Family and Past Histories: Negative, except for a sore on the penis seventeen years before. Had four "shots in the arm" at that time.

Present Illness: About one year ago occasional inconstant but moderately severe bifrontal headaches had appeared.

Three months ago pain had developed in the right eye and orbit, and at the same time the right eye began to bulge and the upper lid to droop. The pain was severe and the proptosis developed rapidly. The patient was then operated upon (through the orbit), at another hospital, and a piece of tissue was removed; it was diagnosed as pseudotumor. He was then treated with iodides and "intravenous injections," probably neosalvarsan, and for a short time seemed to improve. A month after the operation the vision in this

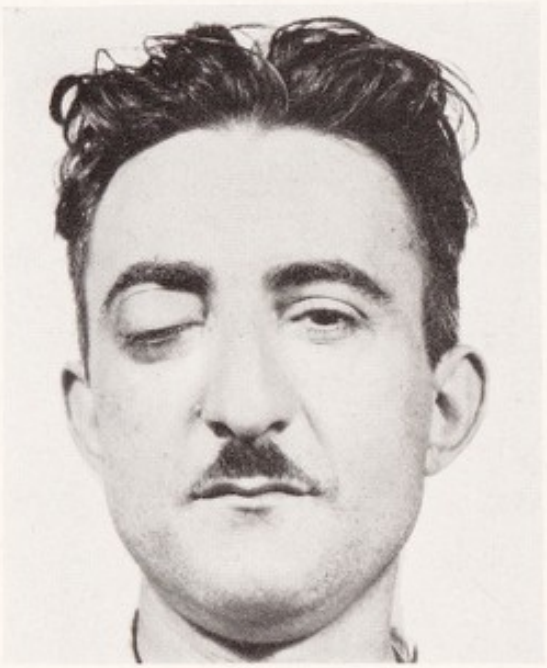


FIG. 47. Case 11. Before operation.

eye was affected and diminished rapidly. The pain in the eye and orbit became much more severe.

Examination: The patient was a large robust man. There was marked proptosis (8 mm.) of the right eye (Fig. 47), complete ptosis of the upper lid, and complete ophthalmoplegia. Visual acuity was 4/200 in the right eye, 20/15 in the left. The right pupil was normal and reacted to light and accommodation. The sclera on the right was much injected, with enlarged vessels. A mass could not be defi-

nitely palpated but there was some suggestion of one. There was papilledema of 2 diopters in the right eye and several small hemorrhages about the nerve head and overfilled and tortuous veins. The left eye ground was normal. The outline of the visual field for form in the affected eye was normal, but color perception was absent. Fields on the left were normal.

There were no other neurological findings except anesthesia of the right forehead resulting from the earlier operation. The corneal reflex was not impaired.

X-rays of the orbit and orbital foramina were normal.

The Wassermann reaction from the blood on numerous tests was negative.

Preoperative Diagnosis: Orbital tumor.

Operation, August 25, 1939: The usual hypophyseal approach with concealed incision was made on the right side. There was ample room after evacuating the cisterna chiasmatis, and no trace of an intracranial tumor was seen. The orbital roof was chiseled away; it was not abnormal. Two ethmoidal cells projected to the middle of the orbit and were opened when the roof was removed. The orbital capsule was roughened and had lost its normal luster; anteriorly there was a small area of tissue that looked like granulation tissue. When the orbital capsule was incised, a hard fibrous mass appeared; it was densely bound to the capsule. Orbital fat was not seen at any time during the operation. An attempt was made to split the tumor and remove it piecemeal, but it was so hard that attempts to incise it with the knife were ineffectual. With a sharp periosteal elevator the tumor was then shelled from its bed (Fig. 48); it appeared to come out completely, but the bed from which it came was dense fibrous tissue, and no normal structures could be seen within the orbit. The tumor was as large as a small pecan. The wound was closed in

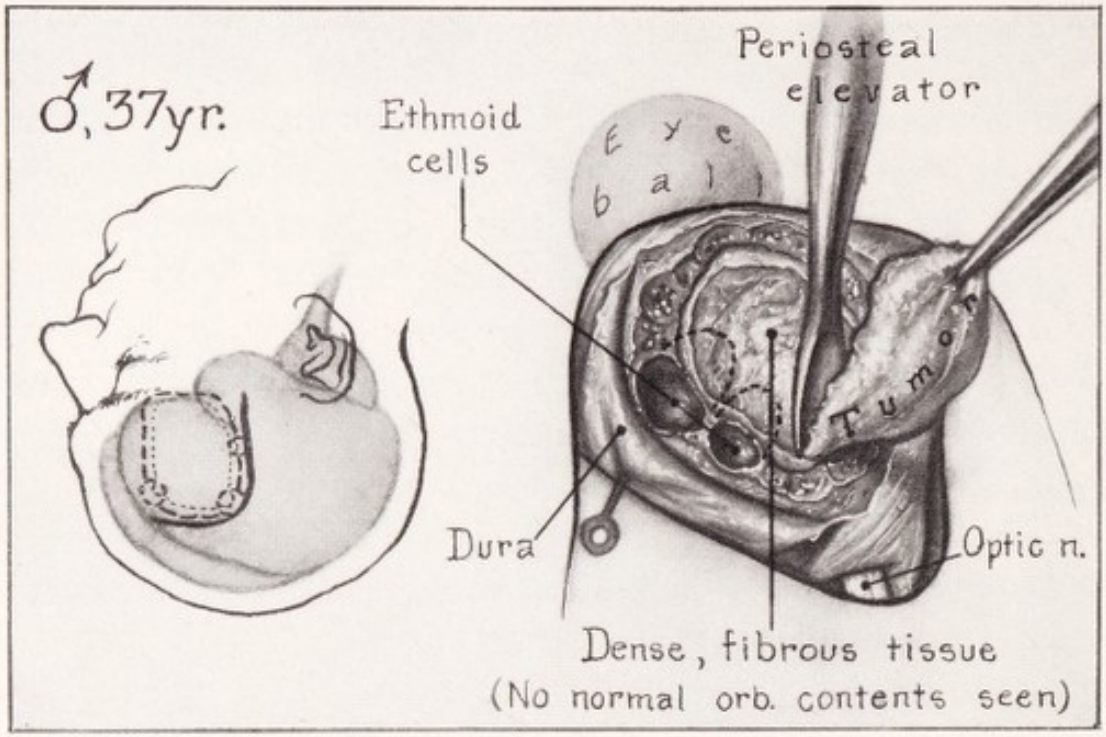


FIG. 48. Case 11. Operative exposure showing removal of tumor from apex of orbit.

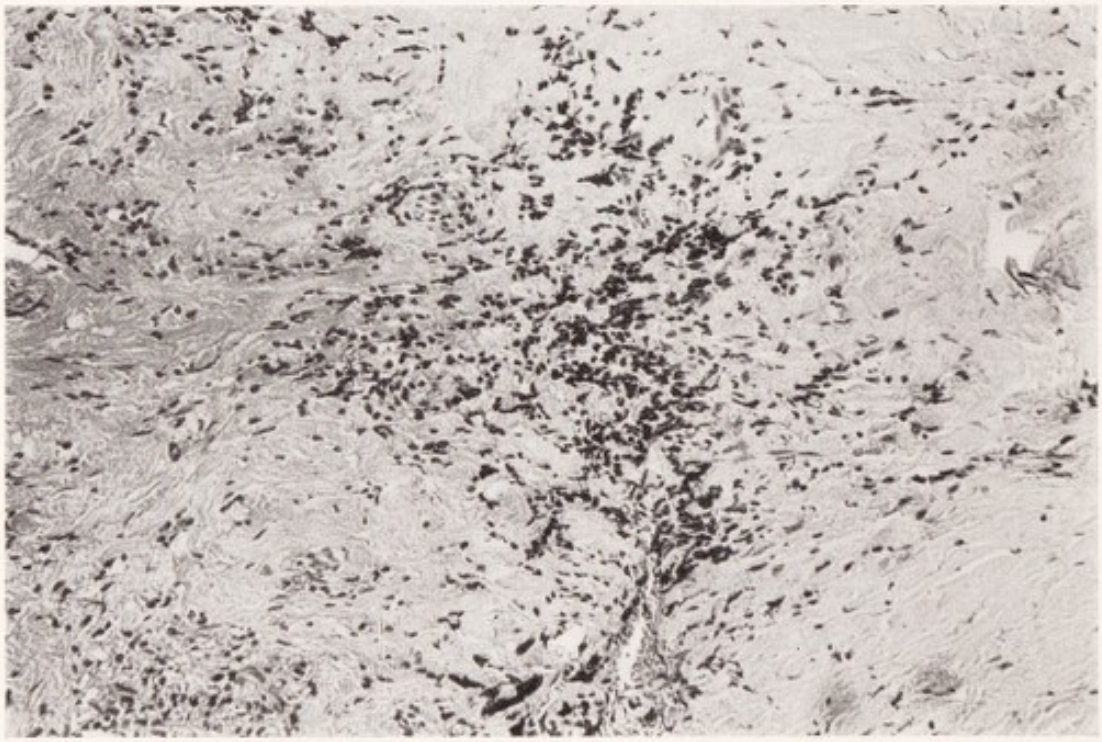


FIG. 49. Case 11. Photomicrograph of tumor: probably Schüller-Christian's disease.

the usual fashion; the orbital dura was placed over the opening in the ethmoidal cells.

On the following day a subdural hematoma was evacuated, after which the immediate recovery was uneventful until evidences of infection developed in the wound, which was reopened and the bone flap removed, on September 4, 1939. A month later the patient was discharged with a well-healed wound. By November 4, he was well and free from pain; the eyeball had receded to normal.

At the time of discharge the vision in the right eye was 20/200 (4/200 before operation) and the fields for form unchanged. The ptosis had not cleared, but there was marked improvement in the function of the extraocular muscles; the downward movement of the eyeball was normal and some return was evident in the remaining movements. The nerve head was flat, i.e., the papilledema had disappeared. There was slight pallor in the disc.

Subsequent Course: Report from Dr. Crump, January 18, 1940: "Patient is well and active, ptosis of right upper lid persists (it doubtless will remain). There is normal vision in the right eye of 20/20, 15° upward, 20° downward, 25° to right, and 20° to left."

Microscopic Pathological Report by Dr. Arnold R. Rich: "There are two sections of the same block. They show dense hyaline connective tissue with abundant mononuclear (lymphoid, plasma cell) infiltrations. In several areas there are loose collections of epithelioid cells and occasional giant cells, and in these foci are many eosinophils. Foam cells are few and are found particularly where adipose tissue is undergoing destruction. These sections are altogether like those of Case 8, except that the areas of necrotic collagen surrounded by palisades of connective tissue cells, as seen in one of the sections of the latter, are not present" (Fig. 49).

Diagnosis: Schüller-Christian's disease (probable).

CHAPTER III

DURAL TUMORS OF THE ORBIT

CASE 12

M. C., History number S-53823. White, female, age 13. Admitted: May 10, 1921. Discharged: June 5, 1921.

Diagnosis: Psammoma of optic sheath, bilateral. Referred by Dr. F. L. Waite, of Hartford, Connecticut. This case has previously been reported (*American Journal of Ophthalmology*, 5: 1, 1922) and was the first in which the intracranial operative procedure was used.

Present Illness: The patient was a very bright and physically normal girl. When she was seven years old, or six years before admission, she had a spell of vomiting lasting a week. There was no associated abdominal pain or other disturbance that should accompany an acute abdominal ailment, but during this period of vomiting, diplopia developed and the left eye turned outward; the intracranial origin of the vomiting was, therefore, evident. There was no headache associated with the attack. Within a week the crossed eye had returned to normal. About this time, dimness of vision first appeared in the right eye. There was no pain or aching in the eyeball and none referred elsewhere. Little further change occurred until three years before admission, when during a mild attack of influenza, another vomiting spell persisted for several days and the right eye again became crossed. The vision was then so much impaired that she had to leave school. The crossed eye gradually returned to normal. During the past three

years there had been three additional attacks of vomiting, each lasting nearly a week, and at each time both eyes were crossed. Coincident with the strabismus there was drooping of the upper eyelids—apparently bilateral. Although the strabismus improved after the subsidence of the vomiting, a more or less permanent weakness of the eye muscles persisted and varied in intensity from time to time. During the last three attacks, both eyes ached slightly, but this had subsided with the disappearance of the vomiting. Exophthalmos was never observed in either eye.

The loss of vision had been very gradually progressive in both eyes, but more in the right. Dr. Waite's careful notes showed that the patient was blind in the right eye at least three years before—at which time she first consulted him. At that time (1918) the vision in the left eye was 20/60. Failing gradually, it was 10/200 in March, 1920, and 8/200 in June, 1920, and was the same on admission to the Johns Hopkins Hospital (May, 1921). The patient had never had a headache in her life and, except for the loss of vision, would have been "in fine shape."

Examinations: From the physical, neurological, and special examinations, the following positive and negative items are summarized:

1) The right eye was totally blind.

2) The visual field for form was preserved in the left eye, but was somewhat contracted; visual acuity was 8/200; there was complete loss of color perception, no hemianopsia.

3) Both discs had a brilliant atrophic pallor; the margins were sharply defined, the lamina cribrosa very distinct.

4) The right eye moved outward, inward, and slightly downward, but not upward; the left eye laterally, slightly downward, not medially or upward. There was a slight but definite ptosis of both upper lids. Both pupils reacted fairly actively to light, directly and consensually.

Operation, May 14, 1921: A fairly large bone flap was turned down on the right (Fig. 50). A small collar-like tumor surrounded the optic nerve at the optic foramen. It was about 6 to 7 mm. thick and of quite uniform depth around the entire circumference of the nerve. It protruded about 1 cm. along the intracranial course of the optic nerve and could be seen extending through the optic foramen into

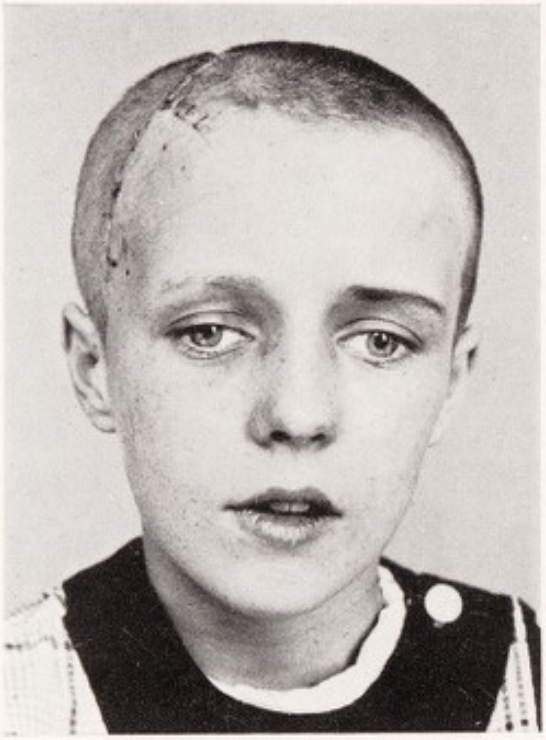
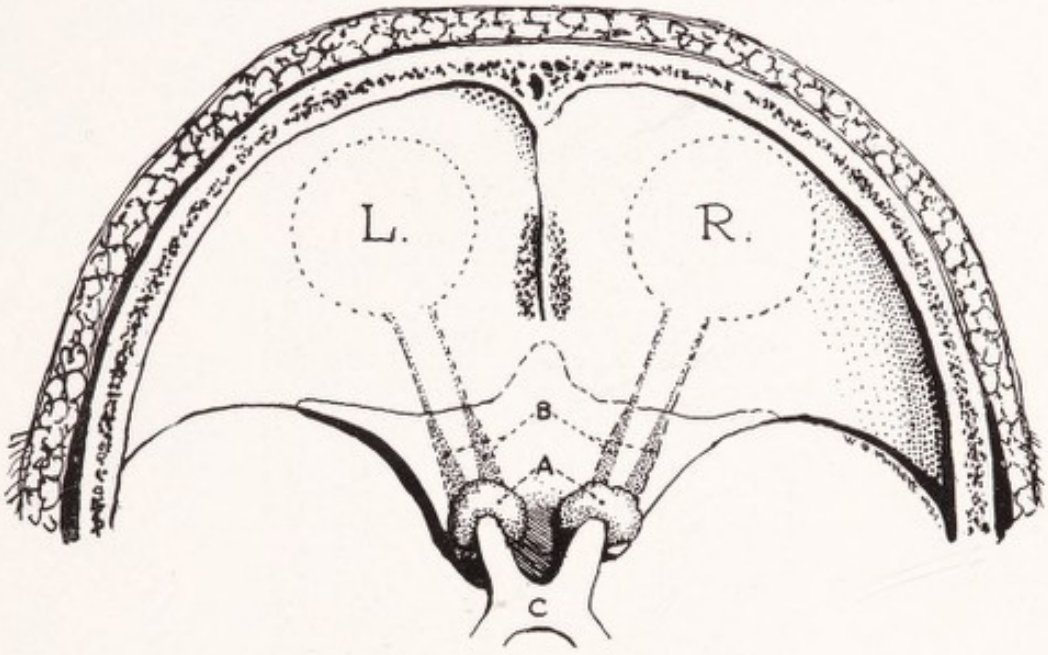


FIG. 50. Case 12. Postoperative photograph. Bilateral symmetrical cuff-like dural endothelioma arising at optic foramina and extending into each orbit. In this, the earliest case in the series, a much larger bone flap was used.

the orbital chamber. It was quite hard and when the outer part of the growth was cut away with scissors, a deep constriction in the nerve could be seen. A similar though smaller tumor was seen on the left optic nerve, the exposure being adequate for the inspection of both nerves. Two weeks later the wound was reopened, the posterior half of the orbital roof chiseled away, and the tumor followed

almost to the eyeball (Fig. 51). The roof of the optic foramen was included in the bony removal. The growth surrounded the optic nerve rather loosely; the maximum depth of the tumor was about 1 cm. The intraorbital portion of the growth was easily shelled from the optic nerve, to which it was but slightly bound. At the optic



A = Exposed Tumor – B = Tumor on Dural Sheath of Nerve in Orbit
C = Optic Chiasma

FIG. 51. Case 12. Sketch of operative findings, bilateral dural tumors. This patient was alive and well, though blind, 18 years later.

foramen, however, the growth was so tenaciously bound and was so deeply constricting the nerve that its removal was impossible.

The postoperative course was uneventful. The patient's vision was unchanged at the time of discharge from the hospital.

Microscopic Report: Dural fibrous tissue filled with psammoma bodies (Fig. 52).

Diagnosis: Psammoma (dural meningioma).

Subsequent Course: Over a period of several years the vision in both eyes gradually diminished until total blindness resulted. When last heard from, in May, 1939, the patient was blind, but otherwise in good health.

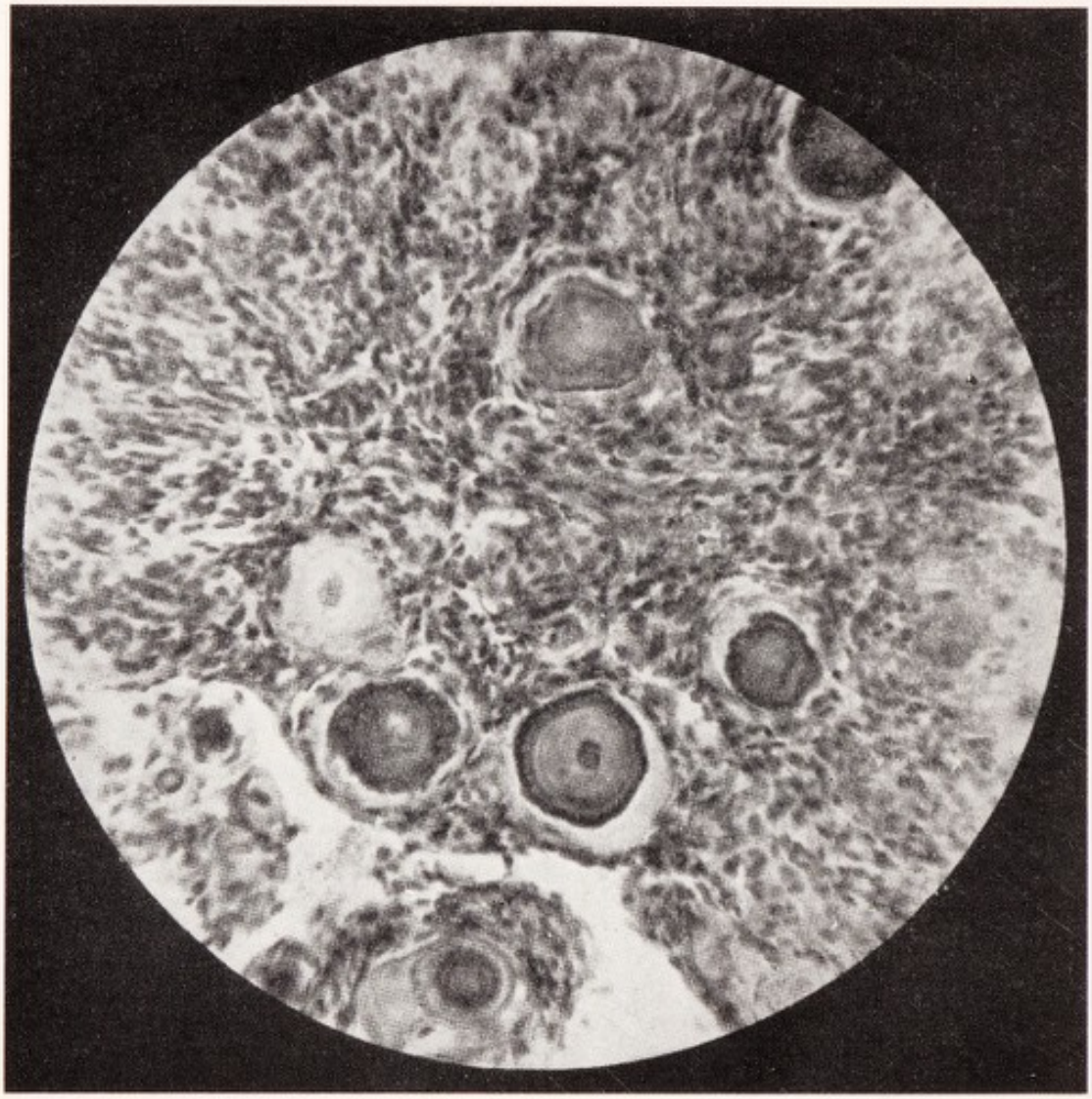


FIG. 52. Case 12. Photomicrograph of tumor, showing numerous psammoma bodies.

Comments: This is the first patient upon whom the intracranial approach was used for a combined intracranial and intraorbital growth. The case was originally reported, as above, in the year following the operation. In view of the

great frequency of direct extension of so many growths from the orbital to the intracranial chamber, or vice versa, the need for such an approach to both chambers simultaneously was emphasized. The succeeding case (below) was operated upon several years previously, but the removal of the orbital roof was not deliberate in this case; the orbital roof came out with the large bony tumor.

CASE 13

J. W., History number 44805. Colored, male, age 23. Admitted: October 18, 1917. Discharged: February 18, 1918.

Diagnosis: Spindle-cell sarcoma with massive hyperostosis of skull.

Complaints: Lump at the left side of the nose and protrusion of the left eye.

Family and Past Histories: Negative.

Present Illness: The patient said he had had an enlargement on the left side of his nose all his life, or at least as long as he could remember, and had had difficulty in breathing through this side of the nose. The lump had gradually increased in size (Fig. 53). Eight months ago it had been operated upon in another hospital and a tumor removed. The wound had broken down and had been discharging ever since. There had been some frontal headaches in the past years; the headaches came in spells, lasting for a couple of hours.

Examination: The patient was a well-developed man with a very marked prominence of the whole frontal region, but more on the left. There was a discharging sinus along the inner canthus of the left eye and a slightly protruding mass which was very hard and extended backward into the orbital cavity. The left eye protruded markedly and was

pushed outward and downward. The mass fused with the supraorbital ridge, the latter protruding markedly. The vision in the left eye was poor; there was double vision.

FIG. 53

FIG. 54

FIG. 53. Case 13. Exophthalmos with marked hyperostosis of the frontal bones. Tumor had projected into inner canthus of the eye and had been opened locally.

FIG. 54. Bilateral frontal hyperostoses removed at operation. One of the nodules is seen projecting to the right. The soft tissue tumor is a spindle-cell sarcoma.

X-rays showed a tremendous thickening of the bone over the entire frontal region and extending back to the parietal bone.

Operation, October 19, 1917: Owing to the discharging sinus we were reluctant to undertake an operation upon the skull, but the tumor was so large and progressing so rapidly, that we felt it necessary to attempt its removal.

A long curved incision was made from one temple to the

other and just back of the hairline. The galea and skin were reflected forward. The bone was so dense even back of the hairline that it was difficult to make the openings with the burr, and it was impossible to use the DeVilbiss or Gigli saw. The large bone-cutting forceps were used to cut the bone between the big burr openings. Finally a curved incision was made through the bone from the middle of the right orbit down to the outer margin of the left orbit. Since the flap of bone was so wide, it was necessary to divide it in the midline in the same manner with burr openings and the bone-cutting forceps, so that the mass was removed first to the right and then to the left. The bone was then broken across at the supraorbital margin and much of the orbital roof and all of the supraorbital ridge came out as the bone was broken (Fig. 54). Much of the roof of the orbit was involved in this great thickening of bone. As the mass came out we saw protruding soft tissue tumors that filled the sphenoid and several others filling the ethmoid, one protruding into the antrum. The main mass of bone was quite hard and in marked contrast to the soft tissue tumors at the periphery. These lifted off the paranasal sinuses like a cluster of grapes. The dura was not opened so that there was no leak of cerebrospinal fluid.

Following the operation the wound became infected. This prolonged the patient's stay in the hospital for four months. At the end of this time there still remained the original discharging sinus, but the rest of the wound had healed.

Pathological Note: Enormous thickening of bone, shown in the accompaning pictures (Figs. 53 and 54). Sections of it show sarcoma in the marrow spaces. The protruding soft tissue tumors are spindle-cell sarcoma of the same type as that in the bone.

Subsequent Course: Several letters have been sent to the patient at his original address, but all have been returned unclaimed.

CASE 14

D. H., History number U-58814. White, female, age 24. Admitted: November 2, 1934. Discharged: November 13, 1934. Readmitted: March 28, 1935. Discharged: April 10, 1935. Died: of recurrence, September 21, 1936.

Diagnosis: Malignant dural meningioma.

Complaints: Headache, vomiting, and blurring of vision.

Family and Past Histories: Negative.

Present Illness: Two and one-half months before admission (August, 1934) severe headaches developed in the left frontal region. At first they were infrequent, but they soon became almost constant, although the intensity varied. The patient insisted that there had been some exophthalmos of the left eye for about twelve years, that it had come and gone, but that it had been more marked and persistent in the past two months. About this time her husband had noticed that her left pupil was constantly dilated, and that she became more irritable and more easily upset than formerly. Two months ago she had noticed a very disagreeable odor as if something were burning. There had also been weak spells in which she felt faint, but there was never any actual loss of consciousness. During these attacks there was ringing in her left ear. There had also been pain along the left side of the nose, but independent of these attacks. For the past six weeks, when her headaches had been severe, she had vomited on several occasions, without being nauseated. In the past two months her vision had been blurred in the left eye; she saw double when looking towards the right, but not when looking ahead or to the left.

Physical examination: Negative.

Neurological Examination: There was impaired vision in the left eye (acuity 20/30), whereas the right was normal (20/15). The visual fields were normal. There was no papilledema, but the veins in the left eye grounds were overfilled though not tortuous. The left pupil was widely dilated and reacted neither to light nor accommodation. The upward and inner movements of the eyeball were

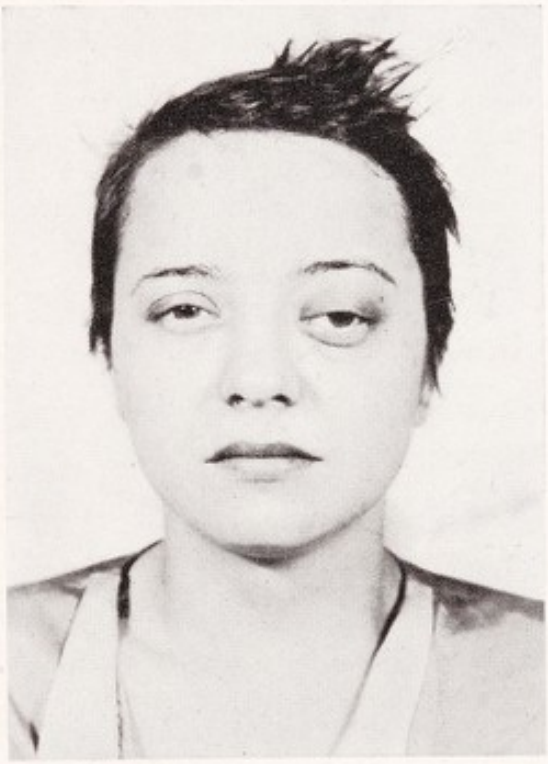


FIG. 55. Case 14. Exophthalmos arising from malignant dural endothelioma in middle fossa and extending into orbit.

markedly limited. There was no ptosis. The proptosis measured 6 mm. (Fig. 55).

Diagnosis: The uncinate attacks, headaches, fainting sensations, and ringing in the ear definitely indicated an intracranial lesion. Moreover, the X-rays showed extensive irregular linear areas of calcification throughout the region of the left temporal lobe. The optic foramen was not enlarged.

Operation, November 3, 1934: The hypophyseal bone flap

(with concealed incision) was made a little larger than usual and fully exposed both the frontal and temporal regions. Far beneath the frontal lobe one could see a hard, well-encapsulated, nodular tumor bulging forward from the temporal fossa and pushing the Sylvian vessels anteriorly; it did not actually project into the anterior fossa. One could feel a very hard temporal lobe, but the tumor was everywhere beneath the cortex. The convolutions were flat and pale; the ventricular needle struck tumor at a depth of 1 cm. The superimposed temporal lobe was excised laterally to the Sylvian vein. A very hard, reddish-brown tumor filled the middle fossa and lay against the base of the skull. An attempt to remove the interior of the tumor was attended with little progress because the tumor was too hard; moreover, it was exceedingly vascular. The tumor was shelled out with the finger after first separating it from, and carefully avoiding injury to, the middle cerebral artery. It had a very dense attachment along the lesser wing of the sphenoid, which was thickened throughout in response to the overlying tumor. We felt that probably this hyperostosis was responsible for the degree of exophthalmos, and therefore did not open the orbital cavity. One could see the exposed middle cerebral artery for a distance of about three inches, but it was intact (Fig. 56). A nodule of the tumor broken off from the main mass was projecting mesially from the margin of the dura lining the cavernous sinus and pushing aside the third nerve, which was elongated to two or three times its normal length and flattened from the pressure. This nodule of the tumor was also removed. The tumor weighed 44½ grams.

The postoperative recovery was uneventful, the patient leaving the hospital eleven days after the operation. The exophthalmos remained unchanged.

Subsequent Course: Five months after the operation she returned because the left eye was blind and the exophthalmos had increased. For a month following the operation the vision in her left eye had not changed.

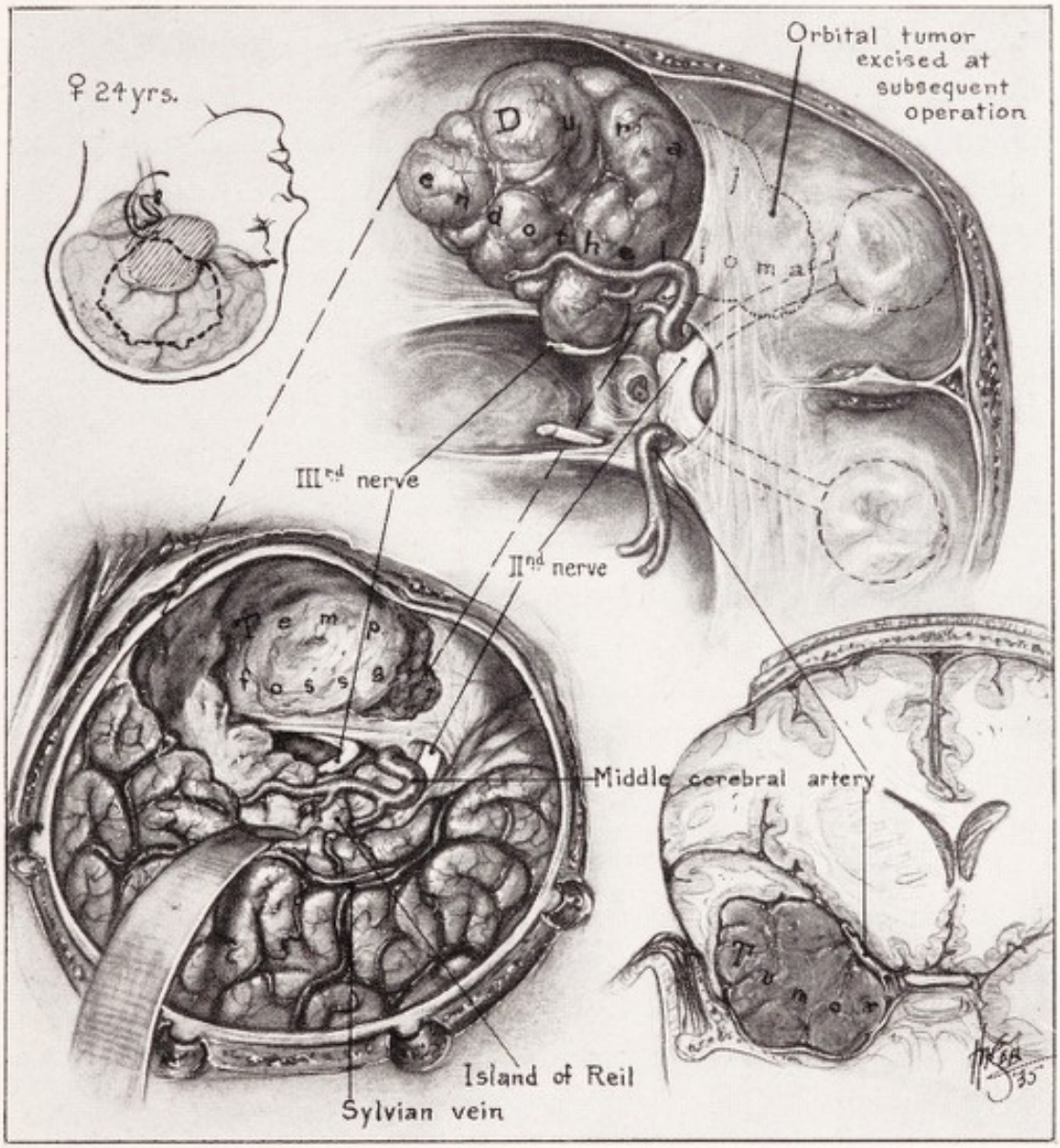


FIG. 56. Case 14. Operative sketch, showing site and relative size of tumor.

For two months she had no headaches, but then these also returned, differing from those before the operation in that the ache was more in and above the region of the left eye. There had been no vomiting and no return of the uncinate

attacks. Neither speech nor mentality had been affected following the removal of the left temporal lobe. The exophthalmos now measured 10 mm. and the movements of the eyeball were decidedly less than on the previous admission. The sixth nerve was completely and the third nerve partially paralyzed.

Operation, March 30, 1935: The old incision was reopened and the bone flap elevated. The middle fossa was first explored and found to be free of tumor. The space was filled with fluid, which was evacuated. The roof of the orbit was then removed; immediately beneath it was a dense mass which felt very much like another orbital roof because it was so hard and smooth. The tumor filled about two-thirds of the orbit and was firmly fixed posteriorly to the apex. The mass, which appeared to be pure bone, was shelled out with a periosteal elevator; it was as large as a hickory nut. One could then see soft or tumor tissue inferiorly and laterally, although it was still quite firm and contained much bone. The mass could be shelled out, except at the sphenoidal fissure, through which it could be seen directly extending. The lesser wing of the sphenoid was then removed to a point alongside the optic foramen. The optic nerve was seen to be surrounded by tumor, and, in a further effort to prevent recurrence, the optic nerve was sectioned just back of the eyeball and again at the optic foramen. The tumor weighed 11.8 grams.

The postoperative course was uneventful, the patient leaving the hospital in twelve days.

Microscopic Report: "Columns of tumor cells of the malignant dural endothelioma type fill many of the marrow spaces [Fig. 57]. The intracranial tumor is very cellular and contains almost no fibrous tissue. The cells are of round and spindle type with little arrangement into groupings of any

kind. The nuclei are round and vesicular and slightly larger than small round cells. The tumor might well pass as a mixed round- and spindle-cell sarcoma."

Subsequent Course: Two months later the patient returned because of corneal ulceration that had developed the night before. The lids were sutured.

In February, 1936 (ten months following the operation), the exophthalmos, which had entirely disappeared after

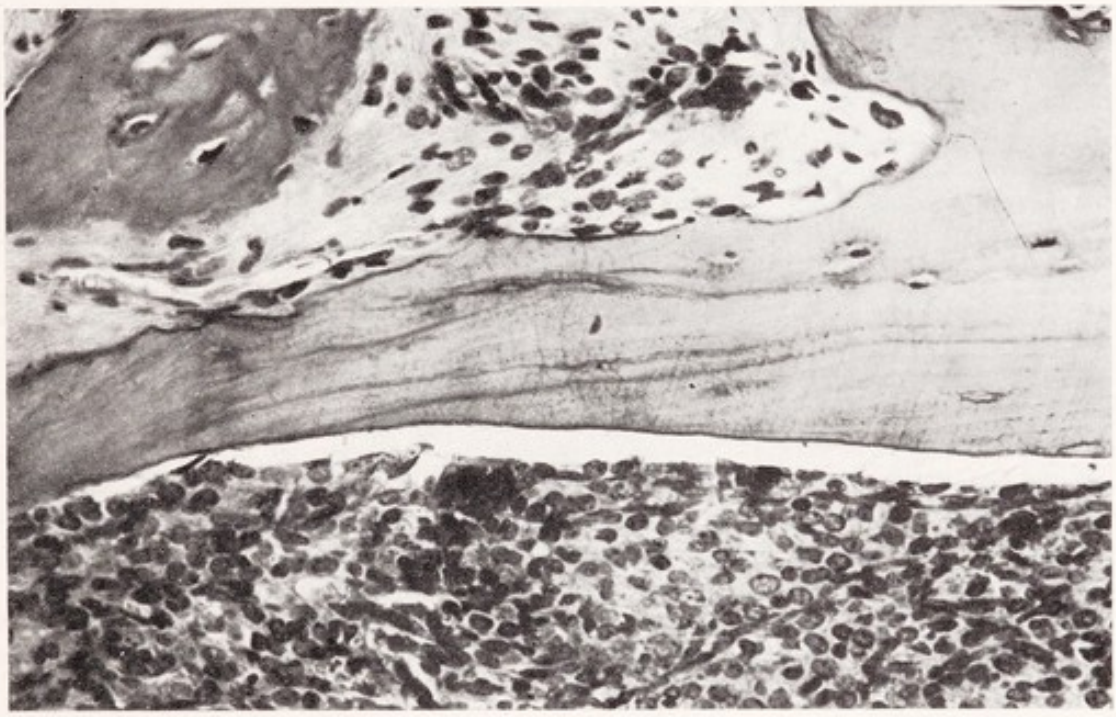


FIG. 57. Case 14. Photomicrograph of tumor, showing invasion of bone. This tumor might well be taken for a round- and spindle-cell sarcoma.

the operation, was again in evidence and was rapidly increasing. Over the forehead and over the eyeball metastatic nodules of tumor could be seen. Vomiting and headaches again returned. There was anesthesia throughout the left side of the face. The patient had a sallow complexion and was losing weight. Repeated X-ray treatments were without any effect. The patient died on September 21, 1936, almost two years after the first operation.

CASE 15

D. H., History number U-100411. White, female, age 27. Admitted: March 18, 1937. Discharged: April 2, 1937.

Diagnosis: Dural meningioma. (Referred by Dr. S. L. Ledbetter, Jr., of Birmingham, Alabama.)

Complaints: Loss of vision in the left eye; protrusion of the left eye; dull ache behind the left eye.

FIG. 58. Case 15. Low grade of exophthalmos from dural tumor in middle fossa.

Family and Past Histories: Negative.

Present Illness: Six months ago the patient had noticed on arising that the central vision in the left eye was gone; the peripheral vision was still present. She had a slight head cold at the time and thought this might have been responsible.

Six weeks ago a dull ache had developed back of the eye, and at the same time the eye was seen to be protruding (Fig. 58). The patient was eight months pregnant at this time

and although there were no signs or symptoms of toxemia of pregnancy, an artificial abortion was advised and carried out. No improvement or increase in symptoms followed. The vision in the right eye had remained normal.

Physical Examination: The patient was a small, slender, but well-nourished woman. Blood pressure was 130/90. Wassermann reactions in the blood and spinal fluid were negative.

Neurological Examination: Vision in the left eye was practically nil; there was a small intact patch of vision in the far nasal and temporal fields, but the acuity in these areas was only 2/100. The vision in the right eye was normal. The left eye protruded 3 mm. The left disk was pale and glistening. Extraocular movements were normal.

X-rays of the head were normal. The left optic foramen was slightly larger than the right and its margins less sharply defined.

Operation, March 20, 1937: The usual hypophyseal approach with concealed incision was made. Across the optic nerves at the beginning of the chiasma was an unusually low anterior cerebral artery that was tightly drawn across the nerves, more so on the left. Had I not known there was a tumor, this artery might well have been suspected of being the cause of the blindness, but the optic nerve was not elevated by the tumor and doubtless, therefore, did not push the optic nerve into the artery.

Evacuation of the cisterna chiasmatis provided ample room and the tumor immediately appeared in the middle fossa and entirely back of the lesser wing of the sphenoid; none therefore was present in the anterior fossa. The tumor was well encapsulated but tightly bound to the dura of the middle fossa, about half of which it covered. It was clearly a dural tumor of the softer, more friable type, so that it easily broke

into fragments when the forceps were applied and much of it could be drawn into the suction tube. I should estimate its weight at about 30 grams. The tumor was attached to the internal carotid artery. A fragment of the tumor had

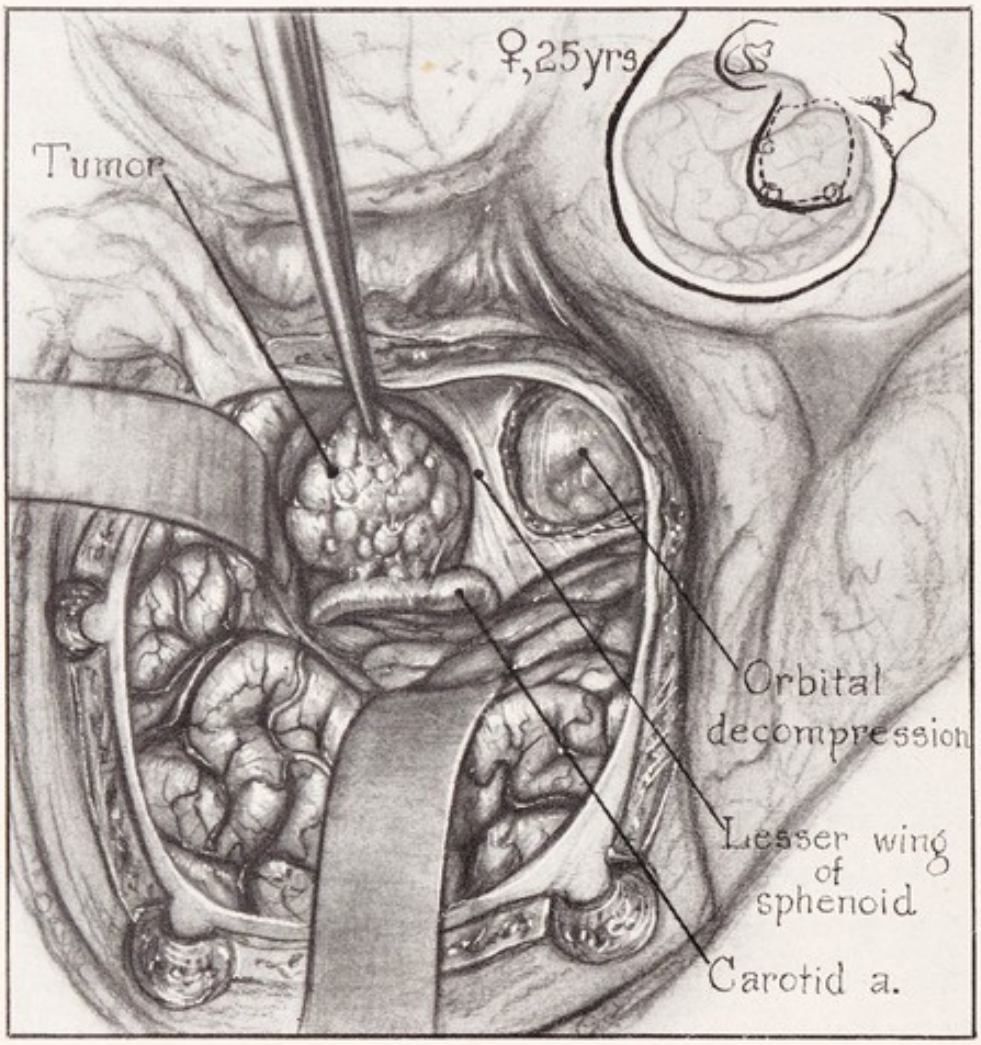


FIG. 59. Case 15. Operative sketch of exposed tumor, a dural meningioma, which had attached itself to carotid artery. In order to avoid injury to this vessel a small fragment of tumor was left attached to it.

to be left on the artery (Fig. 59). The tumor did not actually touch the optic nerve. One could not see prolongation beneath the lesser wing of the sphenoid into the orbit, but presumably there was such. At any rate, the roof of the orbit was removed, leaving only the lesser wing

behind. The orbital fat bulged into the defect and was cut away, but no attempt was made to find and remove tumor tissue because the vision of this eye was permanently lost in any case and the decompression of the orbital roof could be expected to relieve the exophthalmos.

The postoperative course was uneventful; the patient left the hospital two weeks later.

Microscopic Note: "A cellular dural meningioma. In places the cells are grouped in columns of varying size. In most of the section the cells are without arrangement. There is little fibrous tissue. No calcification or psammoma bodies are seen in the section."

Subsequent Course: The patient was last seen October 1, 1938—eighteen months after operation. She thought the exophthalmos was slightly greater; it then measured 5 mm. —an increase of 2 mm. Her headaches had not reappeared. The vision in the left eye was nil; that in the right unimpaired.

Since her operation there has been a profound sexual change. For three days after the operation she menstruated, but has not done so since. Her breasts were lactating at the time of the operation and have continued to do so. They secrete several cc. of milk daily; this drains spontaneously, more at night. Libido and sexual satisfaction have practically disappeared. There has been no diabetes insipidus. She has gained 10 lbs. in weight (now weighs 110 lbs.).

A letter of November 1, 1939, stated that her condition was essentially unchanged.

CASE 16

A. Z., History number 58849. White, female, age 44. Admitted: February 24, 1923. Discharged: March 10, 1923. Died: April 4, 1933.

Diagnosis: Dural meningioma with hyperostosis of skull and orbital roof. (Referred by Dr. Riddick Ackerman, of Waterboro, South Carolina.)

Complaint: Swelling of the eye.

Family and Past Histories: Negative.

Present Illness: After her recovery from an attack of "sleeping sickness," three years before, the patient's husband

FIG. 60. Case 16. Mild degree of exophthalmos due to a hyperostosis of skull and roof of orbit, this in turn due to a dural meningioma "en plaque."

noticed that her right eye was more prominent than the left. Two years ago, the patient had noticed a diffuse hard swelling in the right temporal region; this had gradually increased and now measured 7 cm. in diameter.

Eleven months ago the right eye had become suddenly very much more prominent, and its size increased for six months (Fig. 60). Since then the patient thought the protrusion had actually diminished. However, the swelling

on the right side of the head had slowly increased. There had been no double vision and no loss of vision in the right eye. There had been slight headaches, but never severe, and they always subsided after a short time when she lay down.

Physical and Neurological Examinations: Negative, except for the local condition. A diffuse bulging of the right temporal region extended to the orbital ridge. The right eyeball protruded markedly but with very little displacement. The right pupil was somewhat larger than the left and reacted a little more sluggishly than the left. The ophthalmoscopic picture was not abnormal. The visual fields and the visual acuity were normal.

X-rays showed increased density in the entire roof of the orbit, extending laterally over into the adjacent frontal and temporal bones.

Operation, February 28, 1923: The usual hypophyseal approach, with the concealed incision, was made. Later a secondary bony defect was made posteriorly to provide more room. The bone flap broke just mesially to the thick bony swelling in the temporal region. The bony growth was then rongeured and chiseled from the side of the skull as far as the zygoma. It was a very hard tumor, 9 x 8 cm. in diameter and about 6 cm. thick. It extended directly into and involved the roof of the orbit. The orbital swelling extended to the midline of the skull and backward to the sphenoidal wing and the optic nerve. The entire mass was chiseled away up to but not in the paranasal sinuses and back to but not including the wing of the sphenoid. The upper and outer sides of the optic nerve were uncovered, because the optic nerve was surrounded by the tumor. The orbital growth extended mesially into the paranasal sinuses; this extension could not be removed because of the danger of

infection. For a time the removal of the orbital roof was continued without opening the dura, but when it was found that more room was necessary the dura was opened and the frontal lobe retracted and the cisterna chiasmatis evacuated. It was then found that a soft cellular tumor lined the intracranial surface of the dura covering the roof of the orbit and extended laterally on the side of the skull. This

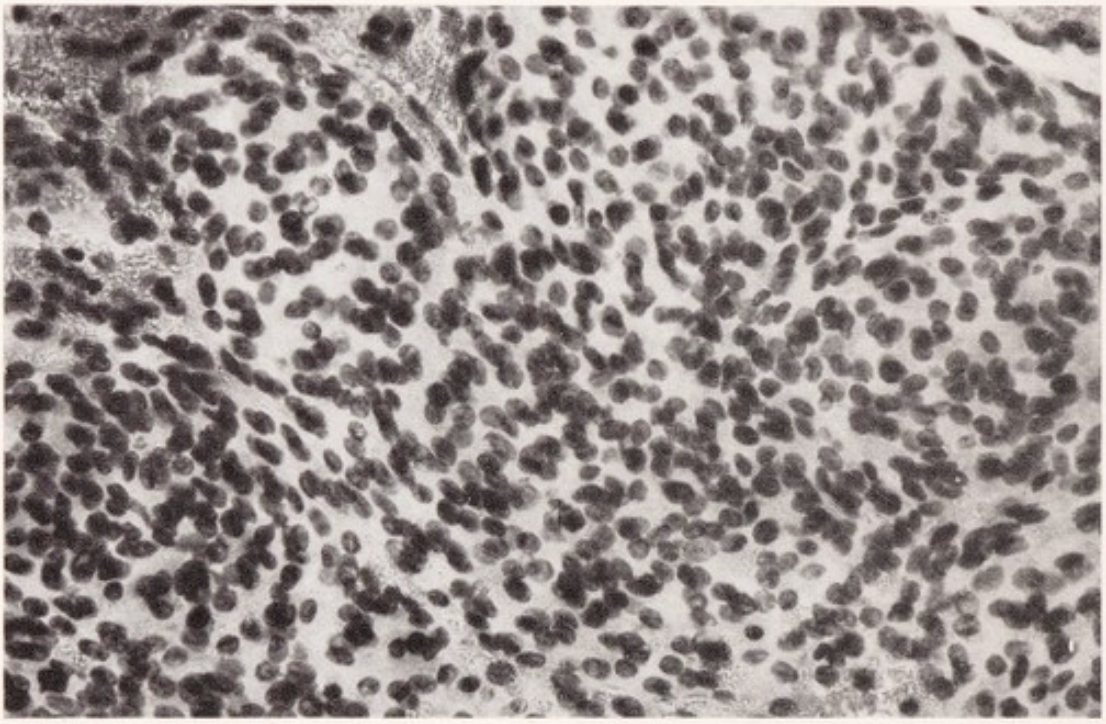


FIG. 61. Case 16. Photomicrograph of tumor: dural meningioma of very cellular type, which might well be taken for sarcoma.

was a thin, evenly distributed layer of soft tissue, fairly firmly bound to the dura; at the outskirts, however, the cells were loosely attached, and in many places parts of the tumor had broken loose and were at a distance from the main mass. At no place was the tumor more than ½ cm. thick, but it covered an area of about 6 x 5 cm., which corresponded fairly well with the orbital roof. Had it not been that the dura was opened to make the operative approach easier, this

tumor would not have been found. Most of the growth was removed, but part of it extended beyond our reach.

Microscopic Study: "The intracranial film of tumor lining the dura, and so loosely attached to it, is made up of masses of small round cells, closely packed, with some separation into clusters by thin lines of connective tissue, of which the amount is exceedingly small. It looks like a round-cell sarcoma [Fig. 61], but there is some resemblance to other cellular dural tumors in this group. Since it lined the dura exclusively it doubtless was of dural derivation."

Subsequent Course: The patient made an uneventful recovery and was discharged two weeks later (March 10). She was last seen by the writer in May, 1928, at which time she was perfectly well. Subsequent letters brought no reply. A recent letter from Dr. Ackerman reported her death on April 4, 1933—ten years after the operation. For two or three years prior to her death there was evidence of recurrence, the protrusion of the eye finally becoming very pronounced.

CASE 17

E. W., History number U-60233. White, female, age 43. Admitted: January 21, 1935. Discharged: February 8, 1935.

Diagnosis: Dural meningioma with hyperostosis of skull and orbital roof. (Referred by Dr. J. Heyward Gibbes, of Columbia, South Carolina.)

Complaints: Protrusion of the right eye and blurring of vision in this eye.

Family and Past Histories: Negative.

Present Illness: About a year ago the patient's family had noticed that the right eye was more prominent (Fig. 62). This had appeared to increase steadily up to four months ago,

when it had suddenly become much greater. During the past month the vision in this eye had become blurred, and there had been double vision when looking to the extreme right. There had been no pain, headache, or other symptoms.

Physical and Neurological Examinations: Negative except for the local condition. The patient was a quite obese woman, but in good health. Examination of the eyes by Dr. Reese

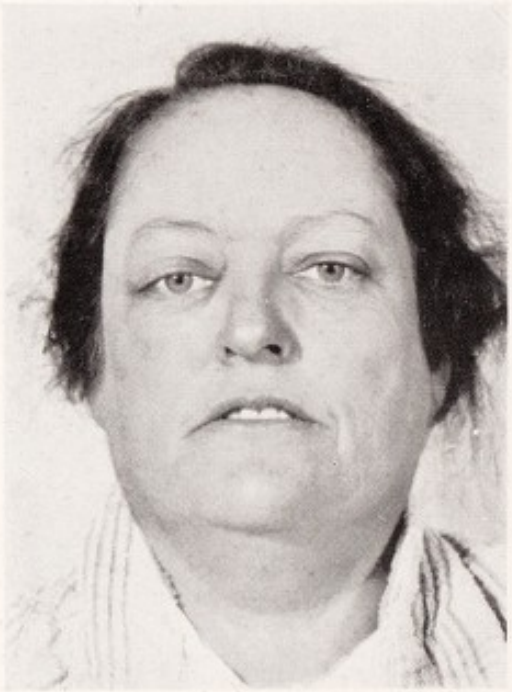


FIG. 62. Case 17. Mild grade of exophthalmos due to hyperostosis of skull and orbital wall.

gave the following findings: There was marked proptosis of the right eye, measuring 8½ mm.; the right disc was a little pale; there was no papilledema; the veins were somewhat engorged. The visual acuity was 20/40 in the right, and 20/20 in the left eye. Visual fields were normal; the blind spots were not enlarged.

X-rays showed a marked diffuse thickening of the outer surface of the skull in the frontal and temporal regions, extending into the roof of the orbit.

Operation, January 23, 1935: A right hypophyseal approach, with concealed incision, was made. The dura was quite tight and despite evacuation of the cisterna chiasmatis there was less intracranial room than usual. The dura was stripped from the orbital roof, which was distinctly more convex than normal. With the chisel, a primary opening was made into the orbital roof and this was extended by further piecemeal chiseling; this thickened bone measured

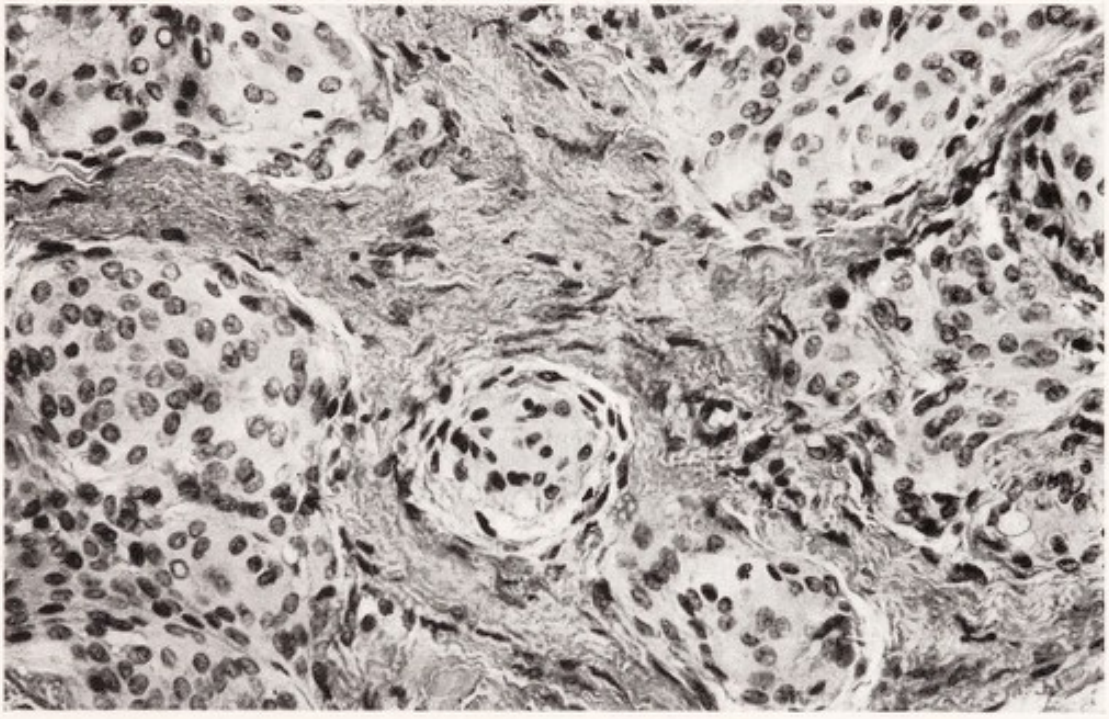


FIG. 63. Case 17. Photomicrograph of tumor, showing columns of dural cells which had invaded bone and were responsible for hyperostosis.

about 2 cm. at the maximum. It extended anteriorly, internally, and laterally to the margin of the orbit, and posteriorly stopped short of the optic foramen and the wing of the sphenoid. All of the mass was chiseled away. At no time were the paranasal sinuses opened. The covering of the orbit was firmly attached to the undersurface of the orbital roof and was thickened and roughened, looking like pale granulation tissue. Much of this thickening was excised and some of the orbital fat was included. Excepting

this tissue there was no gross soft tumor. The increased thickening of the bone in the lateral margin of the skull was also removed.

There was very little bleeding at any time. The wound was closed in the usual fashion, the bone flap replaced and wired. The patient left the hospital two weeks later.

Microscopic Report: Under the microscope the roughened tissue at the posterior part of the orbital covering was found to be tumor tissue. It was made up for the most part of closely packed cells of dural type. In other places, columns of cells of this type were lying in fibrous tissue (Fig. 63).

Sections of the hypertrophied bone showed columns of dural cells in the medullary spaces.

Subsequent Course: A letter from Dr. Gibbes in June, 1939, stated that he had last seen the patient three years previously, but had since been unable to contact her.

CASE 18

F. R., History number U-74229. White, female, age 40. Admitted: November 19, 1936. Discharged: December 12, 1936.

Diagnosis: Bilateral frontal dural meningioma with hyperostosis and destruction of orbital roof.

Complaints: Tumor in the right frontal region and protrusion of the right eye.

Family and Past Histories: Negative.

Present Illness: At the age of five the patient had fallen, striking the right forehead. She had not lost consciousness and the wound healed rapidly. There were no symptoms until the age of seventeen when an enlargement of the forehead developed at (she thought) the site of the old injury in the right frontal region. This swelling had very slowly but steadily increased up to the present time (Fig. 64).

The growth, therefore, was of at least twenty-four years' duration. There had been very few symptoms. During her early twenties there were slight dizzy attacks. Six years ago a dull, generalized headache had lasted two weeks. There had been no further symptoms until two years before, when headache and pain had developed over both frontal regions, but more on the right side. About this time exoph-

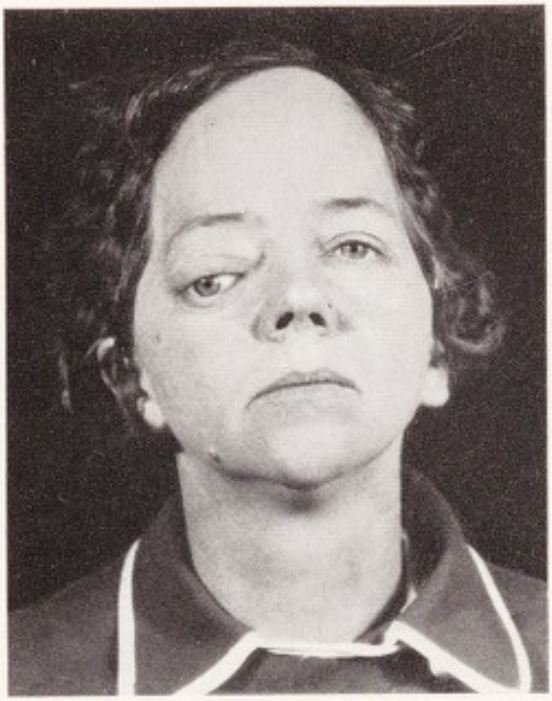


FIG. 64. Case 18. Lateral and front views: tremendous hyperostosis of entire frontal region and marked exophthalmos and downward displacement of right eyeball.

thalmos of the right eye had appeared and gradually increased. The eye was displaced outward and downward. On numerous occasions she had had diplopia and blurring of vision.

Examination: There was marked protrusion of the entire frontal region. From a central boss the swelling extended backward beyond the hairline, gradually becoming less pronounced. The right eye protruded markedly and was displaced downward and outward. Large veins could be

seen leading up to the central mass in the forehead and extending downward into the orbit. No murmurs were heard on either side. Both discs were sharply outlined; both visual fields were normal. Vision was 20/20 in the right eye, and 20/30 in the left. The pupils were equal and reacted to light and accommodation. The corneal reflex was present and equal on the two sides. At the center of the swelling one could feel a small defect in the bone, but

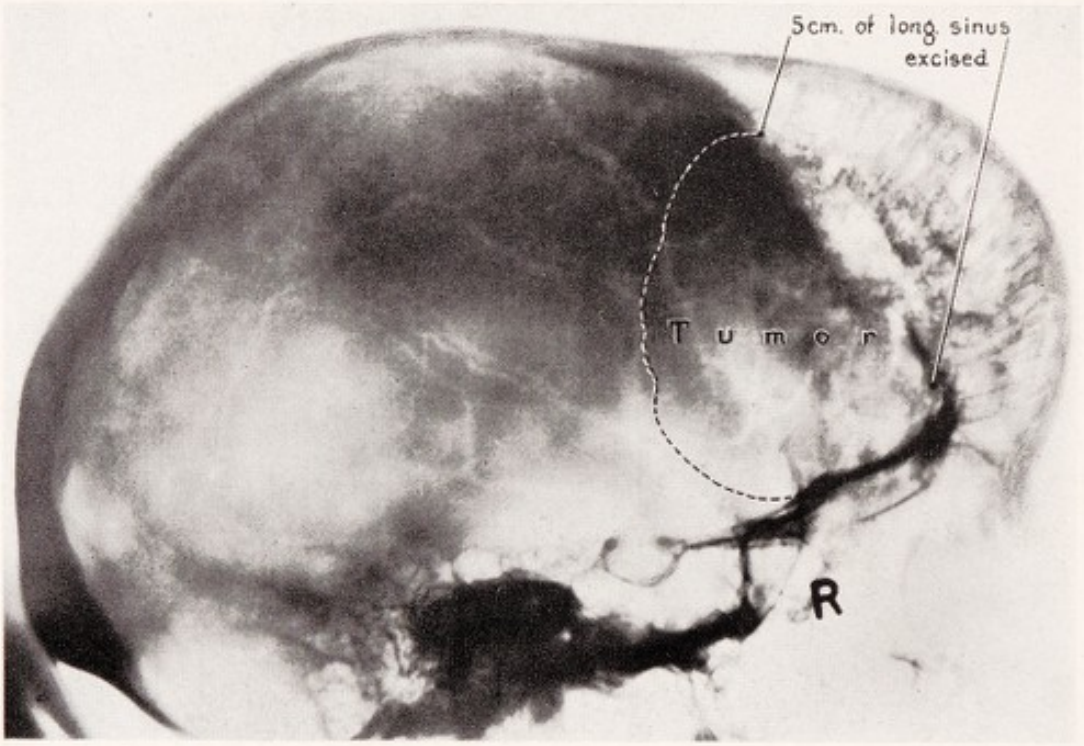


FIG. 65. Case 18. Lateral X-ray of skull, showing tremendous hyperostosis in frontal region.

this area did not pulsate. The blood pressure was 135/80 and the Wassermann reaction negative.

The X-rays showed an area of erosion in the frontal region where palpation revealed the defect. Everywhere the bone was much thicker and more dense than normal (Fig. 65). There was also destruction of the roof of the orbit on the right (Fig. 66).

Clinical Diagnosis: Osteosarcoma.

Operation, November 21, 1936: A long sweeping curved incision was made across the frontal region just under the hairline; it was directed posteriorly and extended from one temporal region to the other (Fig. 67). The skin and galea were then dissected from the bone and thrown forward to the orbital ridges. This step was very bloody; the bleeding was controlled by cauterizing the big vessels and then packing, with wet cotton, the surface of the bone and the

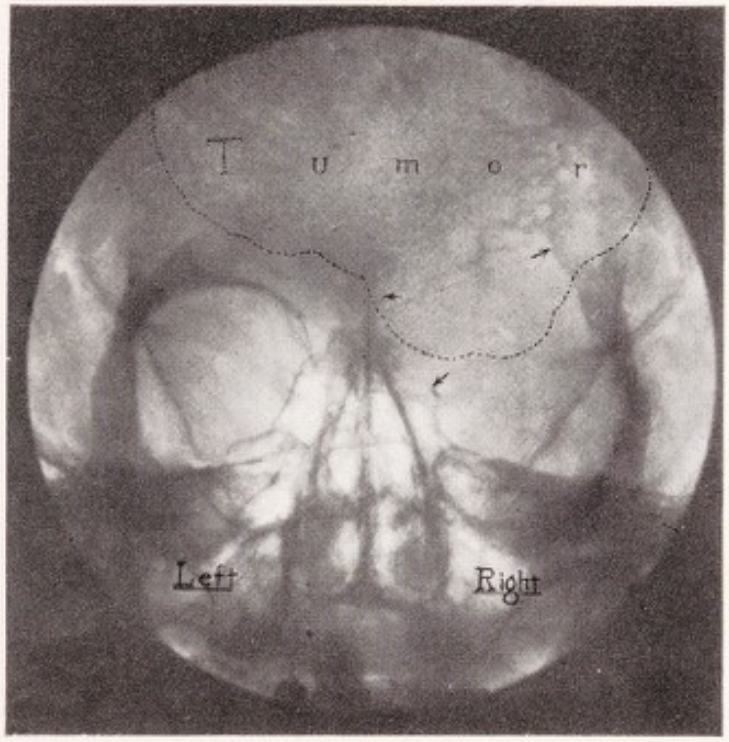


FIG. 66. Case 18. Anteroposterior X-ray, showing destruction of roof of orbit and supraorbital ridge by tumor.

tumor where it had ruptured through the bone. Several perforator openings were made in the bone along the curve outlined by the incision, where the swelling had, to outward appearances, practically disappeared. Everywhere the bone was extremely thick, in places more than an inch, so that it was necessary to use a very large burr; the bone between the openings had to be cut with the DeVilbiss and large bone-cutting forceps. On the right side, the tumor

had eroded through (or nearly through) the entire bone over a fairly large area. The base of the flap along the orbital margin was very long but very weak and it could be broken by turning the entire bony mass forward; it broke at the

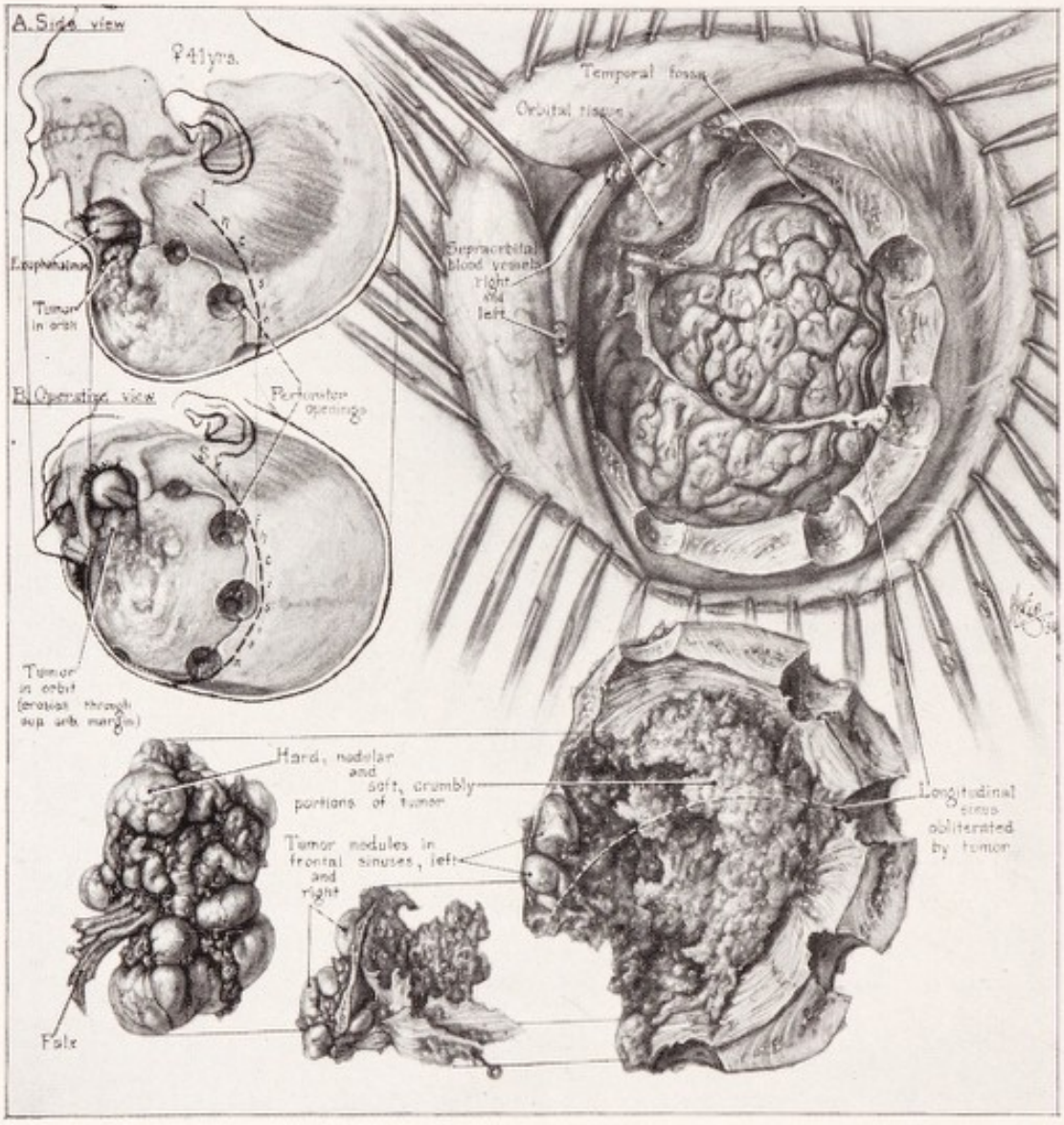


FIG. 67. Case 18. Operative sketch showing removal of bone in both regions, and underlying bilateral tumors, which are dural-type sarcoma. Anterior part of falx and of longitudinal sinus were removed with tumor. There was no sign of recurrence almost 3 years later.

orbital ridge (Fig. 67). Elevation of this huge flap was attended by very little bleeding. The dura, surrounded by a large tumor, came out with the elevated bone, and the anterior portion of the longitudinal sinus was included.

The sinus, however, was so compressed by tumor that very little bleeding resulted. When it was picked up, a small stream of blood was still coursing from the central end. A very large mass of hard fibrous tumor filled the frontal region on both sides; it lifted with the bone flap, to which it was densely bound. It was a white, nodular growth and projected deeply intracranially between the hemispheres and spread out over these. The mass of tumor was about as large as a fist. It also lay upon the olfactory groove; thus

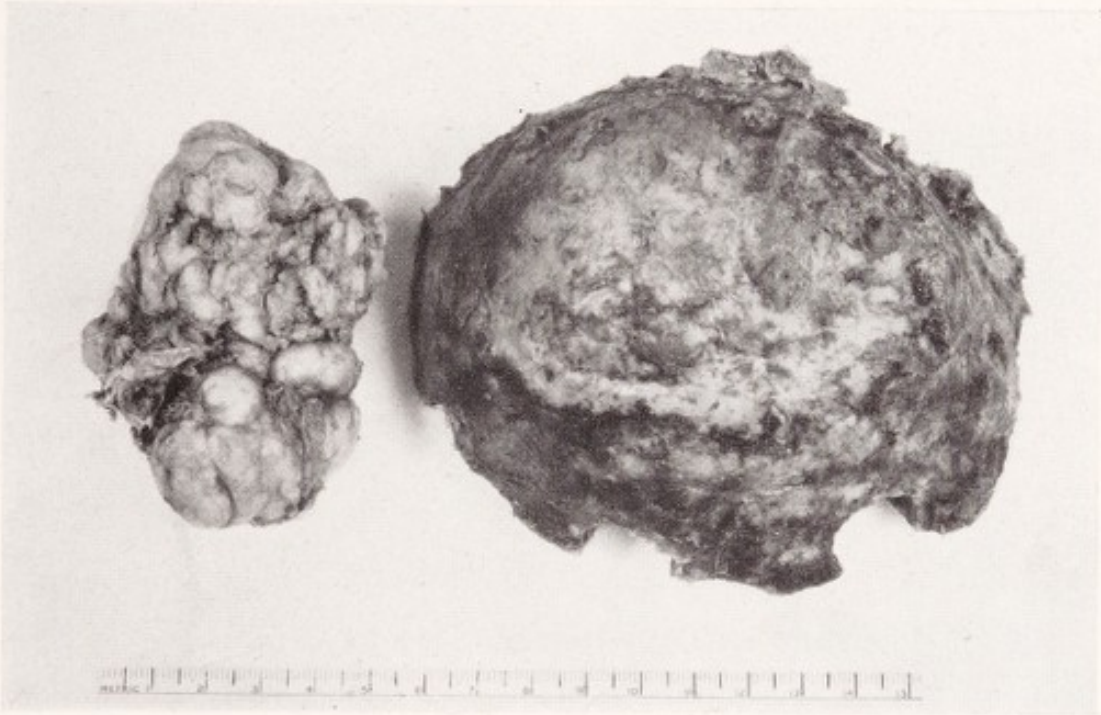


FIG. 68. Case 18. Removed tumor (left) and frontal bone (right).

both frontal lobes were squeezed between the mass of tumor over and beneath them. In some places the outskirts of the tumor were much softer, suggesting a more rapid development, i.e., malignancy. A portion of the tumor projected into the right orbit, the roof having been entirely destroyed (Fig. 66). It lay directly upon the capsule of the eyeball, and was shelled from it. The frontal sinuses had been obliterated by the tumor, as was shown in the X-ray. However, there was one small opening into the ethmoidal cell

far on the left side and from this a mucoid material escaped. This was the only point at which the paranasal sinuses were seen. The veins on the surface of the brain running to the portion of the longitudinal sinus that had been removed with the tumor were thrombosed. The entire bilateral frontal bones and attached tumor were removed. The entire mass weighed 269 grams [Fig. 68].

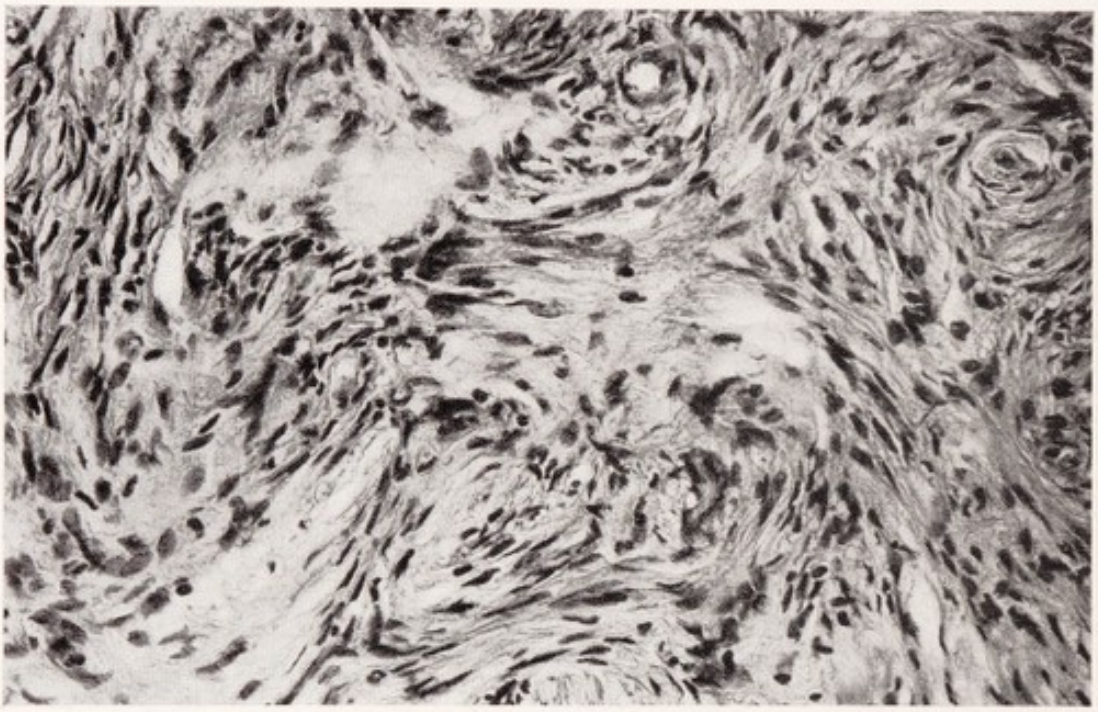


FIG. 69. Case 18. Photomicrograph of tumor, showing character and arrangement of meningioma cells.

It was necessary to give the patient a transfusion of blood at the end of the operation and two additional ones during the night.

Recovery was uneventful, the patient leaving the hospital twenty-three days later.

Microscopic Report: "Dural endothelioma, bilateral. The tumor is seemingly of the benign type with whorls and layers of fibrous tissue [Fig. 69]. It does not have the cellular element of the malignant dural tumors. There are some psammoma bodies."

Subsequent Course: I saw the patient February 5, 1938. She was perfectly well in every way and had been working steadily and at full capacity. She had had no headaches or double vision. There was no discernible exophthalmos, but the exophthalmometer recorded a proptosis of 2 mm. Visual fields were normal and visual acuity 20/30 in each eye. There was a marked depression in the frontal region, but this was masked by hair.

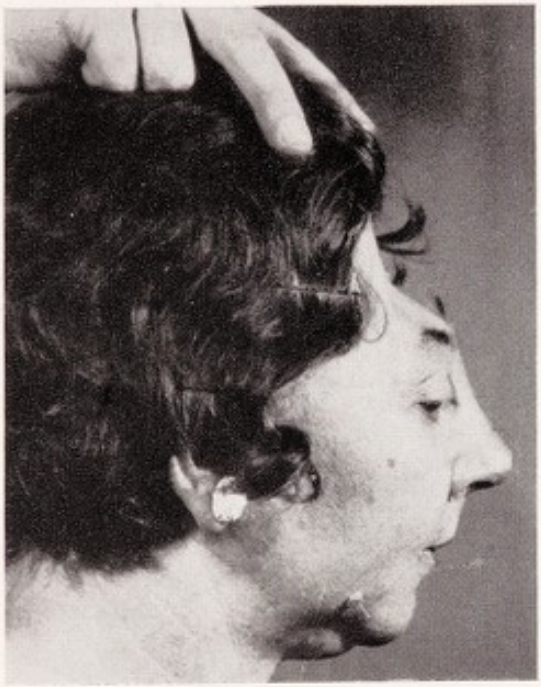


FIG. 70A

FIG. 70B

FIG. 70. Case 18. Fifteen months after operation. (A) Lateral view: big frontal defect is visible. (B) Frontal view: defect is quite well concealed by displacement of hair. Recession of eyeball is practically complete.

The patient was again seen in August, 1939, two and three-quarter years after the operation. She was perfectly well except for a pain in the right supraorbital region. The supraorbital nerve was sectioned for relief of this. The patient had been at work constantly since her discharge from the hospital. There was still the same marked frontal depression and there were no signs of local recurrence (Fig. 70).

CASE 19

R. H., History number U-103341. White, female, age 35. Admitted: April 7, 1937. Discharged: April 21, 1937.

Diagnosis: Dural meningioma with hyperostosis of skull and orbital roof.

Complaint: Slowly progressive right-sided exophthalmos of sixteen months' duration. There had been slight supra-orbital pains and headache, but no visual disturbance.

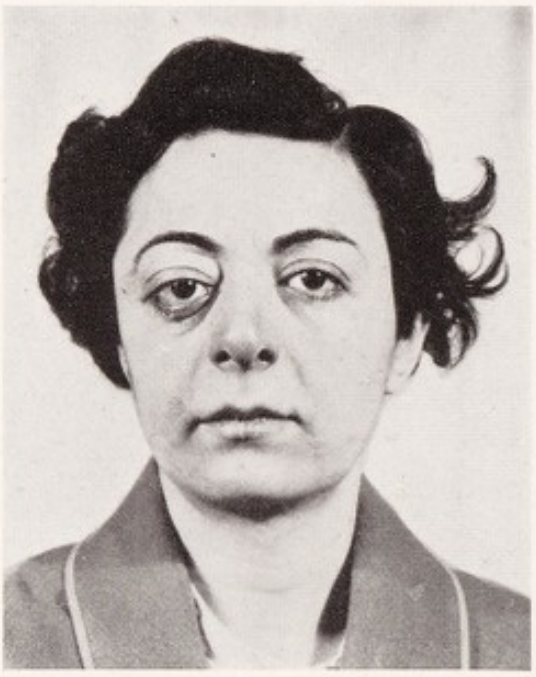


FIG. 71. Case 19. Marked grade of exophthalmos from hyperostosis in roof of orbit, this in turn due to intracranial meningioma.

Physical and Neurological Examinations: Negative except for a marked degree of proptosis (10 mm.) in the right eye (Fig. 71). The vision in the right eye was 20/30, in the left 20/50; the visual fields were normal. The Wassermann reaction was negative. X-rays showed a large diffuse dense shadow of the roof and to a lesser degree of all the walls of the orbit (Fig. 72). The orbital foramen was slightly smaller than on the opposite side (Fig. 73).

Operation, April 9, 1937: The usual approach was made.

The frontal bone was so thick that the supraorbital incision could not be continued laterally with the DeVilbiss forceps. It was necessary to break the bone flap across the osteoma. This thickening was a continuation of the osteoma which involved the roof and outer wall of the orbit; in the frontal

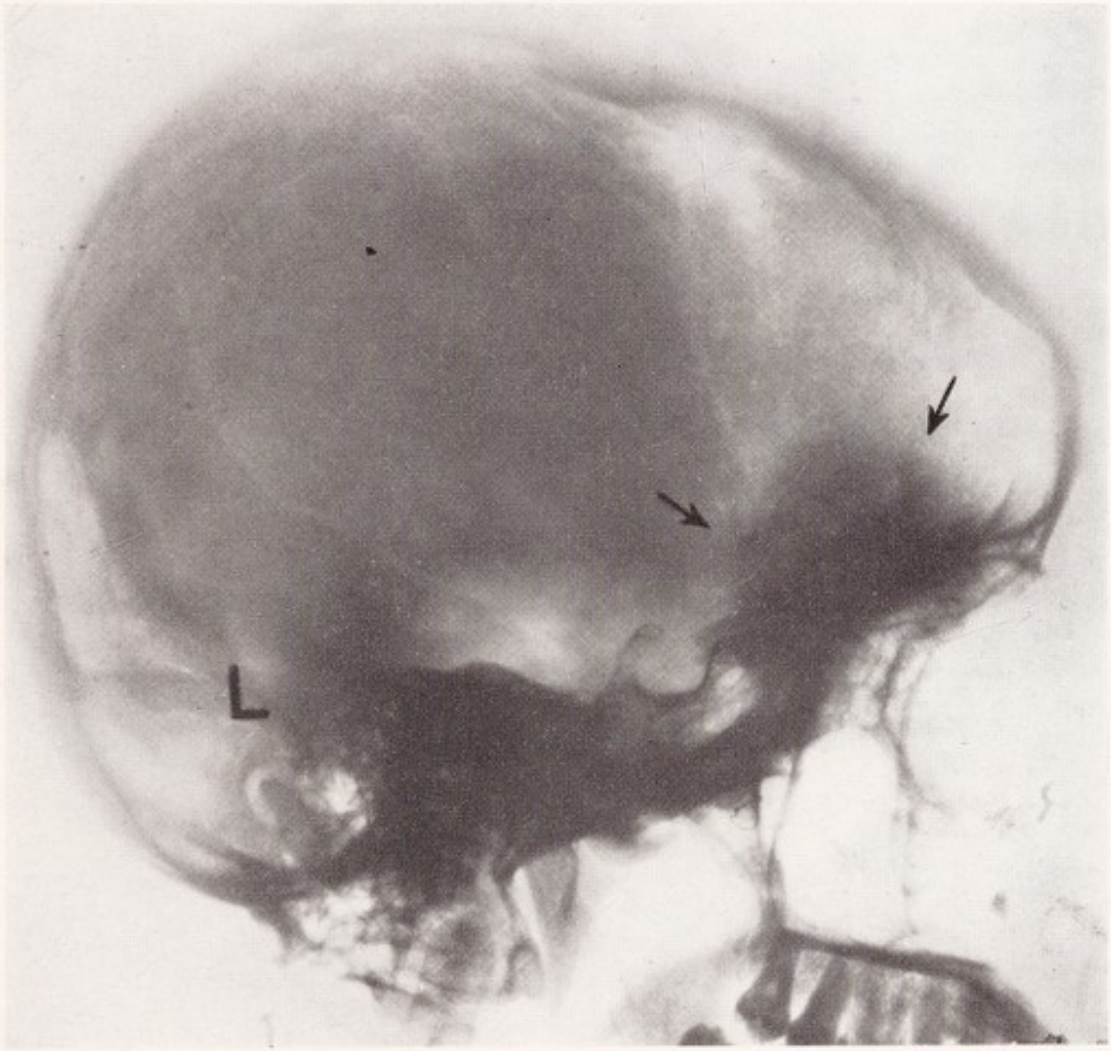


FIG. 72. Case 19. X-ray showing marked hyperostosis of skull and roof of orbit.

bone it was quite well circumscribed laterally. A hazelnut-sized dural tumor projected intracranially from the middle of the orbital roof. It was excised. The roof of the orbit was so greatly thickened (about 2 cm.) that it was necessary to hammer quite severely with the chisel in order to make an

opening into the orbit (Fig. 74). This thickened orbit ended abruptly about $\frac{1}{2}$ cm. from the optic nerve and the posterior border of the lesser wing of the sphenoid was also not affected. Mesially the thickening extended into the cribriform plate, where it also ended abruptly. The orbital roof was removed to the wing of the sphenoid and mesially to the ethmoidal cells, which were entered; from this opening a cerebrospinal fistula immediately developed. Because of this the wound was reopened on the following day; the

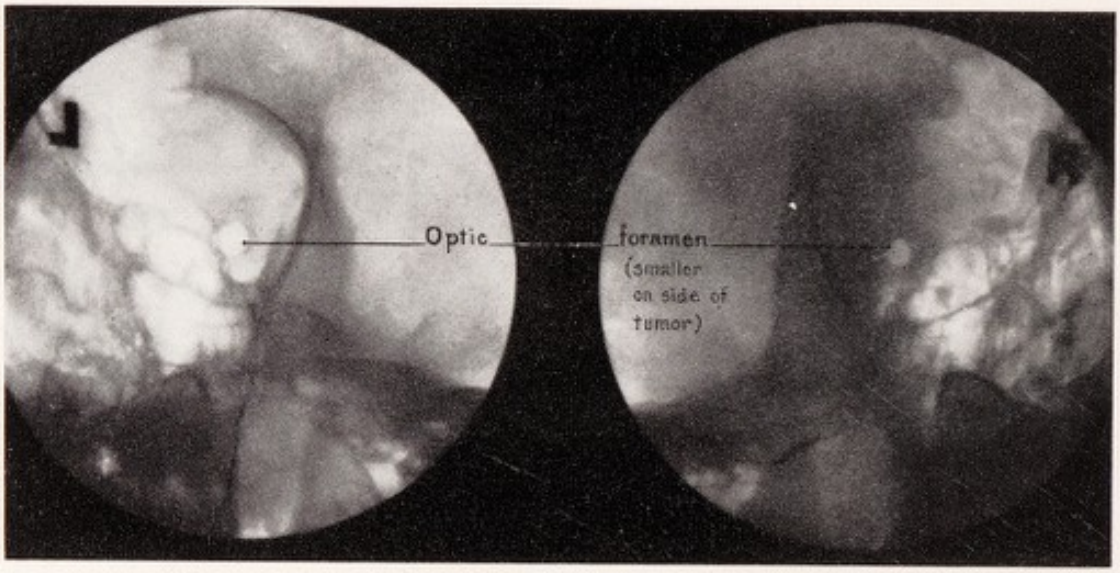


FIG. 73. Case 19. Retouched X-ray showing reduction in size of optic foramen on tumor side.

ethmoidal cells were covered with wax and the orbital capsule split and sutured over the open ethmoidal cells. A little iodine was placed along the undersurface of the brain in this region in order to facilitate the development of adhesions and make the closure of the opening permanent. There was no further leak of cerebrospinal fluid.

The patient's postoperative course was uneventful. She was discharged on April 21, 1937.

Ten months after the first operation (February 24, 1938) the patient returned for observation because of a steady increase in the protrusion of the eye and because the vision

was steadily diminishing; it was now seriously impaired. The headaches had continued and were about the same as before the operation. Her general health and weight had not changed.

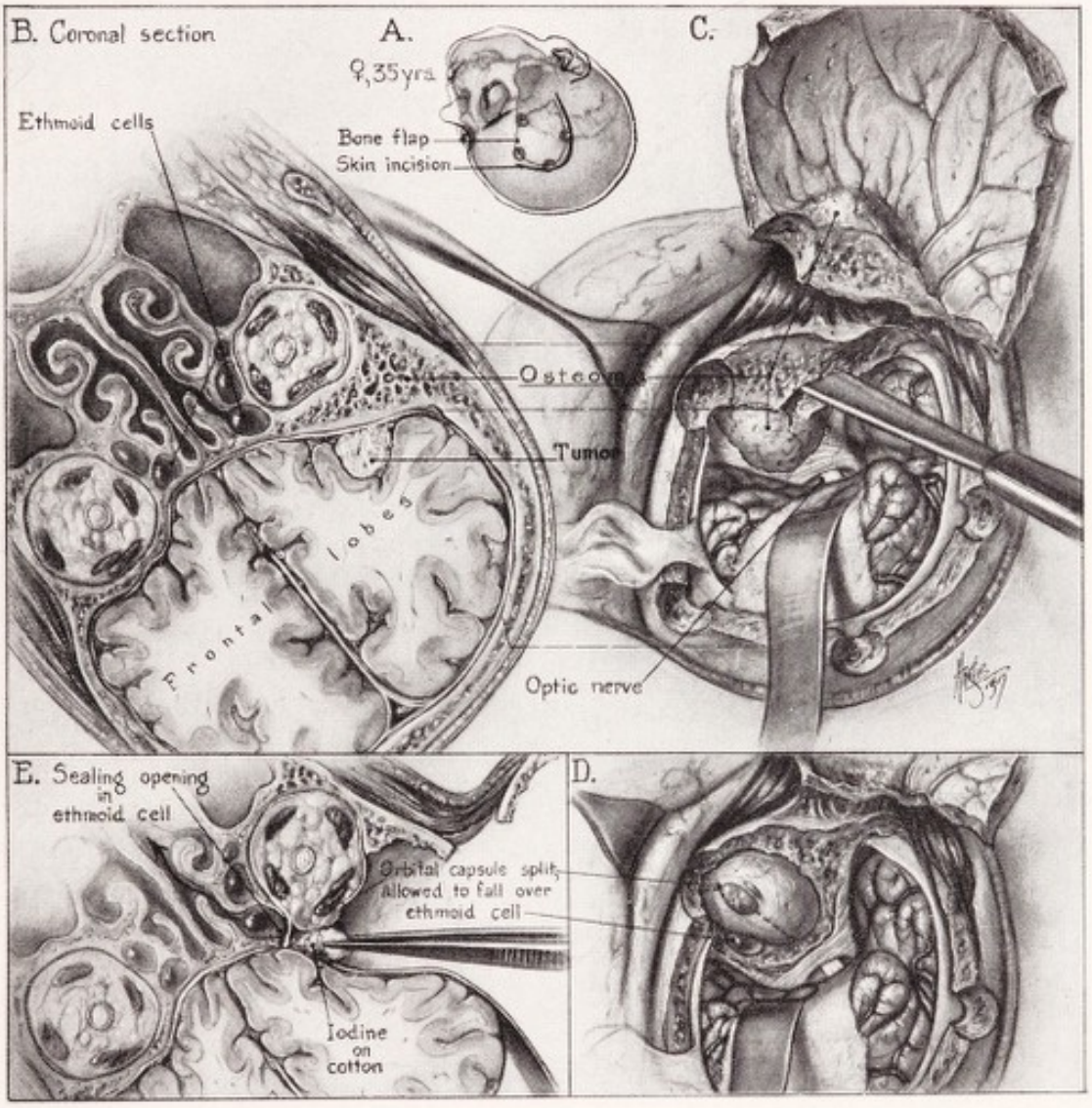


FIG. 74. Case 19. Operative sketch: method of chiseling away roof of orbit and hyperostosis of skull (C); ethmoidal cell opened (D); ethmoidal cell opened and covered with orbital fascia (E).

Dr. Woods's examination showed the protrusion of the eyeball to be 31 mm. on the right and 19 mm. on the left; this was 2 mm. more on the right than on her first admission, ten months before. The visual acuity was now 20/200 in the affected eye, whereas on the first admission it was 20/50.

There was no definite pallor of the nerve head; the visual fields for form and color were normal. However, there was a paracentral scotoma in the lower temporal field near the blind spot, and a scotoma for color running up from this to the point of fixation. On palpation through the upper lid there appeared to be a hard mass which could just be reached with the palpating finger; this, he thought, indicated a local

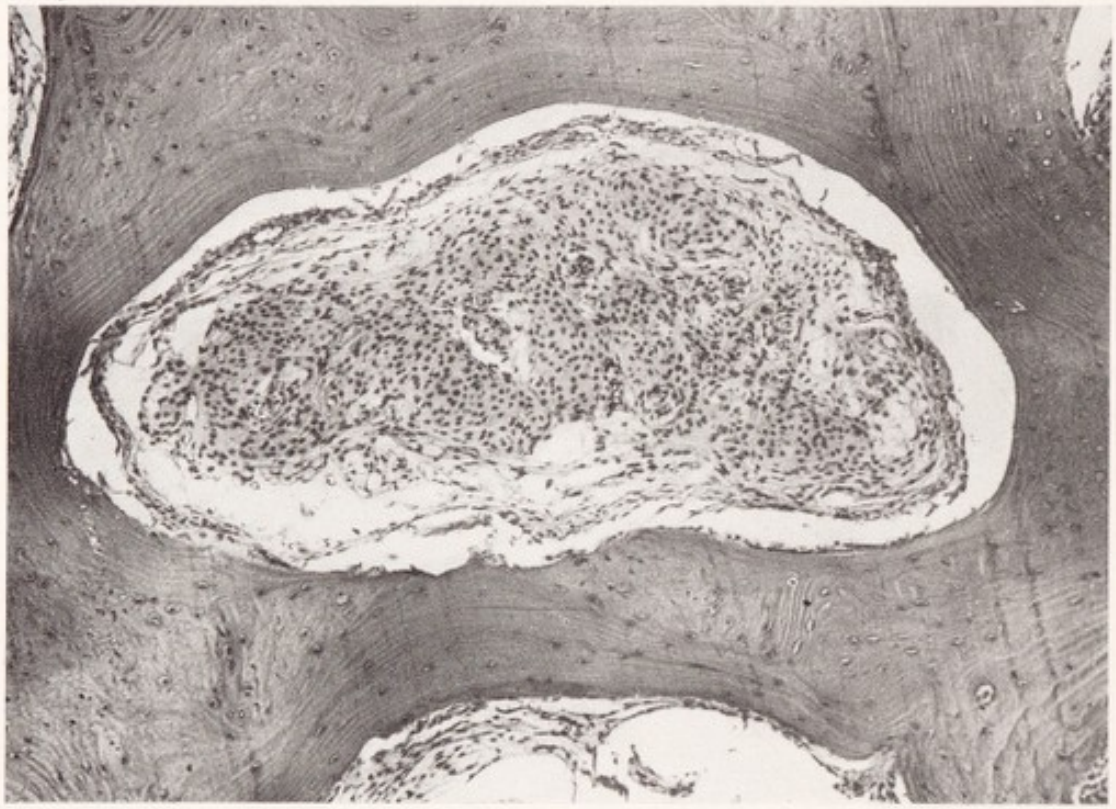


FIG. 75. Case 19. Photomicrograph of tumor, showing columns of cells invading marrow spaces of bone.

recurrence. However, the fact that the ethmoid sinuses were opened at the first operation led me to think that there might be a postinflammatory mass from this source.

Operation, February 24, 1938: The old incision was reopened and the bone flap re-elevated. The orbital defect was readily exposed. There was apparently no sign of tumor whatever, but there was a fair-sized mass of tissue which we took to be inflammatory granulation tissue. This covered the

whole region to the depth of from ½ cm. to a maximum of 1 cm. This was cut away until the covering of the eyeball was reached. There could not be, therefore, any sharply defined excision of this tissue. The superior rectus muscle was well exposed, but was not injured. Although this was taken to be inflammatory tissue, there was no evidence of an open paranasal sinus. Subsequent sections showed the tissue to contain invasive columns of cells of the malignant dural endothelioma type.

Microscopic Report: "The marrow spaces in the bone are filled with columns of cells of the malignant type [Fig. 75]. The dural tumor, both intracranially and intraorbitally, is of the very cellular type of dural endothelioma; the cells are large and closely packed, but in places there are the invasive columns. The tumor contains but little fibrous tissue."

Subsequent Course: The patient left the hospital on March 10, 1938, her condition essentially unchanged. She was last seen on June 15, 1939. The protrusion of the eye had steadily progressed and was now quite obtrusive. A bony swelling had appeared in the temporal region and was doubtless due to tumor.

CASE 20

H. H., History number U-107795. White, female, age 51. Admitted: May 31, 1937. Discharged: June 17, 1937.

Diagnosis: Dural meningioma with hyperostosis of orbital roof. (Referred by Dr. H. B. Gardner, of Pittsburgh, Pennsylvania.)

Complaint: Protrusion of the left eye.

Present Illness: The prominence of the left eye was first noted three and one-half years before; it gradually increased (Fig. 76). For three years there had been dull, bifrontal headaches, not more pronounced on one side than on the

other. For the past four months there had been some pain, not severe, behind the left eye. Vision had not been disturbed and had never been double.

Physical and Neurological Examinations: Negative except for the exophthalmos; the proptosis measured 5 mm. X-rays showed a marked diffuse thickening of the entire posterior part of the orbital roof (Fig. 77).

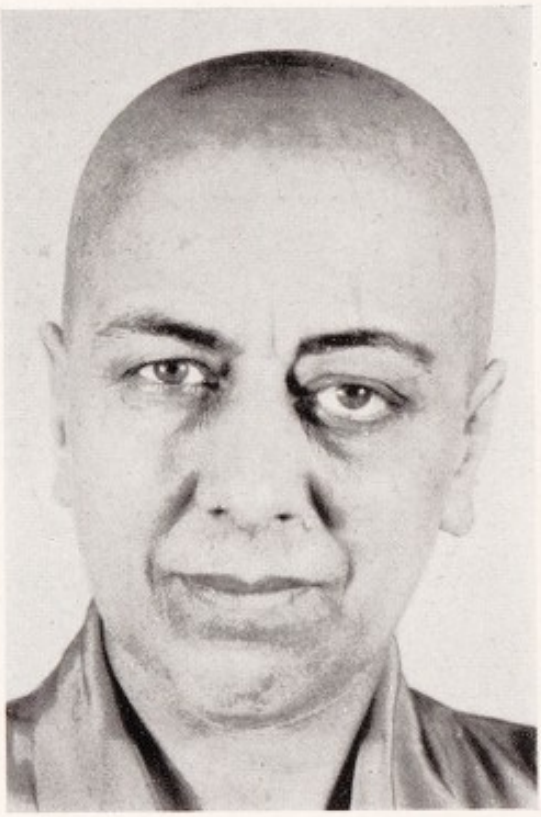


FIG. 76. Case 20. Preoperative photograph, showing mild grade of exophthalmos in left eye.

Diagnosis from X-ray findings: Osteoma.

Operation, June 2, 1937: The usual hypophyseal approach was made on the left side, using the concealed incision. The brain remained quite full during the operation because very little fluid could be obtained from the cisterna chiasmatis, but there was ample room.

The dura was stripped from the orbit, which was markedly

convex. The posterior half of the orbital bone was very reddish and vascular, in contrast to the white, unaffected anterior part; the osteoma was concentrated posteriorly. The orbital roof was first chiseled away anteriorly, where it was less affected, but even here it was so thick that it required much chiseling to make the initial defect. The opening was then enlarged by rongeurs, first toward the front of the orbit and then backward, but it was not long before the

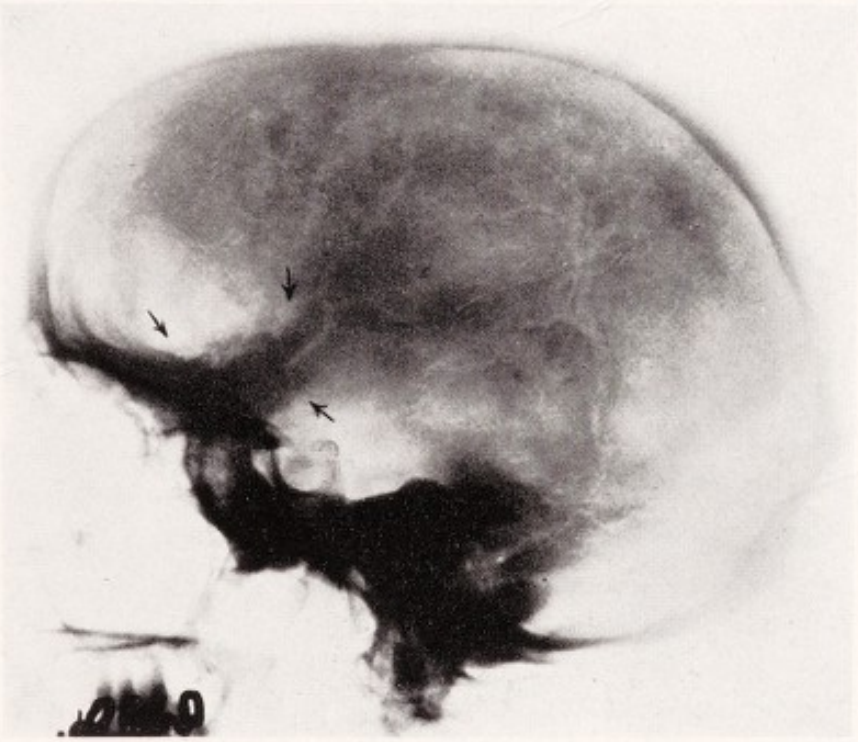


FIG. 77. Case 20. Lateral X-ray, showing degree of hyperostosis associated with intracranial meningioma.

bone became so thick (Fig. 78) that rongeurs were ineffective. It was then necessary to chisel away the bone and because of its great density and thickness much hammering was required. The whole lesser wing of the sphenoid was chiseled away in this manner. The thickened bone extended far out into the lateral wall of the skull, which was several times its normal thickness; this, too, was chiseled from the inside. When the lesser wing of the sphenoid was removed,

a small dural tumor (dural endothelioma) was found beneath it, lying in the middle fossa (Fig. 78). It was not larger than a hazelnut and was easily removed. The capsule of the orbit was not opened.

The brain was a little too tight to permit closure of the dura safely; a small part of the outer side of the frontal lobe was therefore removed. The weight of the dural tumor was 5 grams.

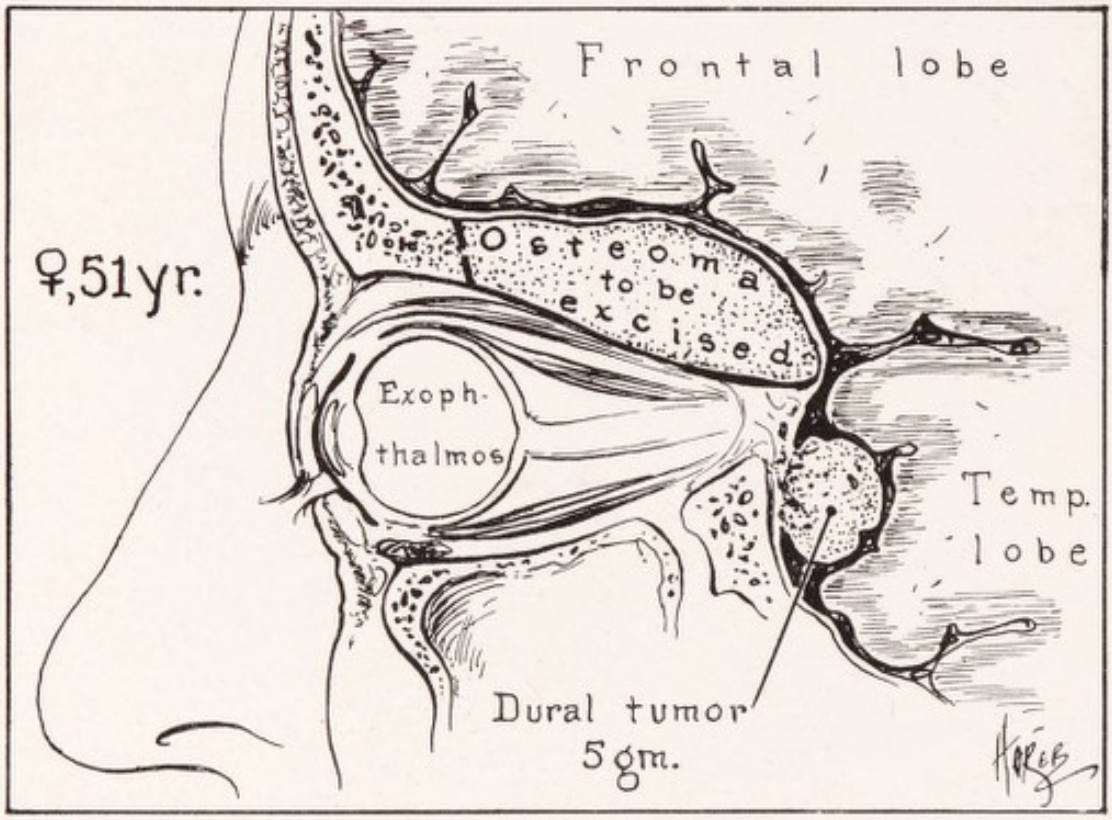


FIG. 78. Case 20. Sketch showing dural tumor.

During the postoperative period in the hospital, the patient's mentality was affected; she was euphoric, forgetful, and disoriented. Gradually these disturbances improved; they disappeared soon after the patient left the hospital.

Microscopic Report: "Small columns of cells of malignant dural endothelioma type invade the marrow spaces in the thickened bone [Fig. 79]. The dural endothelioma is of the cellular type."

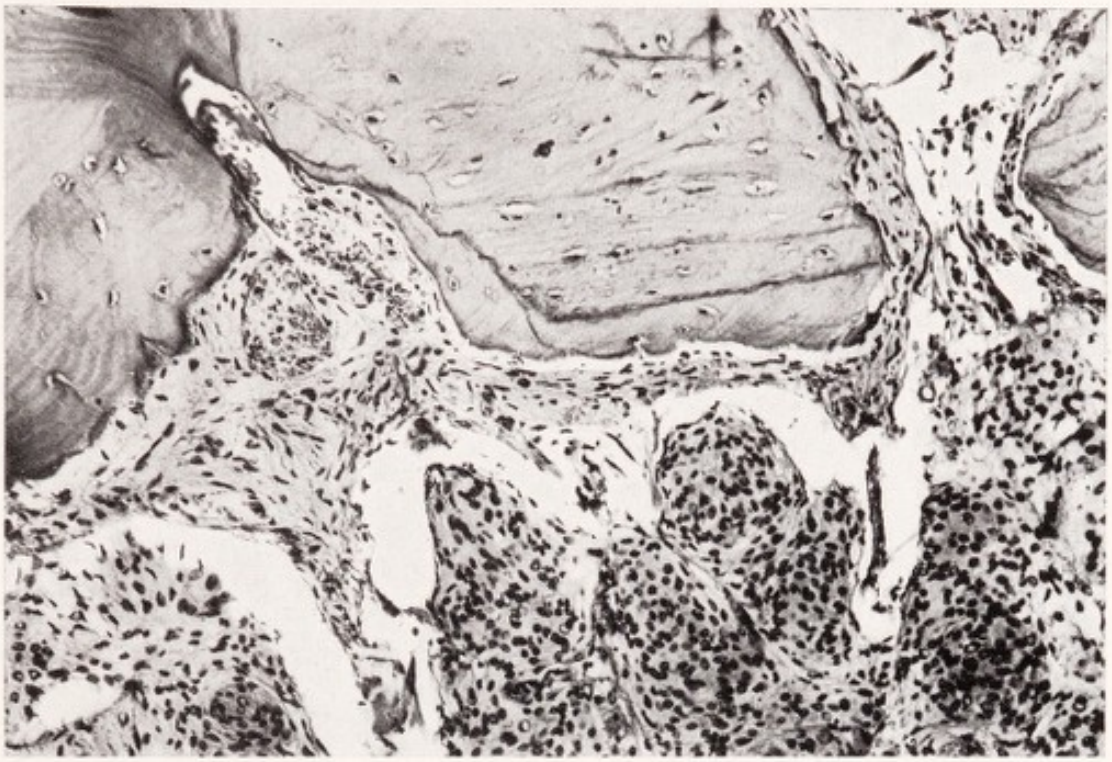


FIG. 79. Case 20. Photomicrograph showing invasion of marrow spaces of bone by numerous dural meningioma cells.

FIG. 80. Case 20. Two and one-quarter years after removal of orbital tumor.

Subsequent Course: A letter received June 20, 1939, two years after the operation, said: "Am feeling normal. My eyes feel all right. The eye is not quite in its normal position and I have some trouble from tears" (Fig. 80).

CASE 21

A. P., History number U-135345. White, female, age 48. Admitted: March 29, 1938. Discharged: May 5, 1938.

Diagnosis: Dural meningioma with hyperostosis of the orbital roof. (Referred by Dr. B. W. Lowry, of Tampa, Florida, with the diagnosis of an orbital tumor.)

Complaint: Protrusion of the right eye.

Family and Past Histories: Negative.

Present Illness: Four years ago, following an attack of influenza, the patient had noticed a fulness of the right eye. Since it was painless and barely noticeable she did not consult a physician (Fig. 81). Until nine months ago no one except members of the family had apparently noticed that the eye was abnormal. The growth had been very gradual and at no time had there been pain, diminution of vision, or diplopia.

An X-ray had recently been taken by her physician and an osteoma at the roof of the orbit was disclosed (Figs. 82A and B). At this time a definite unilateral exophthalmos was noted and the diagnosis of orbital tumor was made.

Physical and Neurological Examinations: Negative, except for the local condition. This was examined by Dr. Woods, who noted the following positive and negative findings: Exophthalmos of 6 mm. on the right; no weakness of the extraocular muscle; no disturbance of the pupillary reaction; no increased tension of the eyeball. The visual fields were normal for form and color. Visual acuity was 20/15 in each eye.

An X-ray showed a marked increased density in the roof of the orbit. The optic foramina were normal in size and equal.

Operation, April 2, 1938: A hypophyseal approach was made on the right side. There was no increased intracranial pressure. When the frontal lobe was elevated, a small, hard dural endothelioma (Fig. 83), almost spherical and about

FIG. 81. Case 21. Mild grade of exophthalmos in right eye, caused by intracranial meningioma and hyperostosis.

2 cm. in diameter, presented on the roof of the orbit. It was about 1 cm. from the lateral margin of the skull and about the same distance from the anterior margin of the skull. When the dura was stripped from the roof of the orbit this tumor came off without attachment to the bone, but the dura remained fast on the orbital surface of the tumor. The roof of the orbit was of fairly normal contour, except that it was definitely elevated. With the chisel the roof of the

orbit was entered anteriorly and gradually the osteoma was removed piecemeal (Fig. 83). The osteoma ended mesially just before reaching the paranasal sinuses, and posteriorly just in front of the lesser wing of the sphenoid. It therefore did not involve the lesser wing of the sphenoid nor the region of the orbital foramen. It did, however, extend into the

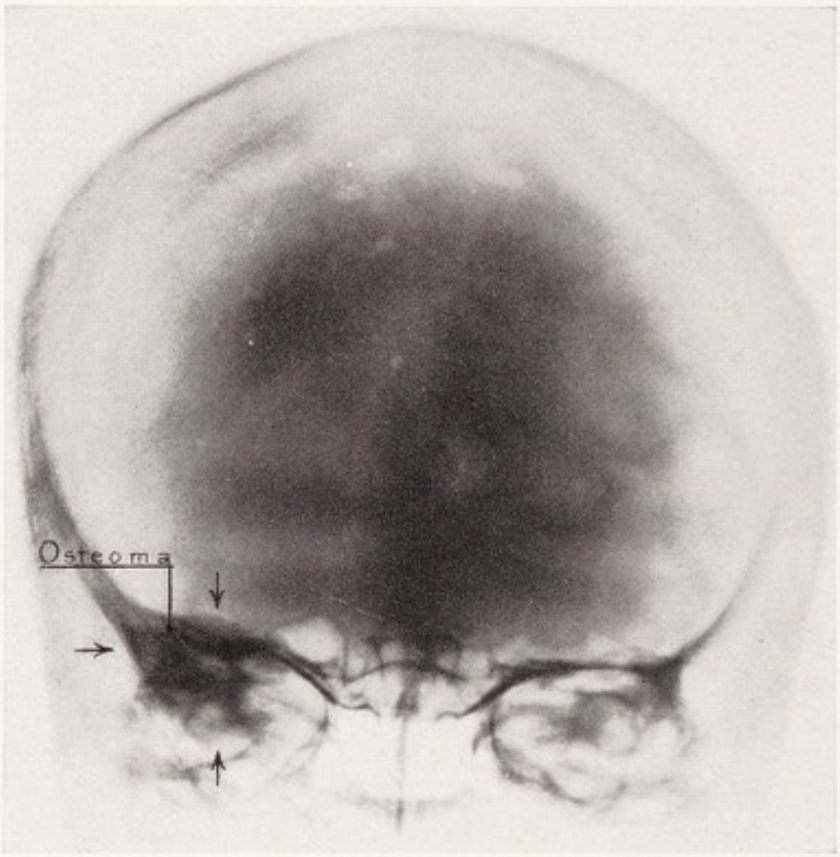


FIG. 82A. Case 21. Lateral view, showing degree of hyperostosis associated with dural meningioma.

lateral surface of the skull and this was removed completely. The bone was exceedingly dense and hard, but there was no sign of tumor within it, and it shelled out so well from the orbital content that, with the exception of one small area about 1 cm. in diameter, Glennon's capsule was smooth and glistening. This roughened, reddish-brown area was at the posterior mesial end of the bony defect and suggested the

possibility of tumor. This was cut away from the orbit and was the only point where the capsule was opened. This tissue had no connection whatever with the dural endothelioma, which was on the cranial side of the orbital roof and

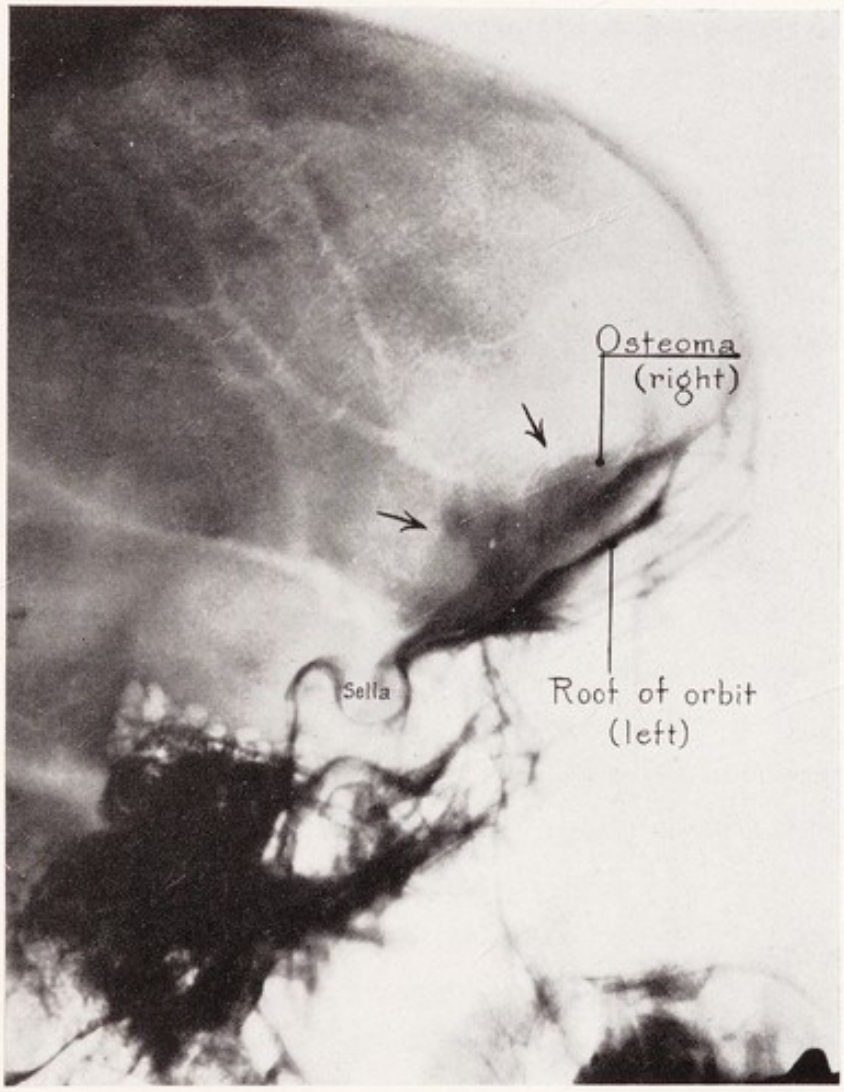


FIG. 82B. Case 21. Anteroposterior view.

far removed from it in the anteroposterior direction; this piece of tissue was removed for section. It was not an encapsulated or even a circumscribed mass; if it had been tumor it would have been infiltrating in character.

Postoperative Course: A wound infection developed two weeks after the operation and delayed the patient's departure

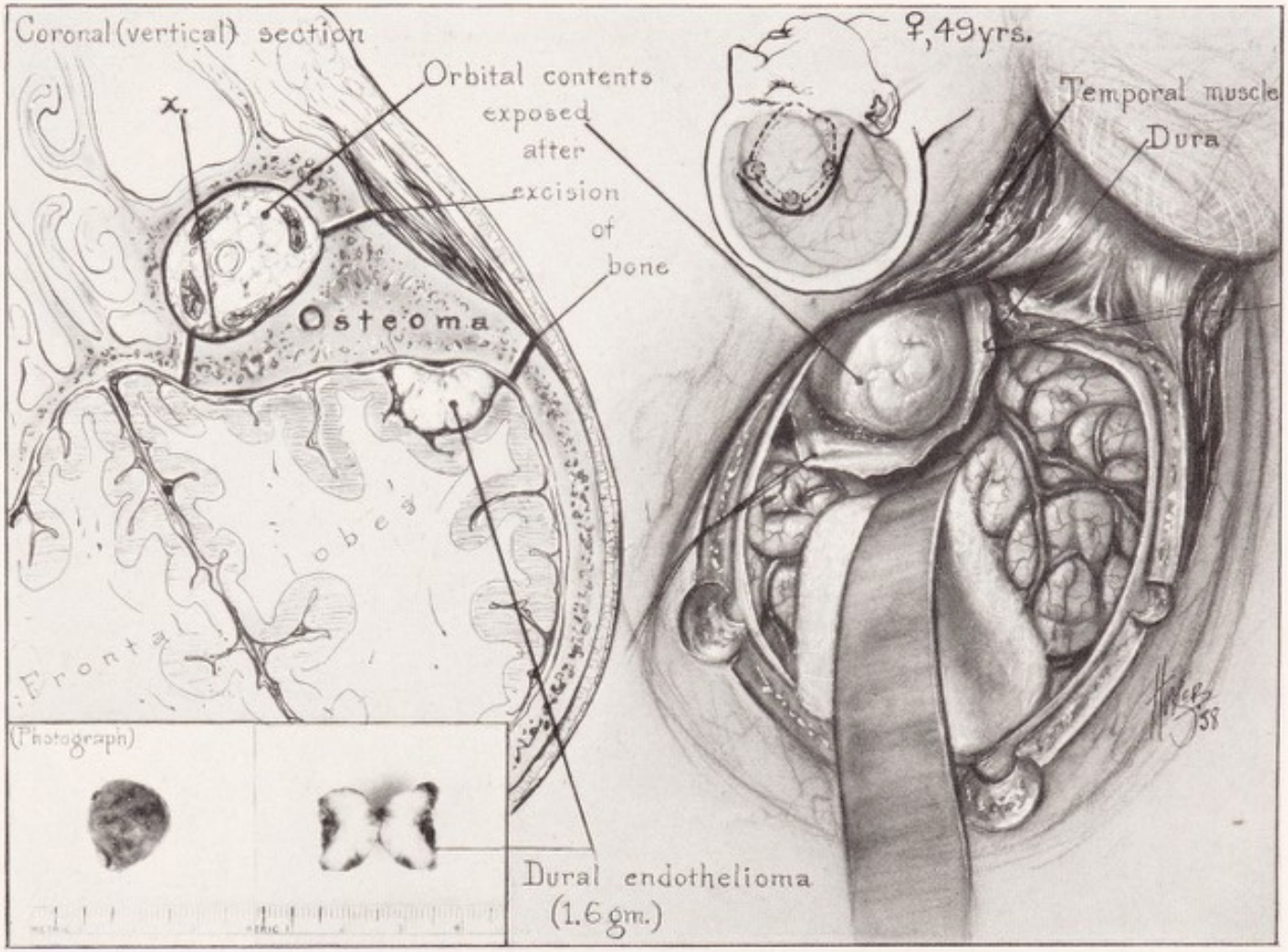


FIG. 83. Case 21. Operative sketch, showing relative position and size of intracranial meningioma and degree of thickening of lateral wall of skull and roof of orbit. Extent of bony removal is also shown.

until May 5. Her exophthalmos had entirely disappeared at that time.

Microscopic Report: The marrow spaces in the thickened bones were filled with strands of cells like those of a malignant form of dural endothelioma (Fig. 84). The dural tumor itself was one of cellular type and contained very little fibrous tissue—no whorls or lamellae of fibrous tissue. The

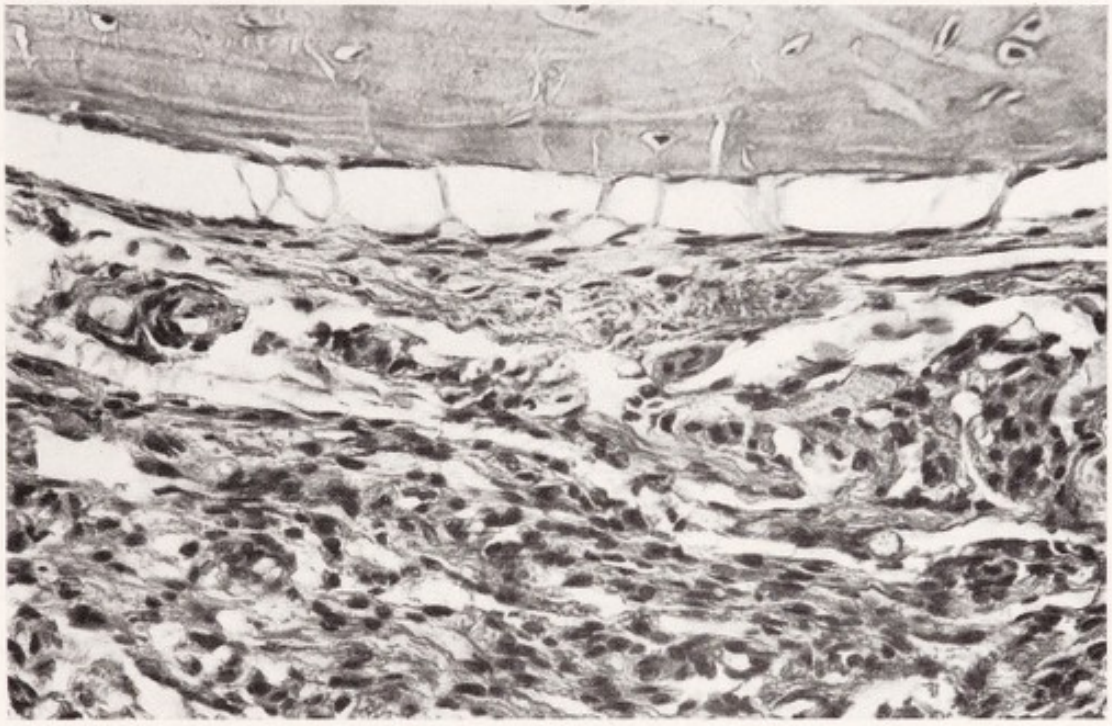


FIG. 84. Case 21. Photomicrograph of tumor, showing invasion of bone.

doubtful-looking tissue removed from the orbital capsule (it was located near the apex of the orbit and far removed from the intracranial dural endothelioma) proved to be tumor and of the same columns of cells that were observed in the marrow spaces.

Subsequent Course: A letter received from the patient, June 22, 1939, stated that she was in excellent health, that the wound had healed, and that the eye did not protrude.

CASE 22

M. S., History number U-143926. White, female, age 43. Admitted: July 29, 1938. Died: July 30, 1938.

Diagnosis: Dural meningioma with enormous hyperostosis of skull and orbital roof.

Complaint: Bone tumor in the skull.

Family and Past Histories: Negative.

Present Illness: Five years ago a small, hard mass had first been noticed in the right temple. This grew very slowly but progressively. After two years' growth it was estimated to be about the size of a hen's egg. At this time the right eye began to bulge, and this also had steadily progressed. Three years ago the mass in the temporal region had been attacked surgically, but little was accomplished and it was soon back to its former size. Although there had been no great discomfort, there was always a slight ache over the mass. There had been no noticeable loss of vision.

Physical and Neurological Examinations: Negative, except in the region involved by the tumor. Beginning at the right zygoma there was a diffuse, bulging, exceedingly hard bony mass measuring 6 cm. vertically, 4 cm. horizontally, and protruding perhaps 5 cm. The tumor was obviously continuous with the bone. Nowhere was it tender. The exophthalmos in the right eye measured 6 mm. Tension in the eyeball was normal; vision was 20/15 on each side. The visual fields were normal. No murmurs were heard over the mass. The Wassermann reaction from the blood was negative; blood pressure was 140/85.

The X-rays showed tremendous thickening of the roof of the orbit and the sphenoidal wing. Thickening through the temporal region was extreme; it passed imperceptibly into the surrounding frontal and parietal bones (Figs. 85A and B). The hyperostosis was much greater than in any of the other cases of this series.

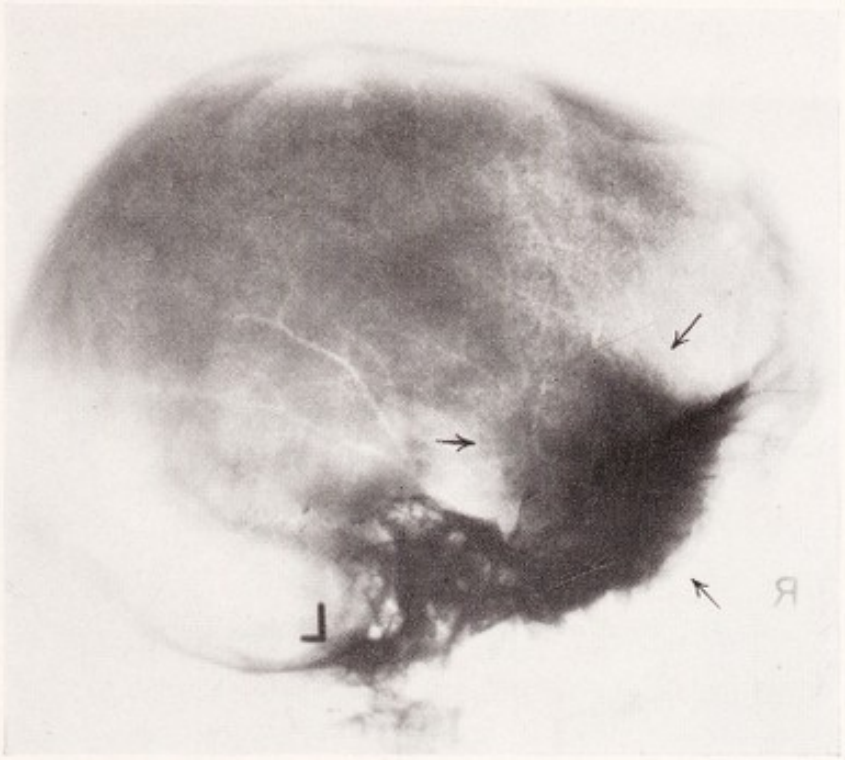


FIG. 85A

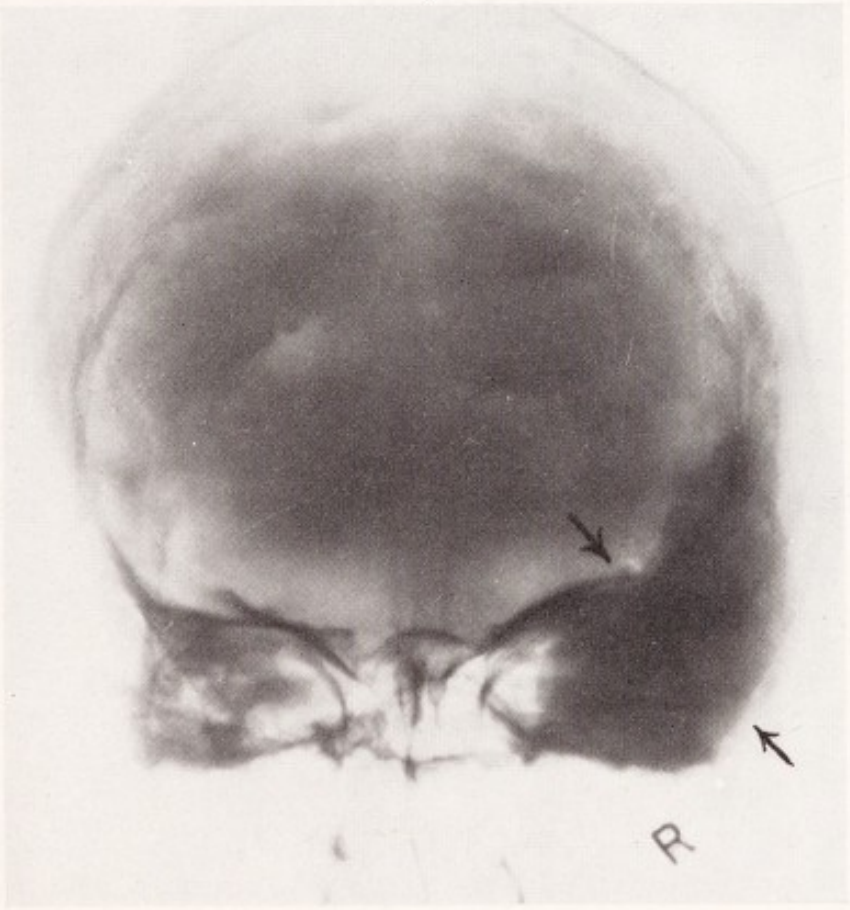


FIG. 85B

FIG. 85. Case 22. X-rays of skull showing location and degree of hyperostosis: (A) lateral view, (B) anteroposterior view.

Operation, July 30, 1938: A hypophyseal flap was made on the right side. The bone along the supraorbital ridge soon became too thick for the blade of the DeVilbiss forceps. The bone flap was then broken very much farther mesially than desirable. From that point on, laterally into the temporal fossa and into the anterior fossa, the bone was so enormously thickened and so dense, that one could make little impression upon it with the chisel and hammer without causing undue trauma to the head. It would have been a serious ordeal to have gradually cut away the bone from such an extensive region. We then hoped to cut the bone around its periphery in all accessible regions and gradually leave as narrow an intracranial portion as could be broken across like a bone flap. Accordingly, an incision was made across the mesial side of the supraorbital ridge and carried backward through the orbital plate and through the sphenoidal ridge just mesial to the optic nerve. A second incision was made through the lower part of the lateral margin of the orbit and carried downward and backward toward the temporal region; the zygoma was also divided. A third incision was made posterior to the mass of tumor in the temporal bones and carried some distance toward the base of the skull. The temporal and masseter muscles were stripped from the skull and zygoma. With these three incisions converging toward the middle fossa, the thinner bone at the mesial part of the middle fossa was broken across and the tumor at the side of the skull lifted from its bed (Fig. 86). But the line of the break in the bone passed through the carotid canal and tore the internal carotid artery. A profuse hemorrhage was quickly controlled by packing and the artery soon picked up with forceps and "clipped" and thrombosed with the cautery. When the mass was removed the eyeball and optic nerves were intact. The

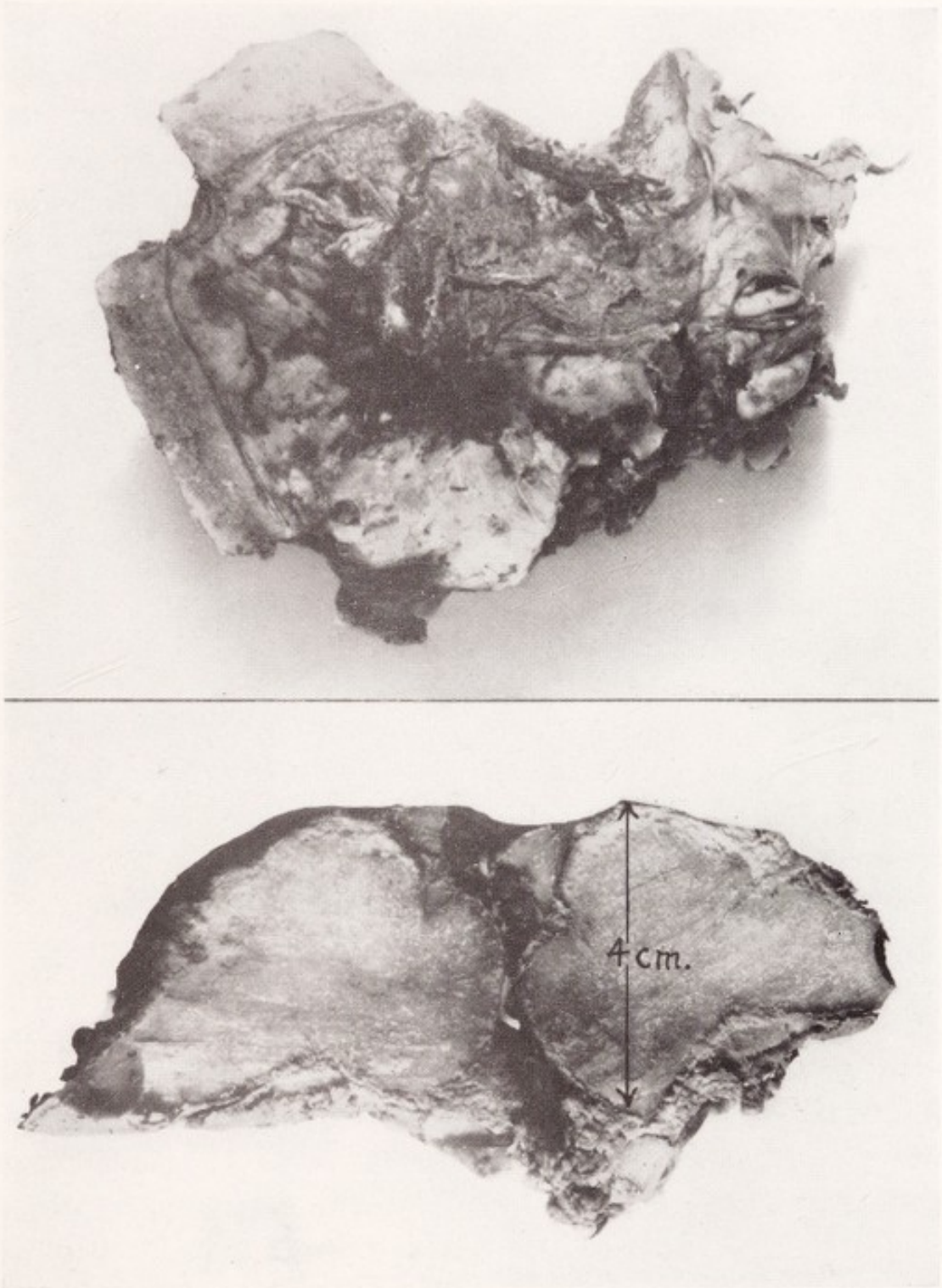


FIG. 86. Case 22. Surface view and cross-section of bony mass removed from orbital roof and lateral wall of skull. Tumor extended to carotid canal.

bone flap broke so far mesially that one could look into the middle fossa and see the hypophysis. On the inferior surface of the orbital mass was a thin layer of soft tissue, of scarcely more than the thickness of paper; this was a dural growth. There was a small irregular fibrous dural tumor projecting into the cranial chamber over the thickened roof of the orbit. The amount of soft tissue was very slight as compared with the

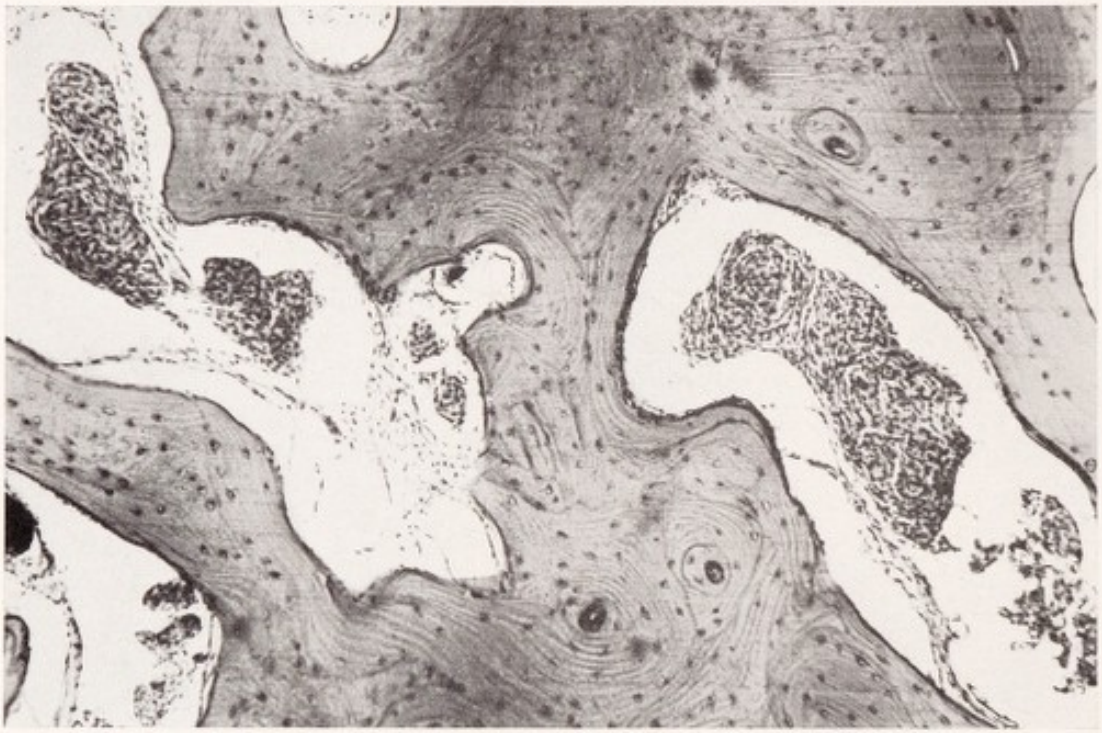


FIG. 87. Case 22. Photomicrograph showing invasion of Haversian canals of thickened orbital bone by meningioma cells.

vast amount of solid bone. It is worthy of note that the roof of the orbit was roughly spherical, curving downward into the orbital cavity and also upward into the cranial chamber. The mass in the middle fossa was roughly of the same shape, continuous with the orbital mass without a neck, and was perhaps three times the size of the latter.

The patient did not regain consciousness and died during the night despite transfusions of blood. There had been no further bleeding. The wound was clear at autopsy.

The specimen was exceedingly hard, dense, eburneous bone; it measured 8 x 7 x 5 cm., and weighed 145.3 grams (Fig. 86).

Microscopic Report: The section of the tumor within the cranial chamber and in the orbital bone showed columns of tumor cells of the typical meningioma type. The same tissue was seen in the medullary spaces of the thickened bone (Fig. 87). There were no psammoma bodies.

CASE 23

N. R., History number U-165565. White, female, age 46. Admitted: March 8, 1939. Discharged: March 31, 1939.

Diagnosis: Dural meningioma with hyperostosis of skull and orbital roof.

Complaint: Protrusion of the right eye.

Family and Past Histories: Negative.

Present Illness: A slight swelling of both the upper and lower lids of the right eye had begun four years ago. The swelling, however, developed so slowly that the patient paid no attention to the mounting edema until two years later when the eyeball itself began to protrude. There had never been any associated pain; the only discomfort had been lacrimation, with some conjunctival infection.

During the past year the exophthalmos had progressively increased and during this time there had been a slight but definite diminution in vision. In November, 1938, a surgeon removed a tumor from the right orbit. Whorls of fibrous tissue were said to be the dominant histological picture, i.e., doubtless a dural tumor. Immediately following this operation the eye was and had since been blind; there was also paralysis of the extraocular muscles. Since the operation there had been no pain or discomfort, but a tremendous edema of both upper and lower lids developed,

which had remained practically unchanged up to the time of the admission (Fig. 88). This edema was doubtless the result of the previous extensive incision around the orbit and was due to inadequate venous return.

Physical and Neurological Examinations: Negative except for the local condition. The upper and lower eyelids were

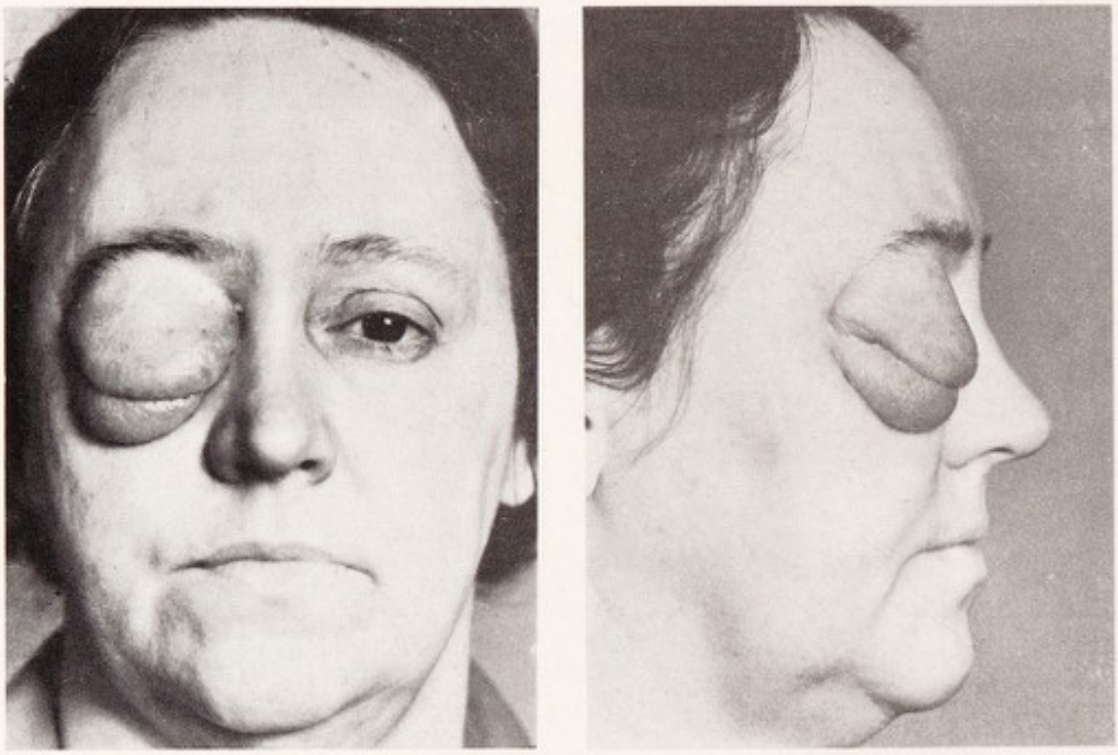


FIG. 88. Case 23. Front and lateral views. Tremendous edema of upper and lower lids due to a peripheral operation performed elsewhere. The swelling is doubtless due to impairment of blood and lymph supply to lids; its terrific size conceals the exophthalmos, which is of mild grade.

so greatly swollen (edema) that the protruding eyeball could not be seen; they protruded about equally and to a distance of $5\frac{1}{2}$ cm. There was practically no movement in the eyelids; this was due in large part to the tremendous edema. Nor were there any movements of the extraocular muscles, excepting a slight internal rotation. Vision was nil, the disc pale and glistening, i.e., primary atrophy. The veins and arteries were small and there was no tortuosity

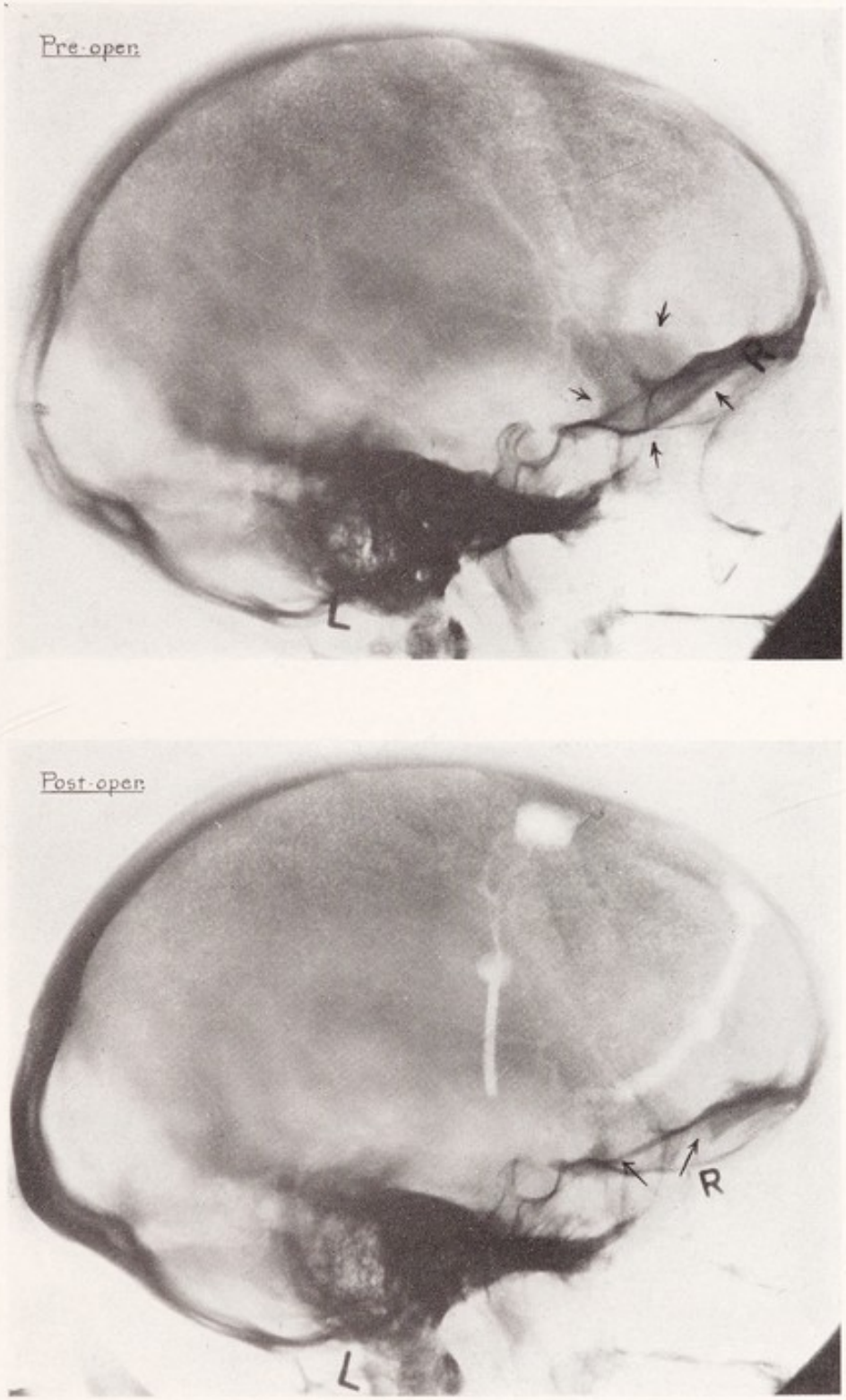


FIG. 89. Case 23. Lateral views of skull, showing degree of hyperostosis of skull and roof of orbit: (A) before operation, (B) after operation.

of the retinal veins; the eyeball protruded 9 mm. The vision and visual fields were normal.

X-rays showed a definite but moderate grade of hyperostosis of the roof of the orbit, extending laterally into the region of the pterion (Fig. 89). This was best shown in the anteroposterior view. The orbital foramen was smaller on the side of the tumor (Fig. 90).

The Wassermann reaction from the blood was negative; the blood pressure was 130/86.

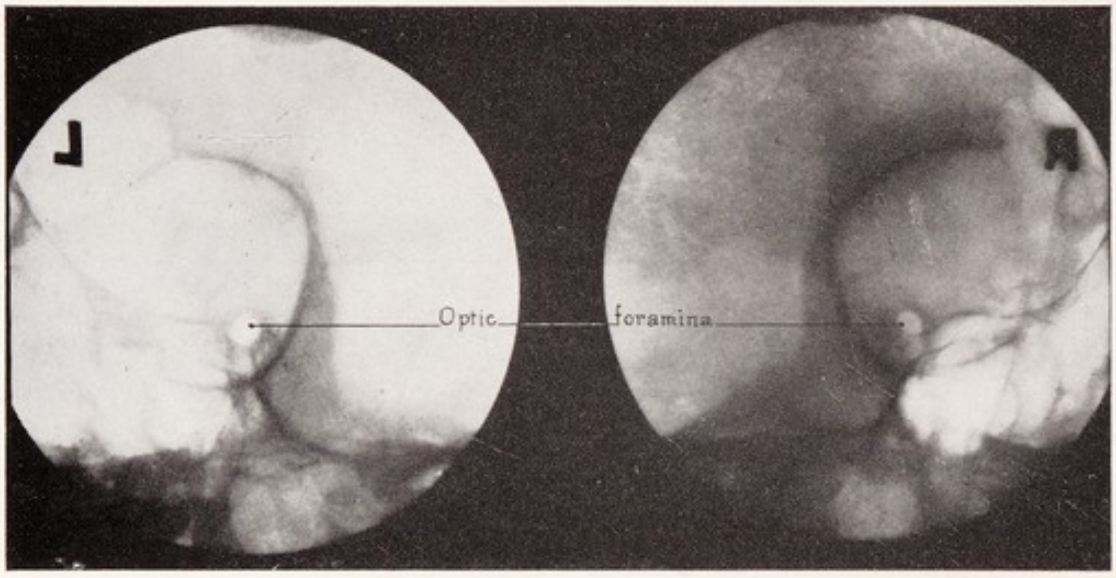


FIG. 90. Case 23. X-rays of orbital foramina, showing reduction in size of foramen on tumor side.

Operation, March 15, 1939: A hypophyseal approach was made on the right side, using the concealed incision. As soon as the bone flap was turned down the first evidence of tumor appeared along the lesser wing of the sphenoid, at which point the growth had penetrated the dura and the bone was thickened. When the dura was opened the dural tumor could be seen covering the roof of the orbit; it was about 1 cm. thick and spread over the entire floor of the anterior fossa. But it also extended posteriorly around the sphenoidal ridge into the middle fossa, covering like a

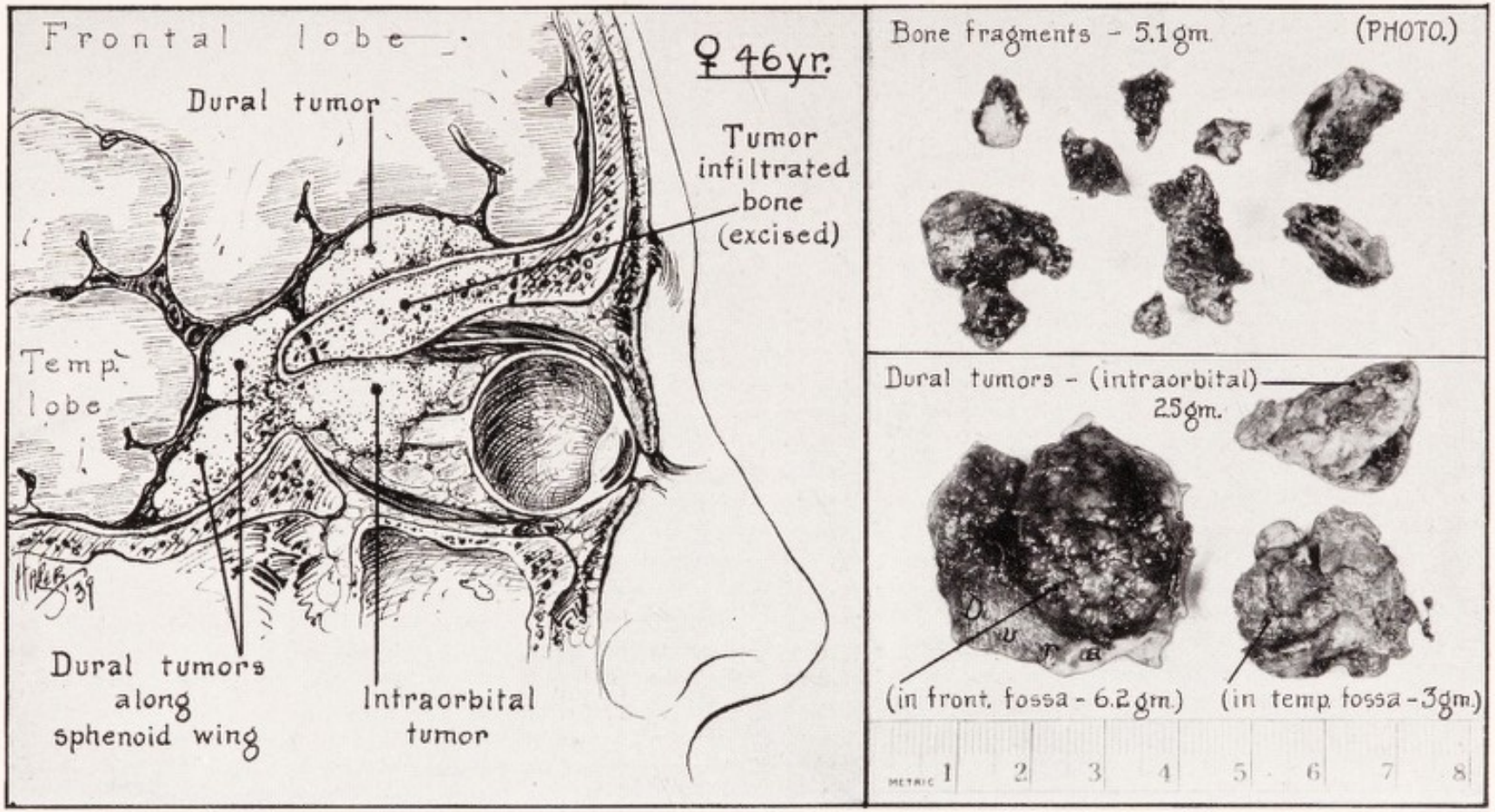


FIG. 91. Case 23. Sketch showing dural meningioma covering roof of orbit and extending back into middle cranial fossa. Photographic insets (right) show portions of orbital roof removed piecemeal, and the three divisions of tumor removed from orbit and cranial chamber.

carpet about two-thirds of the floor of the middle fossa (Fig. 91). It ended just posterior to the foramen spinosum and mesially at the Gasserian ganglion. The depth of the tumor was fairly uniform and essentially the same in both the middle and the anterior fossa. The tumor was very hard and firm. It was so hard and so tightly bound to the dura that no line of separation was possible. To remove the tumor it was necessary to strip the dura from the bone, and both were extirpated together. The tumor and dura were stripped from the roof of the orbit and the dura incised just beyond the tumor's margin, which was just short of the optic nerve. The portion in the middle fossa was much more difficult to remove because it was fed by the middle meningeal artery and was, therefore, exceedingly vascular. The roof of the orbit was then chiseled away; it was much thicker than we had reason to expect from the X-rays—about 1 cm.—and was fairly well distributed along the entire roof of the orbit, but increasingly laterally. One could then see within the orbit a mass of greyish-white tumor, not well-defined and circumscribed like that within the cranial chamber, but more like a thick, diffuse, pale granulation tissue, which nevertheless formed a dense, hard mass at the center (Fig. 91). This mass was excised; it contained islands of orbital fat. A scar from the former operation reached the tumor and was so dense that it had to be cut away by sharp dissection. The removal of the mass was, therefore, not a clean, sharp dissection such as an encapsulated tumor would have permitted.

No attempt was made to expose the optic nerve because the eye was already blind. The ethmoidal cells were not opened. The frontal lobe had been quite severely injured by the removal of the tumor and the tip of it was excised (73.5 grams). The tumor weighed 16.8 grams; this included

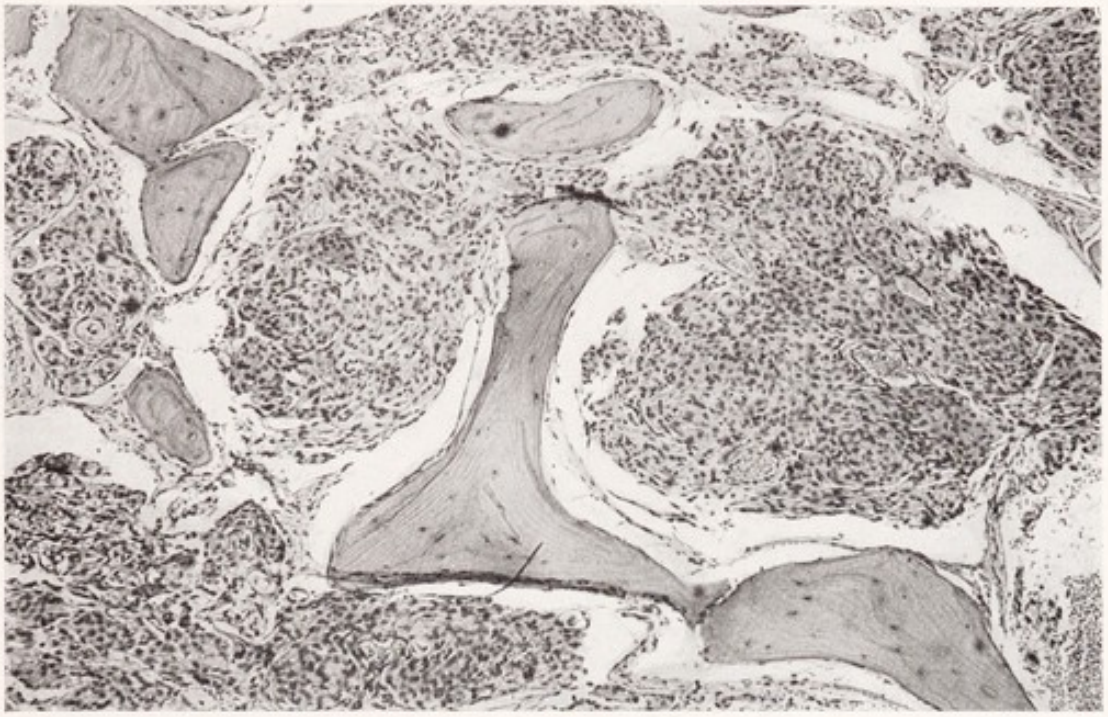


FIG. 92. Case 23. Photomicrograph showing invasion of Haversian canals of orbital roof by tumor cells.

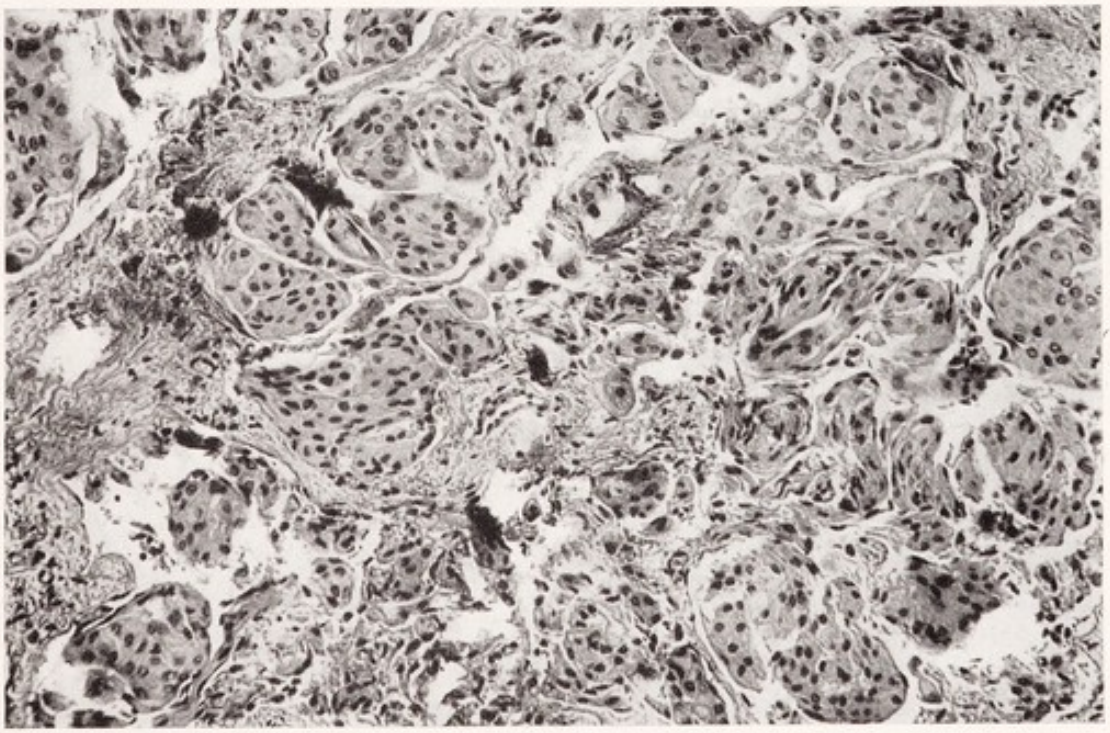


FIG. 93. Case 23. Photomicrograph of tumor within orbit and cranial chamber.

both the intraorbital and the intracranial tumor and the roof of the orbit. The intracranial part of the tumor was probably about two-thirds of the total.

The postoperative course was uneventful and the patient was discharged from the hospital on March 31, 1939. The edematous eyelids remained unchanged.

Microscopic Report: "The medullary spaces of the bone are blown out with masses of cells of dural meningioma type [Fig. 92]. The small area of soft tissue (somewhat suggesting granulation tissue at operation) over the orbital capsule is also made up of columns of the same type of cells [Fig. 93]."

CHAPTER IV

CARCINOMA OF THE ORBIT

CASE 24

E. B., History number 192907. Female, white, age 55. Admitted: February 5, 1940. Discharged: February 23, 1940.

Diagnosis: Carcinoma of the orbit. (Referred by Dr. Curtis Baylor, of Beirut, Syria, with this diagnosis.)

Complaints: Pain on the right side of the face; blindness in the right eye; drooping of the right lid.

Family and Past Histories: Negative.

Present Illness: Eight years ago the patient had begun to have pain in the distribution of the second branch of the fifth nerve. The pain was of a dull, aching character and gradually spread over the entire right side of the face. Five years ago the right eye had become somewhat more prominent; this gradually increased, but the increase had been more marked in the past year; ptosis of the upper lid developed about this time (Fig. 94). Diplopia had appeared only two years ago and then disappeared four months ago when this eye became totally blind. Nine months ago there had been loss of the sense of smell. The patient had never been without the pain, which was dull and aching, and was at this time mainly over the second branch, although the first branch of the trigeminus was also affected. She had had seven sinus operations on the antrum and sphenoid and many tooth extractions, with no relief of this pain.

Physical Examination: Negative, except for the fact that the patient was greatly undernourished, ill-looking, and in

severe pain. There were no signs of tumor in any part of the body.

Neurological Examination: Complete anosmia; total blindness in the right eye; complete ptosis of the right upper lid; 6 mm. of exophthalmos in the right eye. The extraocular movements were limited to 5° upward, 10° downward, 5° inward, and 2 to 3° outward. Tension of the eyeball was normal (eye examination by Dr. Frank Walsh). Vision

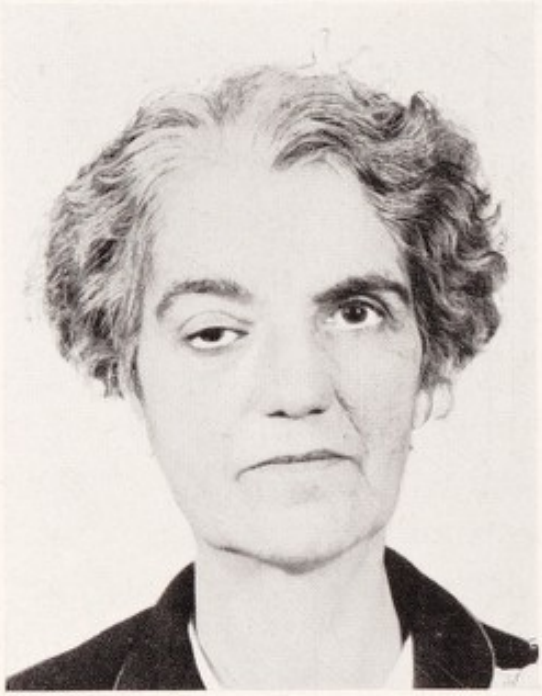


FIG. 94. Case 24. Preoperative photograph.

and visual fields in the left eye were normal. There appeared to be a diminution in the corneal reflex on the right side, but there was no other change in sensation over any part of the trigeminus domain. The right ear was totally deaf, but this had been of many years' standing.

Diagnostic Impression: Because of the long duration of the growth, and largely on the law of probabilities, a dural tumor in the roof of the orbit, or in the middle fossa with projection through into the orbit, was suspected.

Operation, February 10, 1940: For exploration of the orbit by the cranial route, the usual hypophyseal approach was made on the right side. The brain was under no increased

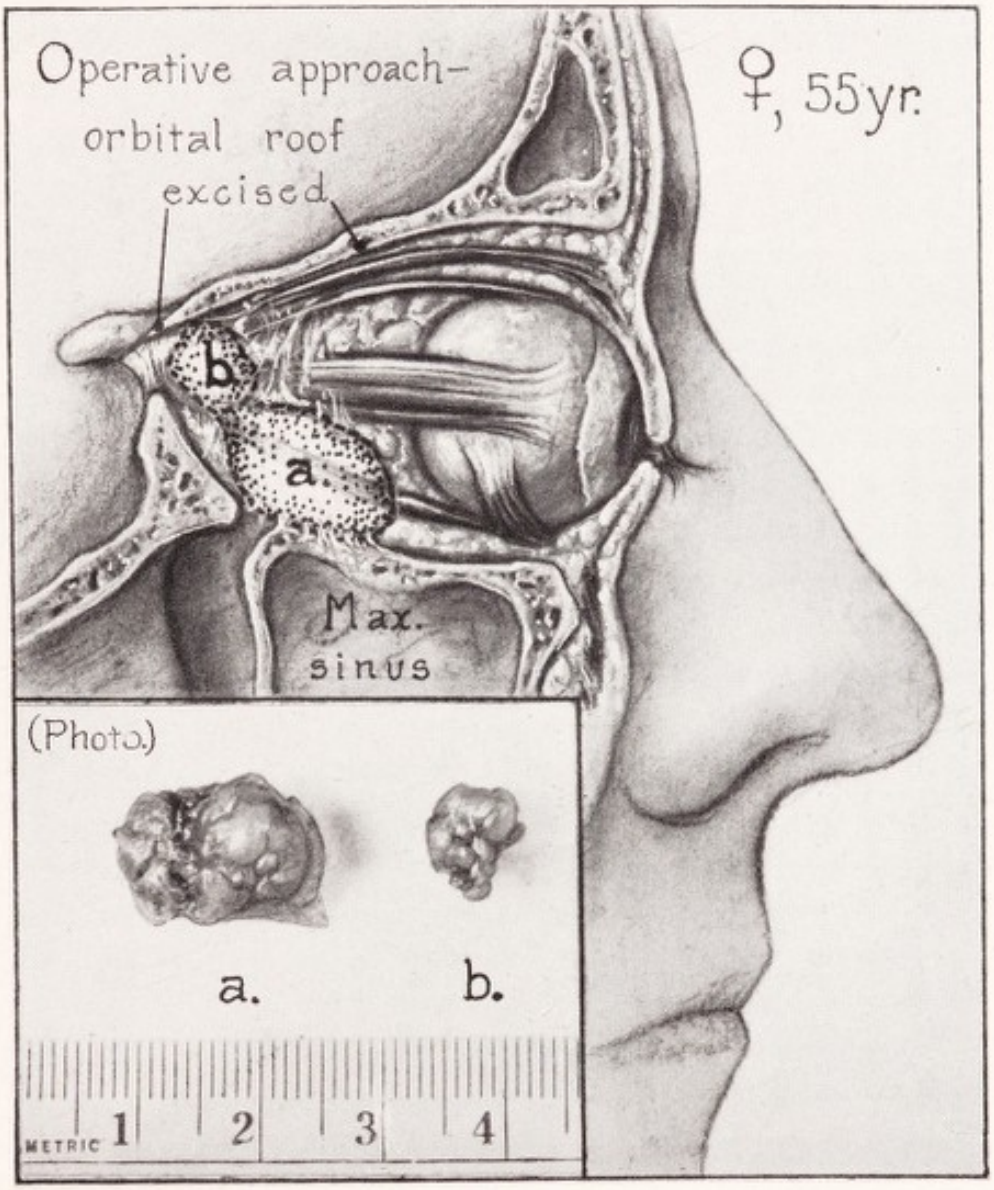


FIG. 95. Case 24. Operative sketch of tumor. Position of tumor on floor of orbit doubtless accounted for pain in distribution of second branch of trigeminus.

tension. The cisterna chiasmatis was evacuated and provided ample room. There was no tumor on the roof of the orbit, and from a thorough exploration under the temporal lobe it could be seen that there also was no tumor in the

middle fossa. The roof of the orbit was then completely removed; it was a little thin posteriorly, but otherwise normal. The capsule of the orbit was incised; there was but a very tiny amount of orbital fat and this was well anterior. A hard mass could be felt at the very apex of the orbit, and this extended over somewhat to the right and along the floor of the orbit; a well-circumscribed tumor was densely bound to the floor of the orbit. At the sphenoidal

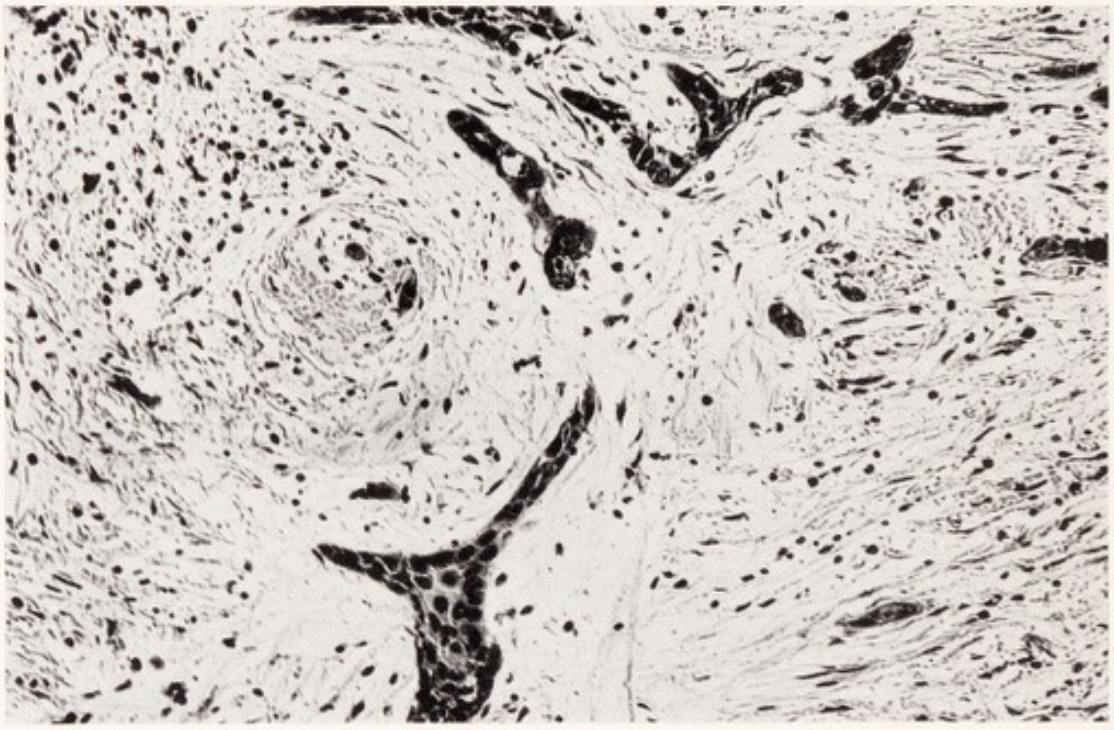


FIG. 96. Case 24. Photomicrograph of tumor, showing strands of carcinoma tissue in dense fibrous mass.

ridge where the extraocular muscles pass through, it was seen that these were densely matted together; a piece of this invasive mass was excised. It was very hard, gritty, and looked like carcinoma. The hard, smooth, well-rounded mass very tightly bound to the floor of the orbit was separated by the periosteal elevator and lifted from its bed (Fig. 95). This was a hard fibrous mass and seemed to have come out completely, although it must have grown into the tightly bound group of muscles at the sphenoidal ridge.

There was no bleeding when the tumor was removed. The wound was closed in the usual manner without drainage.

Postoperative Course: The patient's postoperative course was uneventful. She was entirely relieved of her pain from the time of the operation, but none of her other objective disturbances had been altered, except that her exophthalmos had practically disappeared.

Microscopic Report: The section showed dense fibrous tissue which in places included extraocular muscle. There were occasional little strands of carcinoma tissue (Fig. 96). The origin of the carcinoma could not be told from the section, though the suggestion was made that it was probably from the antrum.

The presence of carcinoma was not suspected because of the very long duration of the tumor's growth; the very small amount of carcinomatous tissue in comparison with the amount of fibrous tissue was most striking.

CHAPTER V

BRIEF SUMMARY OF SEVEN ADDITIONAL NONOPERATIVE CASES

In order to amplify the pathological study of orbital tumors, seven other cases of tumors from my experience, which were not subjected to this operative attack, have been added to the twenty-four cases of our operative series. The pathological study, therefore, is based upon thirty-one cases of tumors of the orbit.

These include Case 25, a diffuse osteoma—the case described as the introductory example of this report; Cases 26, 27, 28, and 29, of gliomatosis of both optic nerves and of the chiasmal region and brain. In each of these cases, the tumor was exposed at the chiasm and the visual tracts found to have been hopelessly destroyed by the growth, which expanded the optic tracts to a relatively enormous size and pushed through the orbital foramina. Despite entrance of the tumor into the orbital cavity, two of these cases had not yet produced exophthalmos. Case 30 was a dural meningioma operated on through the orbit in another hospital, a cerebral abscess following a few months later. The tumor was found at necropsy. Case 31 was a periosteal sarcoma of the orbit and temporal muscle.

CASE 26

M. W., History number U-30076.

A four and one-half-year-old white girl entered the hospital with signs and symptoms of a brain tumor: blindness, weak-

ness, and partial anesthesia of the left side of the body. There was no exophthalmos.

Partial removal of the tumor was made in the parietal-occipital region. The cortex was opened wide and one could see a tremendous mass growing from the base of the brain and into the right parietal and occipital lobes. The tumor arose from both optic tracts. The right optic nerve was diffusely enlarged; this enlargement extended through the orbital foramen, but was not enough to cause exophthalmos. The left optic nerve was also a large round mass and extended through the orbital foramen. The optic chiasm was incorporated in the tumor, which extended up into the temporal, parietal, and occipital lobes, filling the region of the third ventricle. The child died March 19, 1930, two days after admission. An autopsy was obtained.

Microscopic sections showed the tumor to be a glioma (astrocytoma).

CASE 27

T. V., History number U-58061.

A seven-year-old white boy was operated upon for a brain tumor September 25, 1934. The child was totally blind in the right eye and the vision was grossly defective in the left. In addition, there was palsy of the sixth nerve on each side, some anesthesia in the right hand and a weakness on the right side of the face. There was no exophthalmos.

A hypophyseal approach was made on September 29. A round, hard, white tumor, as large as a hickory nut, was found involving the right optic nerve and extending backward to the chiasm and forward through the optic foramen into the orbit. An attempt was made to remove it in order to check the progressive loss of vision in the left eye, but it was soon found that the tumor extended backward into the chiasm and could only be partially removed (Fig. 97).

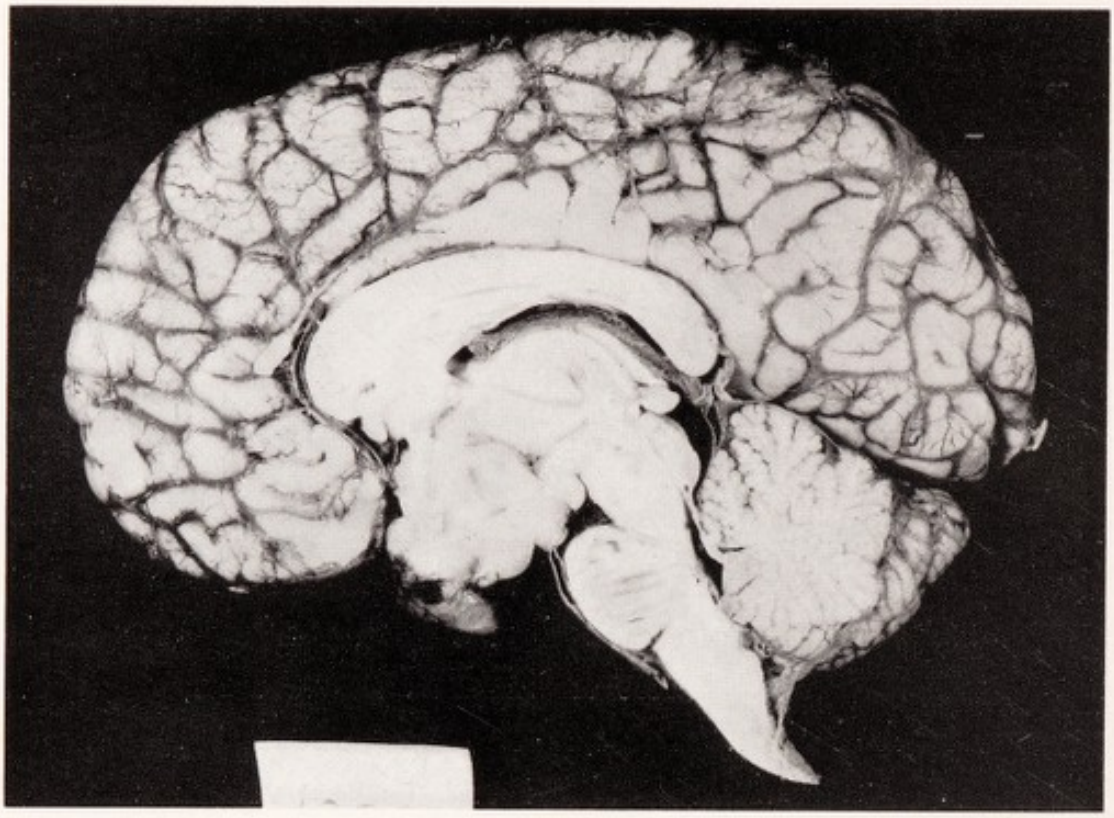


FIG. 97. Case 27. Midsagittal view of brain showing extensive continuation of glioma of optic tracts into hypothalamus and third ventricle.

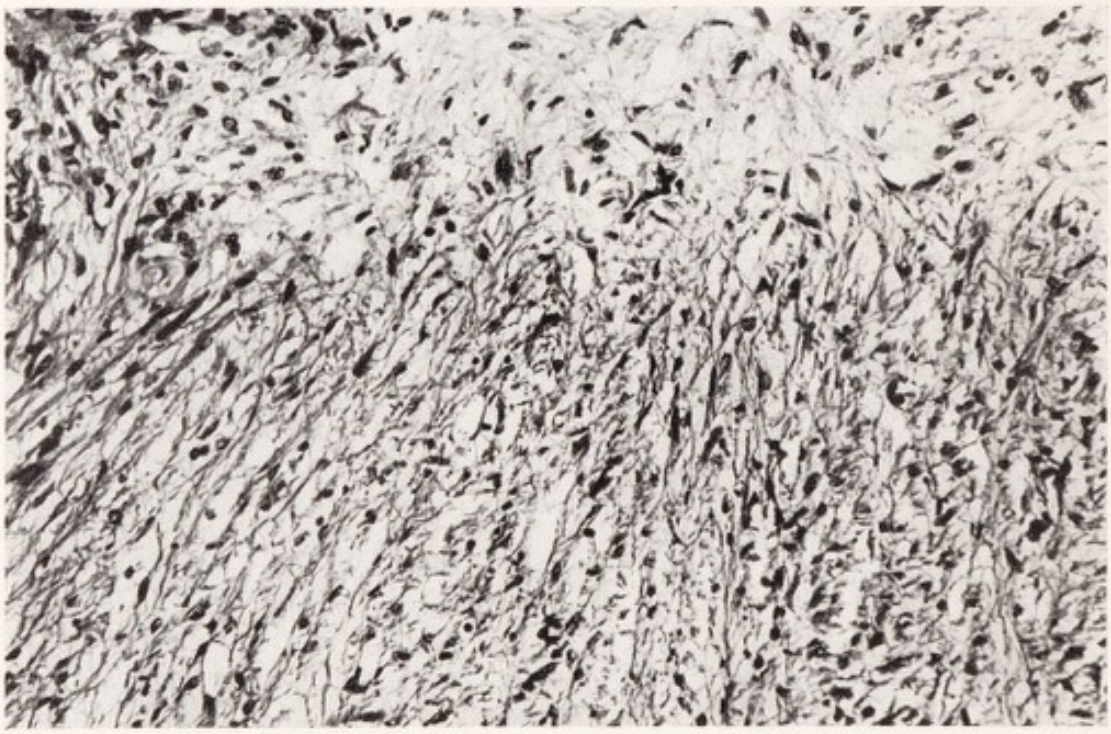


FIG. 98. Case 27. Photomicrograph of a glioma of the optic nerves in the visual tracts.

The patient died during the night. An autopsy was obtained.

Microscopic Diagnosis: Glioma (astrocytoma) (Fig. 98).

CASE 28

J. P., History number U-57831.

A five-year-old white girl had exophthalmos and loss of vision in the right eye. The region of the optic chiasm was exposed September 15, 1934. A white, well-rounded,

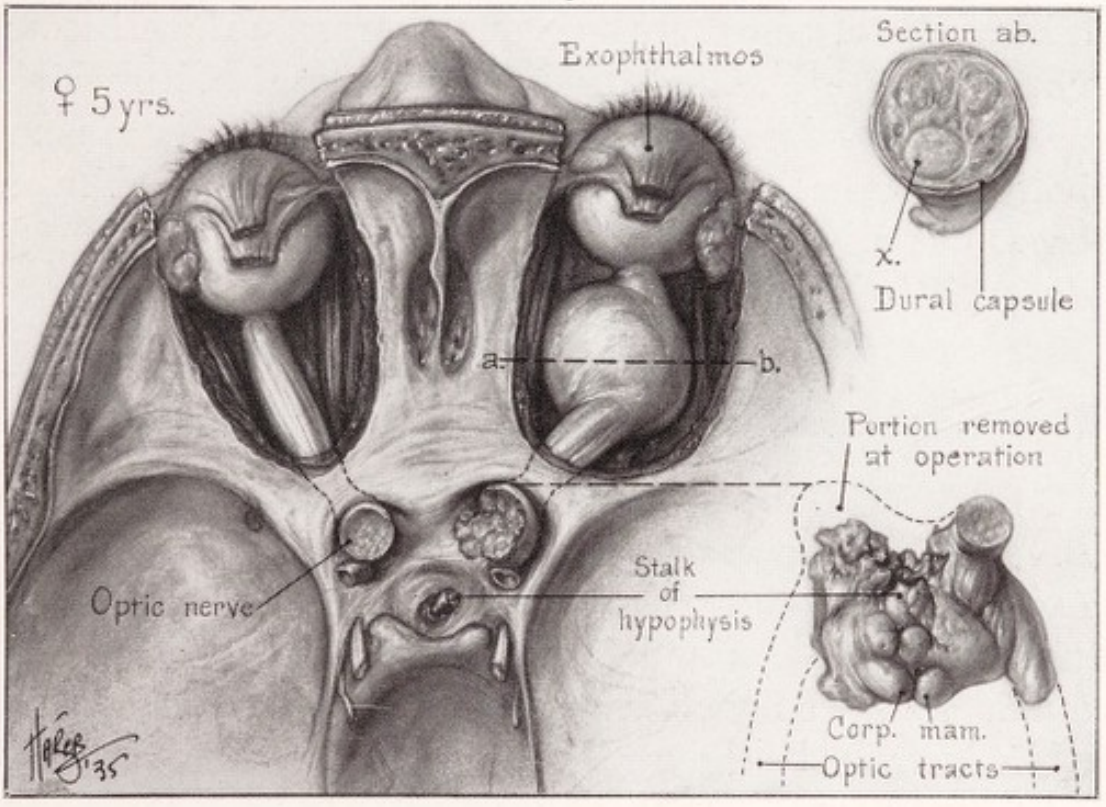


FIG. 99. Case 28. Drawing of diffuse gliomatosis of optic tracts. One large nodule is on nerve in right orbit, and growth has involved entire chiasm, with inclusion of hypophyseal stalk and invasion of overlying brain.

hard tumor of the optic nerve was encountered; it continued on through the optic foramen into the orbit. An attempt was made to remove it by cutting through the optic nerve at the optic foramen and dissecting the tumor backward,

but it was soon found to involve the entire optic chiasm, which was diffusely enlarged. The enlargement, however, extended into the left optic tract. The chiasm was as large as a thumb.

Death followed during the night. An autopsy was obtained. It showed another large tumor of the optic nerve within the right orbit (Fig. 99); this was continuous with the intracranial tumor. At the optic foramen there was a similar but smaller tumor of the left optic nerve; it extended into the orbit.

It is worthy of note that the patient was said to have become pigmented since the onset of her symptoms; also, her weight had doubled and there was some degree of polyuria and polydipsia. The hypophyseal stalk was pushed backward but was not destroyed by the tumor.

Anatomical Diagnosis: Gliomata of the entire visual tracts.

The accompanying drawing (Fig. 99) shows the extent of the tumor.

CASE 29

L. M. G., History number U-130317.

A normal-looking girl of thirteen years complained of blindness of one eye and hemianopsia in the other. The hemianopsia in the good eye had been a recent development; the blindness in the right eye had been first noticed three and one-half years ago. At this time a tumor at the nerve head was diagnosed from an ophthalmoscopic examination. There was exophthalmos (3 mm.) in the right eye.

An intracranial approach was made and a tumor found at the optic chiasm. It extended through the orbital foramen, which was enlarged in the X-ray (Fig. 100). It also extended over to the left optic field and apparently through the optic foramen on this side also. It was a very large mass of

tumor, filling the entire suprasellar region. A small piece of this was removed for microscopic study. It was semigelatinous and looked very much like a myxosarcoma, but microscopically it was a glioma (astrocytoma).

The girl died fifteen days later. An autopsy was obtained.

The accompanying drawing indicates the size and distribution of the tumor.

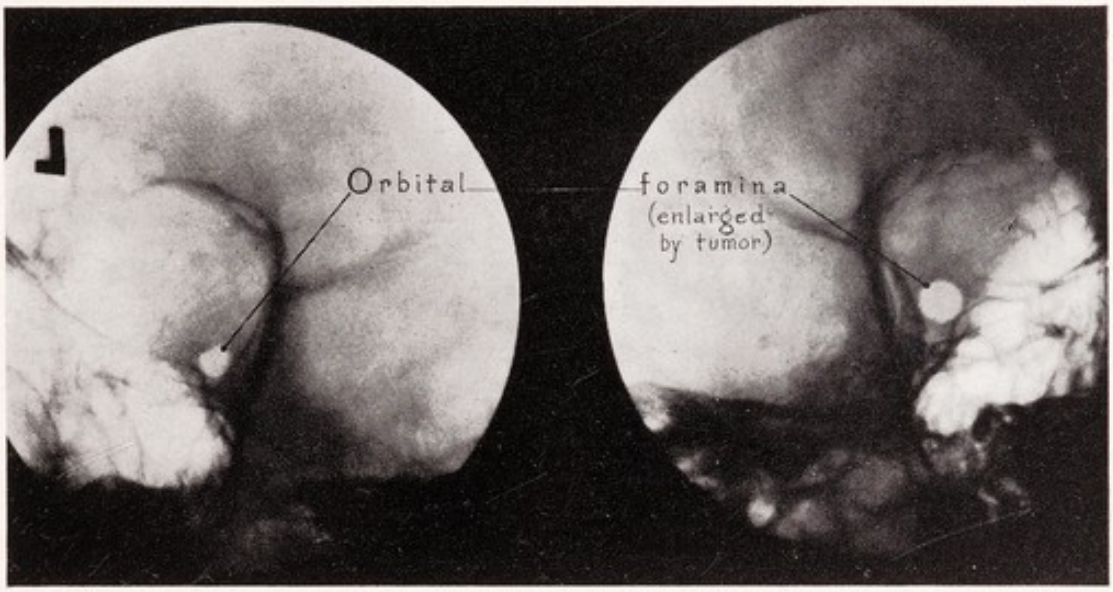


FIG. 100. Case 29. X-rays of orbital foramina showing enlargement of foramen on side of tumor, which has extended directly through this canal.

CASE 30

L. S., History number U-116589.

A white man fifty-five years old was operated upon September 26, 1937, in the Wilmer Clinic, the entire contents of the orbit being removed. The bone was found to be bare in one area and this was curetted. Apparently from this area infection was carried into the cranial chamber and a large abscess developed in the groove between the temporal and frontal lobes. This was opened April 11, 1938. At the time the roof of the orbit was exposed, some of the tissue lining this intracranial growth was removed and this was

found to contain columns of cells of dural endothelioma. Lining the entire surface was a thin layer of granulation tissue, and it was in this that the columns of dural cells were found.

The X-ray showed a marked thickening of the entire roof of the orbit, extending over to both lateral surfaces.

The patient died on May 3, 1938. At autopsy a large abscess filled the frontal lobe; there was a dural perforation in the orbital roof through which the cerebral infection had entered. The wall of the abscess covered a thin plaque of tumor that grew from the dura of the orbital plate.

CASE 31

N. J., History number U-117430.

A well-nourished man fifty-three years old had a rapidly developing tumor in the right temporal region. This was operated upon by Dr. Dean Lewis on September 13, 1937, and a small portion of the tumor in the temporal region removed. The cellular malignant tumor was diagnosed as periosteal sarcoma.

A few months later the patient returned with marked exophthalmos on the same side. The orbital contents were removed and radium applied. The same type of tumor was found. X-ray showed a large area of calcification extending into the roof of the orbit and through it into the orbital cavity. Radium had no effect upon the progress of the tumor and the patient died in March, 1938. No autopsy was obtained.

Microscopic Diagnosis: Periosteal sarcoma.

CHAPTER VI

SUMMARY
PATHOLOGICAL STUDY OF ALL CASES

The character, number, and percentage of tumors from the entire series of thirty-one cases may be summarized as follows:

CHARACTER	NO.	PERCENTAGE
1. Inflammatory mass	1	3.2
2. Pure fibroma	1	3.2
3. Pure osteoma	2	6.5
4. Osteomatous cyst	1	3.2
5. Spindle-cell sarcoma	1	3.2
6. Round-cell sarcoma	1	3.2
7. Schüller-Christian's disease (probable)	5	16.2
8. Dural meningiomata of nerve sheath (bilateral)	1	3.2
9. Dural tumors without hyperostosis of the skull (by X-ray)	2	6.5
10. Sarcoma with tremendous hyperostosis of the skull	1	3.2
11. Dural meningioma with hyperostosis of the skull	9	29.1
12. Periosteal sarcoma	1	3.2
13. Carcinoma	1	3.2
14. Glioma	4	12.9

Inflammatory Nodule: Case 1, well three and one-fourth years later. The proximity of the paranasal sinuses would suggest a much greater frequency of lesions of this type, in view of the fact that it must be the exceptional case that would form a persisting mass. Most infections would

doubtless subside quite spontaneously, or following evacuation of an orbital abscess. There was nothing in the histological picture of this small mass to suggest tuberculosis or syphilis, and the subsequent history of freedom from progression over a period of three and one-fourth years would be ample evidence on this score.

Pure Fibroma: Case 2, apparently well four years later. The origin of this remarkable tumor is not entirely clear. It most probably arose from the inferior surface of the dura, to which it was so tightly attached that the dura had to be excised with the growth. Moreover, judging by its intracranial position, where it must have been for many years before breaking through the roof of the orbit and the cribriform plate, dural origin would appear even more probable. But it was unlike any of the other dural meningiomata in that it contained so few cells. Stokes and Bowers (1934) reported a pure fibroma of the orbit; it was made up of whorls of fibrous tissue.

Pure Osteoma: By pure osteoma is meant a growth of bone that is not due to a soft tissue-invasive tumor that causes hypertrophy of the bone. The latter type of growth occurs with such relative frequency in this series that a degree of skepticism is justified when the diagnosis of a pure osteoma is made. One patient (the first reported in this book but not included in the operated series) has been followed for twenty-one years and is known to have had the growth forty-three years (since the age of six). The tumor was fully exposed intracranially as well as intraorbitally and no sign of an additional soft tumor was found. Moreover, the duration of the tumor's life, with almost no progression in twenty-one years, would safely appear to exclude the possibility of an additional soft tumor. The second patient (Case 3) has been followed only two years. It is only possible to say

that no soft tumor was found when the entire roof of the orbit was removed. The roentgenological picture of a pure osteoma differs but little from that of the larger group of secondary soft tumors. Perhaps in these two cases the growth invaded the paranasal sinuses to a greater extent, but the involvement of the roof and side of the orbit and the walls of the skull appears to be precisely similar. The bony growth was quite diffuse in both cases and its complete removal therefore impossible. It can only be hoped that the very slow growth in the second case will be like that in the first.

Well-defined, localized pure osteomata also occur, but have not been encountered in this series. So frequently associated with the frontal or ethmoidal sinuses and breaking into the orbit, they have been grouped by Cushing (1927) under orbitoethmoidal osteomata. Another case has been reported by Pilcher (1938).

Osteomatous Cyst: This unusual tumor (Case 4) arose in or below the floor of the orbit and projected upward into the orbit. The cyst, with serous fluid, was perhaps as large as a bantam's egg. Lining the exposed part of the cyst wall was a thin layer of tumor tissue. Microscopically, the lining of the tumor was made of cellular connective tissue throughout which were thickly interspersed small roughly spherical and irregularly shaped masses of bone (Fig. 26). There were numerous osteoblasts and large multinuclear osteoclasts. In most of the bony deposits there were osteocytes. The newly formed bone had none of the arrangements of true bone.

Spindle-Cell Sarcoma: Case 5, well and without signs of recurrence nearly three years later. It is worthy of note that this tumor was removed by intracapsular enucleation—a procedure that would favor local implantation at the site

of removal—and yet there has been no recurrence. At the time of the operation the tumor was thought to be malignant and the lapse of time without recurrence has been a pleasant surprise, the more so because the other eye had been removed several years before, following an accident.

Round-Cell Sarcoma: The single case of a pure round-cell sarcoma (Case 6), in a child of two and one-half years, developed symptoms rapidly, and recurrence quickly followed the removal of the growth. In the contiguous region several metastatic or transplanted nodules appeared a few weeks after the operation. It was much the most malignant tumor in the entire series, death occurring five and one-half months after the first sign of exophthalmos.

Schüller-Christian's Disease: Case 7 is a clinical example of a nodule of this type which so characteristically perforates the bone. Histologically there are numerous masses of foam cells, with diffuse infiltration with eosinophils and small round cells in great abundance. The duration of life cannot be told, because the patient has just recently been operated.

The four remaining cases (8, 9, 10, and 11) were entirely dissimilar clinically and from the gross pathological appearance. The growth in Case 8 behaved like any slow malignant tumor, with local recurrence and localized tumor implants about the site of operation. The gross appearance of the mass was like a hard, slowly growing invasive tumor. The patient is still alive six and three-fourth years after the first symptoms. In none of the sections obtained at two operations were there tumor cells. Because of the foam cells and eosinophils, and considering the microscopic, gross, and clinical picture as a whole, Dr. Rich looked upon the diagnosis of Schüller-Christian's disease as the most probable one.

The growth in Case 9 was very rapidly enlarging (total

duration of symptoms until death was ten months) in both orbits, within the cranial chamber, and scattered throughout the body. The tumors were salmon-colored. From the gross appearance Dr. Roger Baker, of Duke University, made the diagnosis of xanthomatosis. From the microscopic study of the various masses removed at operation, Dr. Rich felt that the diagnosis lay between a xanthofibroma and Schüller-Christian's disease. No tumor cells were present.

The tumors in Cases 10 and 11 were quite similar in their gross appearance. They were single masses of tissue which, in the gross, were like hard tumors. Both were firmly fixed, one at the apex of the orbit, the other to the orbital capsule; both had invaded the extraocular muscles; and in both, although the mass could be extirpated, it was clear that the contiguous tissues were infiltrated and that the removal was incomplete. In neither of these, nor in the two preceding cases, were there perforations of the cranial vault, which are so typical of Schüller-Christian's disease. Only Case 7 showed this bony perforation. This diagnosis in the remaining four cases has, therefore, been made solely upon the histological picture, which more closely resembles Schüller-Christian's disease than any other known lesion.

Many cases of Schüller-Christian's disease affecting the orbit have been reported, some as xanthomatosis and others as lipoid granulomatosis, and, since they are not actual tumors, many have been classified as pseudotumors—a designation proposed by Birch-Hirschfeld. Examples of such reports are those of Ringel (1922), Rowland (1928), Chester (1930), Chiari (1931), Heath (1933), Knapp (1934), and Sautter (1938). Among seventy primary orbital tumors Schreck (1939) includes twelve as pseudotumors, a proportion of non-neoplastic masses not greatly different from that in this series. Six of these were reported to be chronic

inflammatory tissue—a diagnosis that could easily have been made in two of my cases of Schüller-Christian's disease that only Dr. Rich's expert eye would have placed in this group. Two of Schreck's twelve cases were called lymphogranuloma and aleukemia lymphadenoma. As Schreck does not mention Schüller-Christian's disease in his cases, one is led to wonder if many of them may not have been of this origin.

Voelkel (1937) collected forty cases of bilateral symmetrical orbital growths, most of which he thought to be of lymphomatous origin. Doubtless some were similar to our Case 9.

Psammoma (Dural Tumor of Optic Sheath): Almost identical tumors were present in both optic nerves (Case 12). The point of origin of the growths was in the dura in the optic canal, from which each projected slightly intracranially and much more intraorbitally. On the operated side the tumor had strangulated the optic nerve like a tight constricting band. Microscopically the tumor was made up of fibrous tissue and was loaded with psammoma bodies. Although our patient's vision was not improved by the operation, she is still alive and well, except for blindness, twenty-four years after the onset of symptoms. Olivecrona (1935) reported an almost identical case—also bilateral. Schott (1877) reported bilateral symmetrical psammomata of the optic nerves at the optic foramen, but they were more circumscribed than mine.

Dural Tumors without Roentgenological Evidence of Hyperostosis of the Skull and Orbit: The qualification of roentgenological evidence is added because it is exceptional in this series (Cases 14 and 15) for a dural tumor to be unassociated with hyperostosis. As a matter of fact, a definite thickening of the sphenoidal ridge was seen at operation in both cases, although not demonstrable in the X-rays either before opera-

tion or on subsequent restudy. The very highly malignant tumor in Case 14 only occasionally showed the arrangement of cells so characteristic of a cellular dural meningioma. Had one not known its character from the gross picture, a diagnosis of sarcoma might well have been made. The duration of life was two years after the first operation. Following operation, transplanted nodules developed about the operative site.

The second tumor (Case 15) was like others of the cellular type of dural growth. The cells in most part were grouped throughout the sections in round or oval columns of varying size. The same columns of cells were found in the marrow spaces of the bone. Histologically this case is similar to the succeeding cases of tumors with hyperostosis.

Tumors with Roentgenographic Evidence of Hyperostosis of the Skull and Orbit: Nine tumors, or over one-third of the operated cases of this series, were associated with such pronounced hypertrophy of the skull and walls of the orbit that it was strikingly shown in the X-ray. The cause of this relationship, recognized by Spiller (1907) and Brissard and Lereboullet (1903), was probably first demonstrated by Cushing, in 1922. Under the microscope he found the marrow spaces in the bone to be filled with tumor cells (dural meningioma in each instance). Phemister (1923), Penfield (1923), and Bérard and Dunet (1924) quickly verified Cushing's findings. This combination of a dural tumor and superimposed thickening of bone occurs not only in orbital growths, but also in those located along all parts of the vault and base of the skull, and always there is the same explanation—the cells of the dural tumor have invaded the overlying skull. That the hyperostosis is not absolutely, although nearly so, pathognomonic of a dural meningioma is shown by the same extensive bony proliferation in a

case of spindle-cell sarcoma (Case 13): the same sarcomatous cells that made an enormous soft tumor also filled the marrow spaces of the skull. Doubtless the bony change is quite similar in its development to that of the well-recognized hypertrophies of the vertebral column associated with metastases from carcinoma of the prostate.

The X-ray pictures of the benign osteomata in this region and elsewhere present no demonstrable difference from those of the invasive meningeal growths. Doubtless the age of the patient is an important differential point, for in both of the benign hyperostoses the bony thickening was present in the first decade. And in none of the dural tumors was the lesion known to be present before the thirtieth year. Although benign osteomata may well appear later, the burden of proof must rest on the acceptance of such a diagnosis for ages later than this.

The size, position, and general character of the dural tumor in relation to the size of the hyperostosis of the orbit and skull are worthy of consideration. In Case 16 the dural growth is especially interesting because there was no dural meningioma on the side of the dura contiguous to the hyperostosis. It was only because the dura was opened that it was seen at all; and then it was only a flat plaque, about the thickness of paper and very loosely applied to the cerebral side of the dura. It gave no gross appearance of having infiltrated the dura. It was stripped from the dura with the greatest ease. Sections of the bone, which was tremendously thickened, showed the same type of tumor. In two other cases (17 and 22), the latter studied at necropsy, there was no gross dural tumor, but in the orbital fat of each there was a small, diffuse, irregular mass looking like firm granulation tissue, and this proved to be tumor. From the gross appearance one would be led to wonder whether

this orbital tumor might not have been a secondary outgrowth from the tumor within the bone. Could this dural tumor have arisen from dural rests within the bone, which in each instance was of enormous size? In Case 22 the hyperostosis was the most extensive of the entire series. In Case 23 the dural growth was hard and flat, about 1 cm. thick, and spread like a carpet over the entire orbital roof, to which it was firmly attached. It extended posteriorly, covering the dura over two-thirds of the floor of the middle fossa; the hyperostosis was of moderate grade. In Case 19 the dural tumor, arising from the orbital roof, was about the size of a hazelnut. In Case 20 a hazelnut-sized dural tumor projected from the sphenoidal wing. In Case 21 a small round dural tumor (2 cm. in diameter) projected intracranially from the orbital roof, and the irregular mass of greyish-brown tissue-like granulation tissue projected into the posterior part of the orbit.

On the other hand, the dural tumor in Case 18 was of great size and very hard. In this case the orbital roof was destroyed and the hyperostosis was confined to both frontal bones.

Viewed histologically, the tumors are—with one exception (Case 18)—all of the very cellular type of meningioma and are lacking in the whorls and palisades of fibrous tissue. In Case 18, however, the tumor is made up of whorls and palisades and is lacking in the solid masses of cells. Both types of dural tumor can, therefore, be responsible for the extensive contiguous hyperostosis. Moreover, the degree of hyperostosis is just as great with small circumscribed or thick carpet-like tumors as with the larger growths. Cushing (1938) commented upon the fact that a greater proportion of dural tumors "en plaque" produced hyperostosis, than of the large, hard, rounded tumors which he terms

the "global type." However, the same change in the bone is produced by both types. Surprising indeed is the extensive and widespread distribution of the hyperostosis, which so frequently involves much of the orbital bones, the side and front of the skull, and all too frequently the paranasal sinuses. So much are the paranasal sinuses involved that these growths have been included by Cushing under "orbitoethmoidal" tumors.

Operative removals of tumors of this type have been reported by Cushing (1922), Armitage (1930), Patterson and Cairns (1931), Vorhis and Adson (1934), Love (1935), Hoover and Horrax (1935), Vincent and Mahondeau (1935), Pilcher (1938), and Cairns (1938). Knapp (1938) reported two cases, one operated locally, the other treated by X-rays. Elsberg, Hare, and Dyke (1932) reported five cases of tumors of this type, in some of which the dural tumor was removed, but apparently without removal of the orbital hyperostosis.

Glioma of the Optic Tracts: All of the four cases added for pathological study were explored surgically, but in none did the operation include the orbital attack, the tumor being clearly inoperable. In two of the cases there was no exophthalmos, despite the fact that the growth had continued into the orbit. All were widespread growths involving the chiasm, one or both optic nerves within the cranial chamber and also in the orbits, and the brain. In one instance the tumor extended through the third ventricle and the temporal, frontal, parietal, and occipital lobes on one side. All were in young children, the ages being four, five, seven, and thirteen. This type of tumor may be suspected when hemianopsia without exophthalmos is present or beginning. An early case of this type was reported by Willemer (1879); an excellent drawing was included. Other cases have been reported by Martin and Cushing (1923).

Lundberg (1935) made an extended study of this tumor, which he classified as an "oligodendrocytoma." He emphasized the early age of onset. Busch (1937) suggests partial extirpation of the growth and records two cases with improvement after two and one-half and seven and one-half years. However, very little operative trauma can be sustained without mortality, because the subsequent edema involves the hypothalamus, which tolerates trauma poorly.

Other Tumors in the Orbit: The foregoing pathological report by no means exhausts the orbital tumors. It represents merely the cases that have occurred in the experience of a neurosurgeon, and very probably are fairly representative of the commoner types. Coulter and Coats (1910) reported an orbital teratoma and Gow (1934) a large cyst that was probably of this character. Mancilla (1923) and Byers (1924) each reported a cavernous angioma of the orbit; Adson and Benedict (1934) a hemangioendothelioma; Franklin and Cordes (1924) a lymphangioma; Knapp (1923) an oil cyst lined by epithelial cells and containing many sebaceous glands and a few hair follicles; Wheeler (1937) an orbital cyst with epithelial lining; Stallard (1938) a cavernous angioma; DeSchweinitz (1915) a myxosarcoma and a lipoma; Pinkus (1933) two cases of cholesteatoma of the orbit; and Cairns (1938) a cholesteatoma arising beneath the orbital roof. Van der Hoeve (1925) reported a Recklinghausen's neurofibroma, and Steuer (1922) two others of this type; the latter also included an epidermoid with slight hyperostosis of the orbital roof with six cases of dural hyperostosis causing exophthalmos. Davis (1939) reported two cases of plexiform neurofibromatosis, one of which was associated with a glioma of the optic nerve. There are doubtless few types of tumors encountered elsewhere in the body that do not also appear in the orbit.

CHAPTER VII

THE OPERATIVE PROCEDURE

The size of the cranial exposure has been greatly reduced since the earlier report. This is in large part due to the use of avertin as the routine anesthetic in operations upon the brain. The great operative disadvantage of ether lies in the swelling of the brain that usually follows. It was to make allowance for this that the larger bony exposure was required. With avertin the brain does not swell and therefore a much smaller bony opening will serve just as effectively as the larger one required when ether is used.

For exposure of the orbital roof, the regular hypophyseal approach is made. The concealed incision has for many years been employed in all operations upon the brain, with no loss of room or increased difficulty of approach. The cutaneous incision begins about 2 cm. anterior to the ear, passes straight toward the midline and, when about 3 cm. from it, makes a sharp curve forward and ends anteriorly at, or slightly in front of, the hairline. The galea and skin are then stripped from the bone and temporal muscle and retracted anteriorly.

The bone flap is made so that the anterior border misses the frontal sinus and skirts the supraorbital ridge as far laterally as possible. When turned back and broken under the temporal muscle it is retracted laterally and is well out of the way. The dura is opened just within the bony margin and the roughly circular incision is so complete that it becomes practically an autotransplant, thus being a factor in preventing postoperative extradural bleeding. The

frontal lobe is elevated and the cisterna chiasmatis evacuated, thus reducing the volume of the intracranial contents and providing room for attack upon the roof of the orbit. The head is then lowered to permit the frontal lobe to fall away without the need of traction.

If the cisterna chiasmatis is not opened, the brain so completely fills the operative field that undue pressure would usually be necessary to retract the frontal lobe to a degree sufficient to permit the attack upon the orbit and its contents.

From the optic foramen the dural covering of the orbital roof is incised in a curve sweeping laterally around the outer margin of the orbit, then curving anteriorly almost to the cribriform plate. The dural flap is stripped toward the midline with a periosteal elevator.

The initial opening in the orbital roof can frequently be made by applying slight force with the periosteal elevator at the thinnest point of the bone. The remainder of the roof is removed with rongeurs, care being taken not to extend the defect into the ethmoidal cells. In two of our cases this happened and rhinorrhea promptly followed, necessitating reopening of the wound and covering of the opening with bone wax and the flap of dura. At times the bone is so thickened (by an osteoma) that the chisel is required to make the primary defect, and it may then be necessary to complete the removal of the roof with numerous applications of this instrument.

The capsule of the orbit then presents and is incised longitudinally and the edges retracted. Either the tumor or orbital fat presents, depending upon the position of the growth. If fat presents, it is under pressure and extrudes and must be excised. Identification of the superior rectus muscle is usually immediate, unless the tumor is superimposed. This muscle is surrounded by a silk suture and

retracted to one side, thus providing inspection of the entire superior half of the orbit. From this view tumors in any part of the orbit would be found and their extirpation can be carried out deliberately. The optic nerve, the posterior part of the eyeball, and the ophthalmic artery and vein are readily identified.

In only one case in which evacuation of the cisterna provided much less room than normally was there any trauma of the frontal lobe. Rather than take any chance with subsequent postoperative edema and intracranial pressure, a small part of the frontal lobe was removed. In closing, the dura is snugly resutured, the bone flap replaced and wired, and the galea and skin closed without drainage.

There has been but one death in the series (Case 22). With the huge mass of bone along the base of the middle and anterior fossae, it doubtless would have been wiser to have been content with a partial removal of the growth with the chisel, rather than to attempt such an extensive extirpation.

The thought doubtless occurs whether a pulsating exophthalmos does not follow removal of the orbital roof. I have looked carefully for such a sequela but have never found it, despite the fact that pulsating exophthalmos is known to occur when the orbital roof is congenitally absent.

Although I have never done a lateral Krönlein operation for an orbital tumor, it does not seem possible that it can provide the exposure for the deliberate painstaking dissection of a tumor needed in order to avoid injury to important structures within the orbit. And for tumors that extend beyond the orbital cavity, the Krönlein operation would, of course, be perfectly futile. When an orbital tumor is small and protrudes anteriorly beyond the orbital ridge, the attack upon the tumor by an incision under the supraorbital ridge is doubtless adequate; but when the tumor extends back of the eyeball, the intracranial approach is advisable.

Before the operation is begun, the size and position of the frontal sinuses should be known. In making the anterior bony incision, it is important that the frontal sinus be avoided. Entry into a frontal sinus is a potential source of infection and of subsequent rhinorrhea. If a frontal sinus is opened, a flap of dura should be tightly sutured over the opening. If an ethmoidal cell is opened, it can be covered with wax and the dural flap replaced over it. A small opening in the frontal sinus may also be covered with wax, but the larger openings would require a dural flap. In none of the cases in this series has the frontal sinus been opened. In Case 18, the enormous tumor of long standing had completely obliterated the frontal sinuses, but the paranasal sinuses communicated with the wide-open cerebral chamber (the whole frontal dura on both sides was removed with the attached tumor). No attempt was made to close this defect and neither rhinorrhea nor infection followed. In two cases of this series, infection followed entry into the ethmoid sinus and the bone flap had to be sacrificed.

The method of attack upon the intracranial part of the tumor is dependent upon the nature and position of the growth. Usually the hypophyseal approach affords ample room. If the middle fossa or temporal lobe contains a large dural growth, it may be necessary to rongeur away an additional area of bone beneath the temporal muscle. This was done in two of our cases. It has not been necessary to enlarge the cutaneous incision or to turn down an additional bone flap, either of which, however, is easily possible if the situation should demand. Retraction of the temporal muscle with or without transverse division of its fibers will, after the underlying bone has been removed, expose most of the temporal lobe and even permit its resection in large part if necessary. The removal of a large dural endothelioma is always a difficult feat and requires ample room. Since the

arterial bleeding is from the middle meningeal artery, the tributaries of which cover the middle fossa to the foramen spinosum, it is essential that access to this area be unimpeded by inadequate exposure. The dural attachments should, when possible, be totally excised. As long as the dura along the floor of the middle fossa is the site of origin, this is not difficult; in fact, the removal of this dura also greatly facilitates control of the severe bleeding from the middle meningeal artery. Not infrequently control of this bleeding is best accomplished by immediately following the middle meningeal artery to the foramen spinosum. Ligation or cauterization of the trunk of this vessel is usually far easier and safer than attempting to control the numerous bleeding points from its many branches.

The problems associated with the intracranial portion of these tumors are those encountered in any series of brain tumors. The risk involved in removing combined intraorbital and intracranial growths is confined almost exclusively to the part of the tumor within the cranial chamber, and the group of cases described shows that, even with enormous growths, the danger is relatively slight.

Occasionally the internal carotid artery is surrounded by tumor, or at least attached to it. This complication occurred once (Case 15) in this series of cases. To injure the internal carotid artery or the middle cerebral artery would mean disaster and must be avoided at all hazards. Rather than invite hemiplegia and probably death from arterial thrombosis by shaving the tumor too closely, there is no choice but to leave a nest of cells on this vessel.

When there is some exophthalmos and a dural endothelioma has been completely removed from the middle or anterior fossa, should one open the orbit? The answer is dictated by the fact that the bone about the tumor is thick-

ened because of tumor contained within it. In one patient with this condition (Case 15) I removed the orbital roof, but since the eye was already blind I did not incise the orbital fascia. Since the additional exposure of the orbit is simple and not time-consuming and adds practically nothing to the operative risk, the surgical possibilities should be completed in one stage. Moreover, a second stage is much more difficult and the possibilities of a complete cure are reduced.

The most difficult surgical problem in this series of tumors has been in attacking the bilateral frontal tumors of enormous size. This was done in two cases. A bilateral approach was made with a long sweeping curve from one ear to the other and just under the hairline. The bone was so thick that the ordinary instruments used for making bone flaps were of little avail. Between large deep burr openings the bone was cut in a curve from the supraorbital ridge on one side to that on the other, with large bone-cutting forceps. On turning back the huge bone flap on the supraorbital ridge, the dura, which was densely attached, tore with it; this included the anterior part of the frontal sinuses which, however, were practically obliterated by the tumor masses. This bone must, of course, be removed.

In May, 1884, a remarkable tumor extirpation was performed by Durante of Rome—perhaps the earliest successful tumor removal. It antedated by seven months the celebrated case of Rickman Godlee of England, to whom is usually accredited the first successful extirpation, although the removal was only partial. Whether or not Lister's aseptic methods were used is not stated in Durante's publication. His patient survived for many years, and was even apparently cured, the tumor doubtless being a dural endothelioma. His case happened to be one with exophthalmos

secondary to rupture of the orbital roof. A drain was placed from the depths which the surgeon thought to be the nasal cavity from rupture of the ethmoids, but one is led to wonder whether on account of the exophthalmos it might not have been the orbit; no infection followed. Durante's diagnosis was based upon the loss of smell, and is, therefore, one of the very earliest cases operated after a localization of the growth had been made from neurological signs. The study of cerebral localization, based upon the experiments of Fritsch and Hitzig and of Ferrier, was just beginning to stimulate interest and bring results. Except for the fact that the frontal bone was chiseled away and therefore sacrificed, and that no attempt was made to remove the orbital roof, the operative approach was strikingly similar to that presented in this book.

In his publication on meningiomas (1938) Cushing called attention to this approach, which he expected to use in future operations on tumors of the orbit. Adson and Benedict (1934), Hoover and Horrax (1935), and doubtless others, have used this approach to remove orbital tumors.

CHAPTER VIII

CONCLUSION

Proportion of (1) Purely Orbital and (2) Combined Orbital and Intracranial Tumors

Of all the tumors included in this report only six, or 20%, were confined solely to the orbit, and one of these was an inflammatory nodule. The combined orbital and intracranial tumors were, therefore, 80%. If only the operative series of twenty-four cases were analyzed, 25% would be solely orbital and 75% combined orbital and intracranial. Among the pathological and operative cases together, it is quite probable that all of the combined tumors arose primarily in the cranial chamber, except perhaps in the cases where the tumors were multiple (the four gliomata, and one case of Schüller-Christian's disease—Case 9). The earlier papers of Hudson and Byers prepared us for the very high percentage of combined orbital and intracranial tumors, but our figures are even higher than theirs. They, however, did not have the advantage of direct inspection of both cavities simultaneously, but were dependent upon the late-appearing necropsies or histories providing unmistakable evidence of subsequent intracranial lesions. If, however, we take the twenty-three postmortem examinations of orbital tumors reported in the papers of Byers and Hudson with twenty-one intracranial extensions of the growth, it will be seen that the involvement of both the orbital and cranial chambers actually exceeds the percentages in this report.

Malignancy of Tumors in This Series

In the series of twenty-four operated cases, the outcome is unknown in only one (Case 13). Five are known to have died: (1) Case 6, a round-cell sarcoma, three and one-half months after operation; (2) Case 9, a Schüller-Christian's disease or xanthoma, eight months after operation; (3) Case 14, a malignant dural meningioma, twenty-two months after operation; (4) Case 16, a dural meningioma with hyperostosis, ten years after operation; (5) Case 22, an operative death.

Three cases at least, and probably more, were benign and should be permanently cured: (1) Case 1, an inflammatory nodule, operated three and one-quarter years ago at this writing; (2) Case 2, a fibroma, four years ago; (3) Case 5, a spindle-cell sarcoma, three years ago.

The remaining cases must be looked upon as malignant, or as benign growths that could not be completely removed. The great group of dural tumors must be considered malignant because the orbital roof and frequently the skull over a large area had been invaded by the growth and its complete extirpation was, therefore, impossible. Their growth, however, is usually very slow and there should be relief for many years; Case 16 survived ten years, and all have survived a number of years. The same is doubtless true of the sarcoma with hyperostosis (Case 13). This is the patient whose subsequent course cannot be traced; he was well when he left the hospital. Two other potentially benign tumors could not be completely removed (Case 3, an osteoma, and Case 12, a bilateral dural psammoma of the optic sheath). The latter case has gone eighteen years since the operation and the tumor has made no observable progress. It is hoped that the osteoma (Case 3, operated two years ago)

will grow as slowly as that in the introductory case in this study, which has shown no recognizable progress in twenty-one years.

The four living cases of Schüller-Christian's disease must be considered malignant. One (Case 8) has gone nearly three years, but local recurrence is slowly progressive. The other three patients (Cases 7, 10 and 11) have been operated upon in the past few months, and although other lesions cannot be found and the orbital growth has been thoroughly removed with contiguous tissue, a cure can hardly be expected.

Of the series of twenty-four intraorbital tumors that have been operated upon by the intracranial route, six, or 20%, were confined to the orbit; eighteen, 80%, were combined intraorbital and intracranial growths; one of the former and two of the latter were metastatic. With the seven pathological cases added, the respective percentages are in even more pronounced contrast, $16\frac{2}{3}\%$ and $83\frac{1}{3}\%$. The pathological features of the tumors have been discussed.

The operative attack, proposed in 1921, is through an intracranial (hypophyseal) approach. The roof of the orbit is removed after evacuating the cisterna chiasmatis; retraction of the frontal lobe then provides sufficient room.

The operation is offered not only for all combined intraorbital and intracranial tumors, but also for those that are restricted to the orbital cavity. As a matter of fact, it is rarely possible before the operation to be certain whether or not the tumor also lies within the cranial chamber, as so many of them do (roughly 75–80% in this series). Their co-existence should, therefore, be assumed by the law of probability.

For tumors confined to the orbit this operation offers a far better exposure than is possible by any other method.

Deliberate, careful dissection of the tumor is possible only by this approach. There is, therefore, much less chance of injury to the extraocular muscles, their nerve supply, the optic nerve, and the ophthalmic vessels by this approach.

It offers the only hope of a permanent cure when the tumor is in both cavities, and for incurable cases it offers the maximum palliative result.

The operative risk—in safe hands—should be very low (4.1% in this series) both for tumors confined to the orbit and for those with intracranial extensions. Prior exenteration of the orbit, or removal of the eyeball, will prevent the utilization of this operation because the orbital tissues will be infected.

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