Congenital cysts of the fourth ventricle : a report of two cases associated with tumor of the optic thalamus and crus cerebri / by J. Ramsay Hunt.

Extracted from The American Journal of the Medical Sciences, March, 1904.

CONGENITAL CYSTS OF THE FOURTH VENTRICLE.[1]

A REPORT OF TWO CASES ASSOCIATED WITH TUMOR OF THE OPTIC THALAMUS AND CRUS CEREBRI.

By J. Ramsay Hunt, M.D.,
(of New York)
ASSISTANT INSTRUCTOR IN NERVOUS DISEASES IN THE CORNELL UNIVERSITY MEDICAL COLLEGE, NEW YORK; NEUROLOGIST TO THE CITY HOSPITAL; ASSISTANT PHYSICIAN TO THE MONTEFIORE HOME.

(From the Pathological Laboratory of the Cornell University Medical College.)

Introduction. The pathological conditions resulting in the formation of cysts within the fourth ventricle or a cystic dilatation of the ventricle are as follows:

1. Parasitic: Cysticercus or echinococcus cysts.
2. Cystic degeneration of a tumor.
3. Cystic degeneration of the choroid plexus.
4. Cystic dilatation of the fourth ventricle from occlusion of its communications.
5. Congenital cysts.

These are of pathological rather than clinical importance because of their rarity and the atypical character of the symptoms produced.

Both cases forming the subject of the present study are examples of brain tumor in young subjects with *congenital* cysts of the brain stem projecting into the fourth ventricle.

In both the intimate structural relation of cyst and tumor was demonstrable.

Summary.

Case I.—A boy, aged seven years, previously healthy, developed symptoms of brain tumor referable to the left optic thalamus. The important objective symptoms were: hemiparesis and hemiataxia of the right arm and leg; paralysis of the emotional innervation of the right side of the face; paresis of the left external rectus; nystagmus; hearing impaired on the right, both to aerial and bone conduction; no hemianopsia; no objective sensory disturbances.

Autopsy. Large tumor (mixed-cell sarcoma) situated in the left

[1] Read in abstract at a meeting of the New York Neurological Society, January 5, 1904.

optic thalamus, infiltrating the subjacent structures and appearing on the basilar surface of the pons.

The fourth ventricle is dilated by a large cyst, which is firmly attached to the floor, penetrates the substance of the pons Varolii, terminating in the midst of the neoplasm. Serial sections show the presence of glia and nerve fibres in all parts of the cyst wall continuous with the nerve structures of the pons.[1]

History. A boy, aged seven and a half years, previous history negative, was admitted to the Montefiore Home September 12, 1896. Complains of headaches, vomiting, defective vision, uncertainty of gait, and an awkwardness of the right side; mentally clear, but apathetic and lacks energy; speech stammering; awkwardness and weakness of the whole right side; tongue deviates to the right. Station uncertain; gait ataxic; hemiplegic. Pupils medium size, the right larger than the left; all reactions prompt. Lateral nystagmus; slight convergent squint; vision $\frac{3}{4\,6}$ on both sides. Optic neuritis greater on the right side. Hearing both to bone and aerial conduction is impaired on the right. Percussion of skull negative. Smell and taste normal.

Note November 26, 1897. Is dull and forgetful; explosive laughter; hearing on right defective; skin reflexes active; tendon reflexes active and greater in the right side; right ankle clonus. The active innervation of the face is alike on the two sides. In repose and when responding to psychical and emotional stimuli, paralysis of the right side of the face is evident. Inco-ordination of the right arm and leg, which are flabby and thinner than in the left; no disturbance of sensation; left parietal region, slight tenderness on percussion. (Under antisyphilitic treatment the general symptoms subsided, including the optic neuritis, and the objective symptoms showed great improvement.) Discharged.

Readmitted April 9, 1900 (Dr. Abrahamson). Ataxia, atony, and weakness of the right side. The volume of the extremities is diminished on the right side. Asymmetry of the face; in repose the right side drooping; this inequality is reversed on emotional innervation, and disappears on voluntary innervation. Paresis of left external rectus with internal strabismus; lateral nystagmus; no hemianopsia; no limitation of visual fields. Vision: O. S., $\frac{2\,0}{2\,0}$; O. D., $\frac{2\,0}{3\,0}$. No objective sensory disturbance; knee-jerks elicited by reinforcement, left $>$ right; Achilles jerks lively; abdominal reflexes present, more active on the left side. Right ear defective to bone and aerial conduction.

Note September 10, 1900. Apathetic and somnolent; vertigo, headache, and vomiting; pupils equal and rather small. The direct and consensual reactions to light on the left side are sluggish and

[1] Patient was presented at the January meeting of the New York Neurological Society, and subsequently was reported clinically by Dr. Joseph Fraenkel in the Journal of Nervous and Mental Disease, 1899, p. 427.

at times absent; accommodation sluggish on both sides. Paresis of left external rectus; nystagmus; knee jerks are variable, diminished, sometimes absent; Babinsky phenomenon on the right; Achilles jerks lively on both sides; no objective sensory disturbances; abdominal reflex is absent on the right, feeble on the left.

Grew rapidly worse; progressive increase of the general cerebral symptoms. Died September 30, 1900.

Autopsy. Only the brain was removed. The skull, meninges, and vessels of Willis were normal. The pons contains a tumor within its substance on the left side throwing its normal contour into irregular nodulations. The left fifth nerve at its exit from the pons is compressed and flattened, while the left third, fourth, and sixth nerves stand in dangerous proximity to the growth.

Section through the hemispheres on a plane with the corpus callosum exposes a large tumor situated in the left optic thalamus and continuous with the infiltration in the pons. (Fig. 1.) It measures three inches in the longitudinal and two and one-half inches in the transverse diameter, and is rather sharply circumscribed from the surrounding brain substance by a lamellated periphery. It is of firm consistency and of a deep-red color (hemorrhagic). Some portions, chiefly on the periphery, are of a pinkish hue; others of a translucent appearance.

Hemorrhagic dots and minute areas of necrosis are scattered over the surface of section. The growth bulges into the third ventricle, the middle commissure of which is flattened out into a thin membrane, and encroaches posteriorly on the corpora quadrigemina.

The pineal gland is enclosed in a dense accumulation of connective tissue, but is otherwise normal. The hypophysis cerebri was normal.

On separating the medulla and the cerebellum at the foramen of Magendi a firm epiglottis-like prolongation appears. This on removal of the right lobe of the cerebellum is found to be the posterior tip of a large cyst, filling up and distending the fourth ventricle. (Fig. 1.) This cyst is one and three-quarter inches long and one and one-half inches wide. Its walls are thick, wrinkled posteriorly, smooth, and distended anteriorly and laterally. The outer layer of the wall strips readily, as in a fibrinous exudation, exposing a smooth surface with prominent and injected vessels beneath. It is accurately moulded to the interior of the fourth ventricle and on its basilar surface is firmly attached to the floor. The attachments of its upper surface to the cerebellum are slight and easily separated. The width of the vermis of the cerebellum is reduced by pressure to five-eighths of an inch and the lateral recesses appear as deep excavations.

The medulla is broader and flatter than normal. The posterior orifice of the aqueduct of Sylvius is free. The whole ventricular system of the brain is distended, more so on the left side. Numerous

ependyma granulations. A small portion of the cyst wall was cut away, exposing the interior, which was smooth, containing only a small quantity of clear fluid.

Microscopic Examination. The medulla, pons, left hemisphere of the cerebellum, and the enclosed cyst were hardened and embedded in bulk and cut serially. Approximately every tenth section was prepared and mounted by the Weigert-Pal method; half of these were subjected to a contrast stain (acid-rubrin).

Cyst. The cyst begins on a level with the tip of the calamus scriptorius, gradually increasing in size to the plane of the striæ acusticæ. From this level it becomes smaller and the walls thicker, dipping down into and becoming an integral part of the left side of the pons. On a level with the posterior corpora quadrigemina it terminates as a small triangular cleft, in the neighborhood of the left pyramidal tract.

From the tip of the thalamus to the striæ acusticæ the cyst wall has no connections with the floor of the ventricle, save to the ala pontis (ponticulus) on either side. From the striæ acusticæ to its termination the cyst wall is intimately associated with the neural structures of the pons.

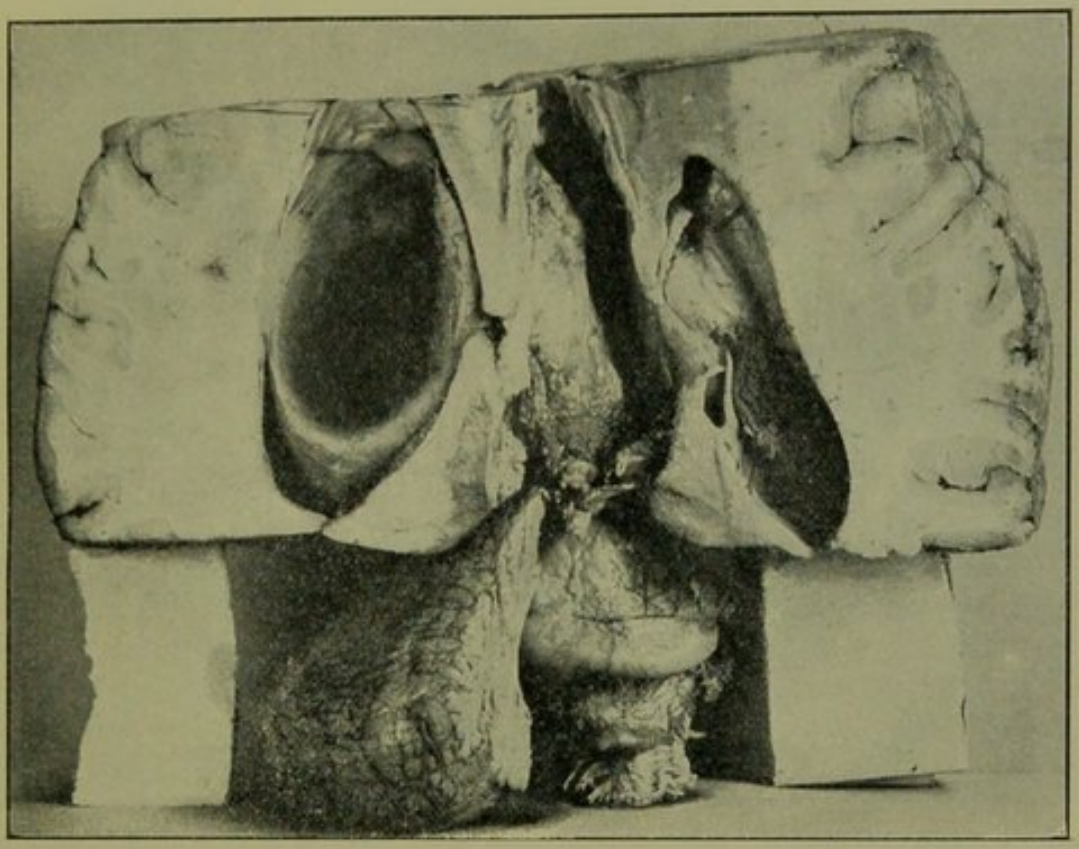
The nerve tracts form a decussation on its under surface, which pass upward into the cyst wall on either side. (Figs. 2, 3, and 4.) The cyst wall may be divided into three layers. An inner one is composed of glia; a thick middle layer of medullated nerve fibres which encircle the wall as a heavy band; and an outer layer in which the network of medullated nerve fibres is sparse, resembling gray matter. No ganglion cells could be demonstrated in the cyst wall, which possessed a rich supply of bloodvessels. No ependyma cells could be demonstrated lining the cyst wall. Here and there flat cells were seen, apparently of epithelial origin.

Cerebellum. The vermis and hemispheres of the cerebellum were compressed, and as a result showed changes in their configuration but without atrophy.

Medulla and Pons. The left pyramidal tract is small and pale. The lemniscus and fasciculus long. posticus are normal on both sides. The columns of Goll and Burdach are paler than normal, probably due to ascending intramedullary posterior root degenerations. (As the cord was not removed this cannot be definitely stated.)

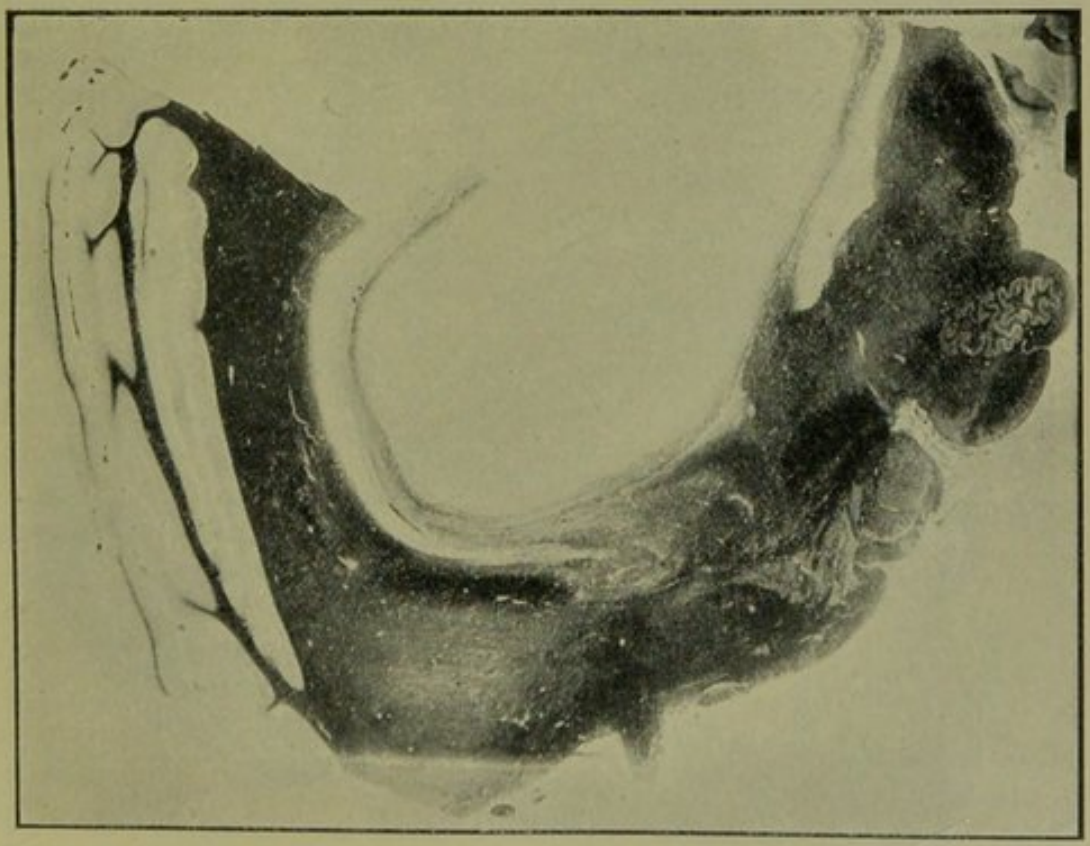
The medulla, except for a moderate flattening and lateral elongation, maintains its normal configuration to a level with the acoustic nuclei. Here the cyst becomes incorporated with the floor of the ventricle on the left side, nerve fibres decussating on its under surface and passing up into the cyst wall. This decussation is largely formed of the coarse fibres composing the middle peduncles of the pons, and increases considerably as the level of the superior olive is approached. Tegmental fibres, as well as a few fibres from

FIG. 1.



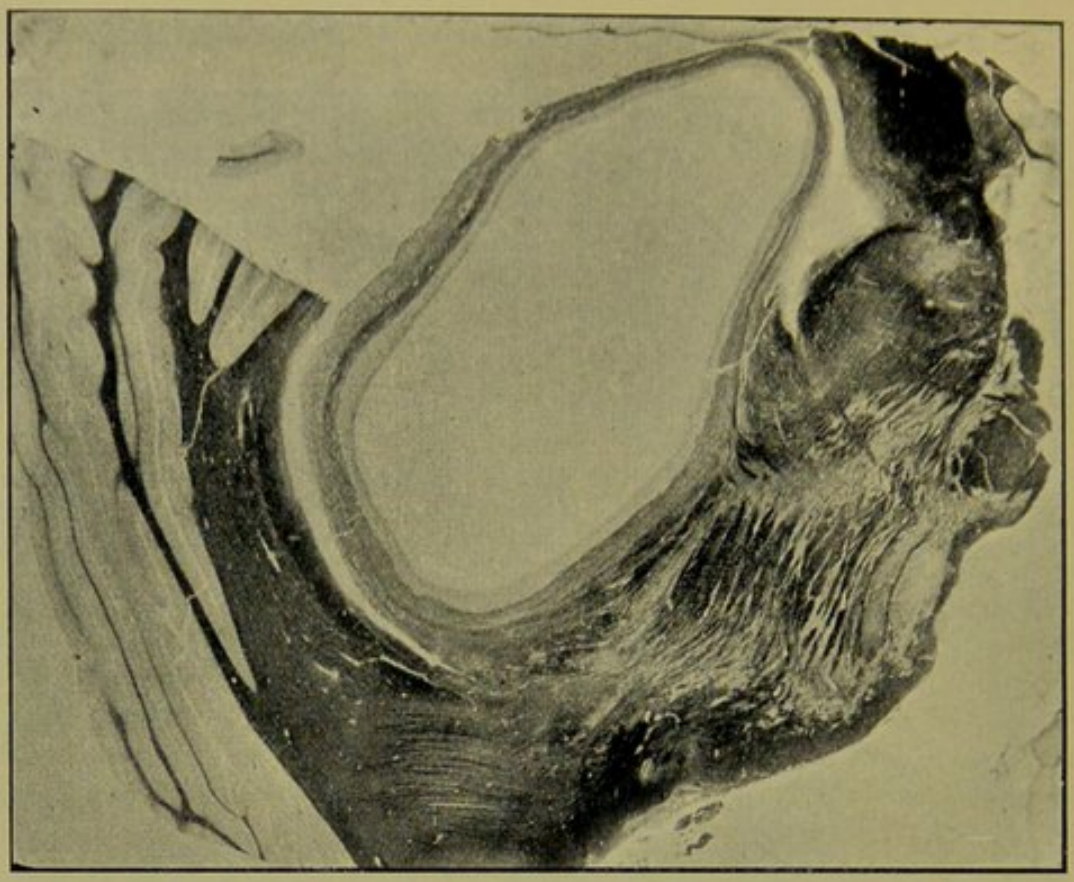
CASE I.—Tumor of left optic thalamus and congenital cyst distending the fourth ventricle.

FIG. 2.



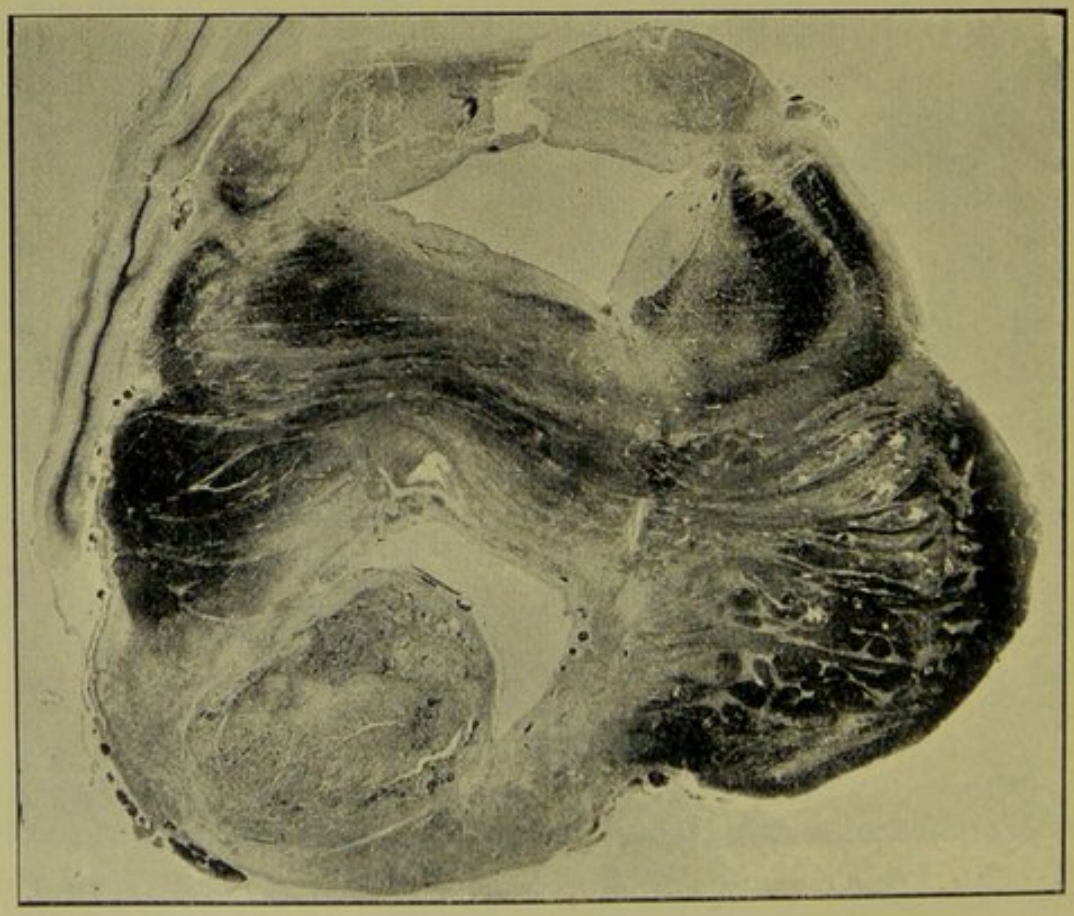
CASE I.—Weigert-Pal method. Level of striæ acusticæ. Showing intimate relation of cyst wall to floor of ventricle.

FIG. 3.



CASE I.—Weigert-Pal method. Level of the superior olive. Showing thick band of medullated nerve fibres in wall of cyst, continuous with the neural structures of the pons.

FIG. 4.



CASE I.—Weigert-Pal. Level of the posterior corpora quadrigemina. Cyst terminating in tumor as a narrow cleft.

the left median fillet and fasciculus long. posticus may be traced upward into the wall of the cyst.

At a level with the superior olivary body, where the cyst begins to sink gradually into the pons, the number of nerve fibres decussating on its under surface and passing up into its walls is very large. Most of these are derived from the pontine peduncle, so that these fibres on the right side of the pons are comparatively sparse.

Notwithstanding the distortions and changes in the configuration of this area, the important structures may all be discerned—*i. e.*, the superior olive, the ascending root of the fifth, lemniscus, and the fasciculus long. posticus.

On a level with the posterior corpora quadrigemina the cyst has tapered off to a small triangular slit in the region of the left pyramidal tract, and is here completely encircled by the tumor, which projects ventrally as a nodular swelling. (Fig. 4.)

Tumor. The pontine tumor and the large thalamus tumor are continuous and have the same histological peculiarities—*i. e.*, a polymorphous cell sarcoma, composed of round cells, spindle cells, and giant cells. The growth is extremely vascular, with numerous hemorrhages.

CASE II. *Summary.* A boy, aged seventeen years, previously healthy, following a head injury developed symptoms of brain tumor referable to the right crus cerebri. Weber's syndrome; left hemiplegia with extreme spasticity; complete right and partial left oculomotor paralysis; explosive laughter.

Autopsy. Glioma of the right crus cerebri extending posteriorly and infiltrating adjacent structures in the pons. A cyst of the fourth ventricle penetrating its floor, perforating the substance of the pons beneath and distinct from the aqueduct of Sylvius and appearing on the under surface of the right crus, terminating in the tumor mass.

Microscopically the cyst wall is composed of medullated nerve fibres and glia continuous with the nerve structures of the pons.

History. M. H., admitted to the Montefiore Home November 8, 1901; cash boy, aged seventeen years; family history negative. Previous history: was always a healthy boy, not subject to headaches or vertigo. (*Would frequently become nauseated while riding in street cars.*)

On June 7, 1901, he was hit in the back of the head by a swinging door, falling down a short flight of stairs. He received a scalp wound in the occipital region, but was not unconscious and in a short time was able to return home. In the street car, as had happened frequently before, he became nauseated and vomited. For a day or so he had moderate headache and in three days was able to return to his occupation apparently as well as ever.

One month later internal strabismus of right eye with ptosis

developed and was soon followed by a weakness and stiffness in the left arm. The weakness in the arm increased and became apparent in the left leg as well. He was inclined to somnolence and got very dizzy, but no headaches or vomiting. About the same time it was noticed that he laughed inordinately on slight provocation and sometimes spontaneously.

The patient was examined in the neurological department of the German Hospital and was referred to the surgical ward for operation, under the supposition that a post-traumatic cyst had developed at the base of the brain in the region of the right crus cerebri.

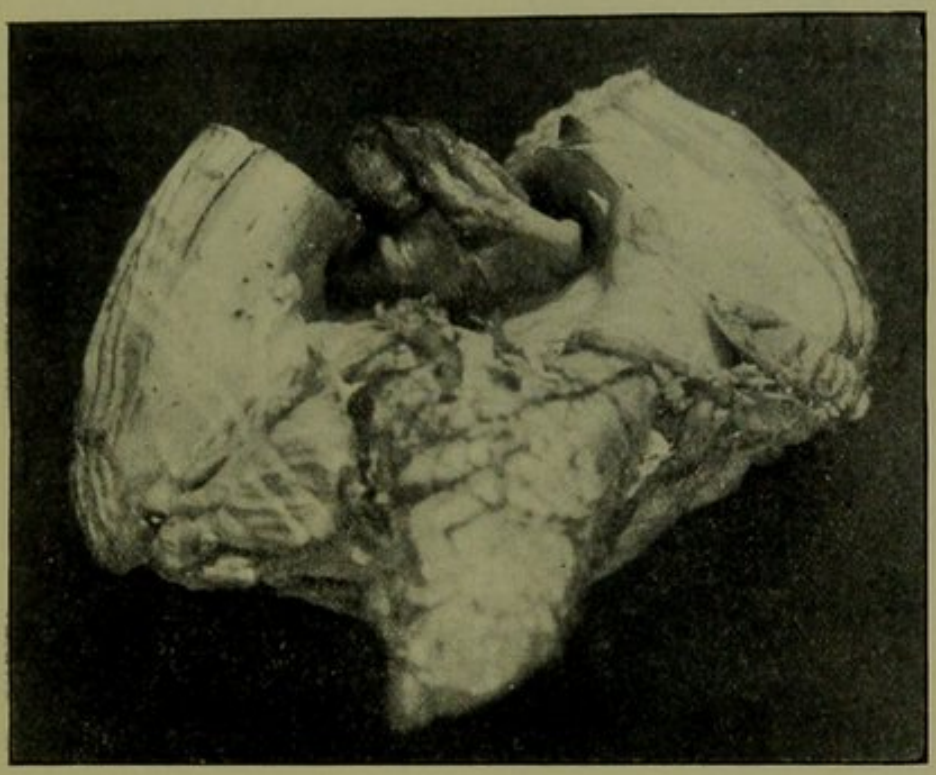
Operation August 28, 1901. An osteoplastic flap five inches in diameter was thrown back; the middle meningeal artery was resected and, after retraction of the temporal lobe, four exploratory punctures were made in the direction of the right crus, with negative results. Discharged November 7, 1901, unimproved.

Status præsens, November 9, 1901. Head dolichocephalic; face asymmetrical; teeth viciously implanted; torus palatinus; complete ptosis of right eye, partial of left; divergent strabismus on both sides; paralysis of the upward, downward, and inward movements of the right eye. The inward and downward movements of the left eye are limited in their excursion and accompanied by nystagmus. Nystagmoid twitchings were present in the right eye on attempted movement. The right pupil is widely dilated and fixed. The left pupil is normal in size and all reactions are present but sluggish. Paralysis of the left face, arm, and leg, with spasticity; clonus; Babinsky phenomenon; no objective sensory disturbances. Skin reflexes were absent on the left side; hearing, smell, and taste normal; no limitation of visual fields. Station; body is bent toward the left side with a tendency to fall in the same direction. There was uncontrollable and involuntary laughter from time to time, especially when starting to speak. Mentally, he was bright and clear; speech normal.

Optic Nerves. The vessels of the disks were congested and the upper margin was hazy; urine negative.

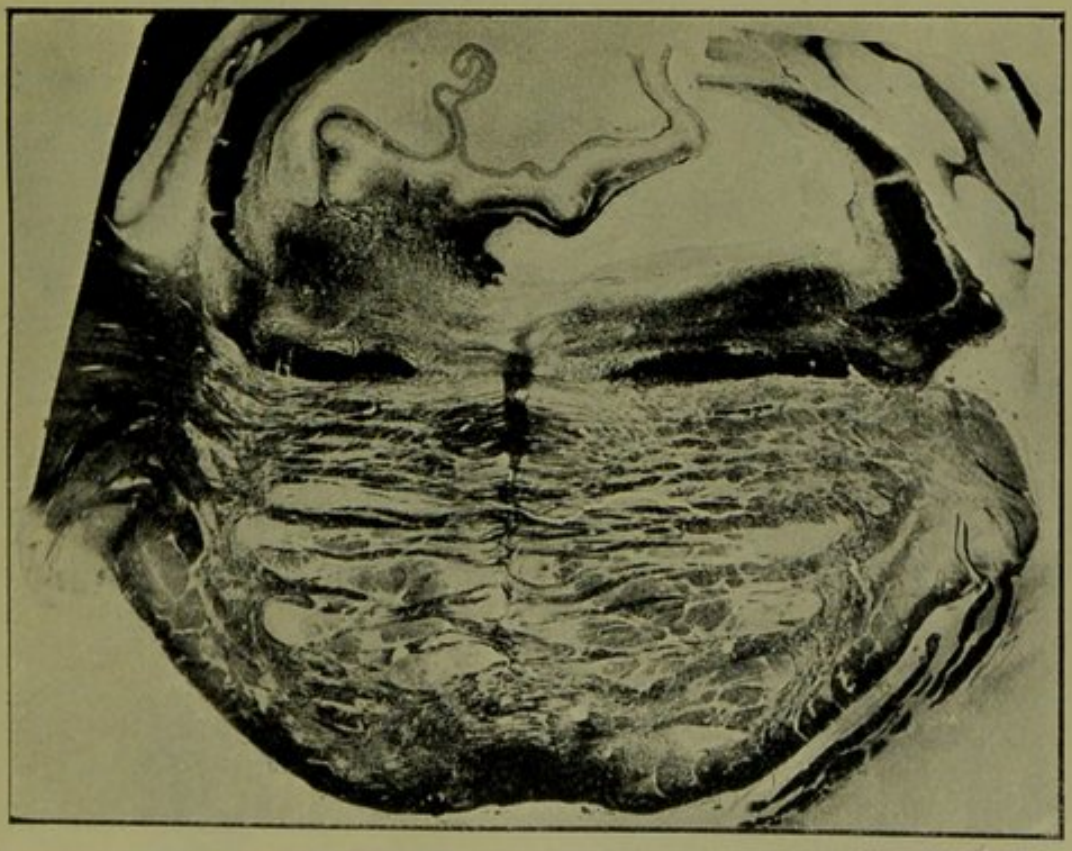
Note, March 12, 1902 (Dr. Abrahamson). General condition is much worse. His speech is indistinct. Any question or command induced uncontrollable laughter; no weeping. The right arm and leg are the seat of slow co-ordinated movements, which may be controlled on command. The cessation is only temporary, however. There is contraction of the left upper extremity, the clenched hand resting in contact with the shoulder; extensor contraction of the left lower extremity. The tongue deviates toward the left. There is complete ptosis of the right eyelid and partial of the left. The right eye is turned upward and slightly outward, the only movement preserved being feeble external rotation. The left eye is turned outward and downward, with preservation of the excursions downward and inward. Right pupil is large, with

FIG. 5.



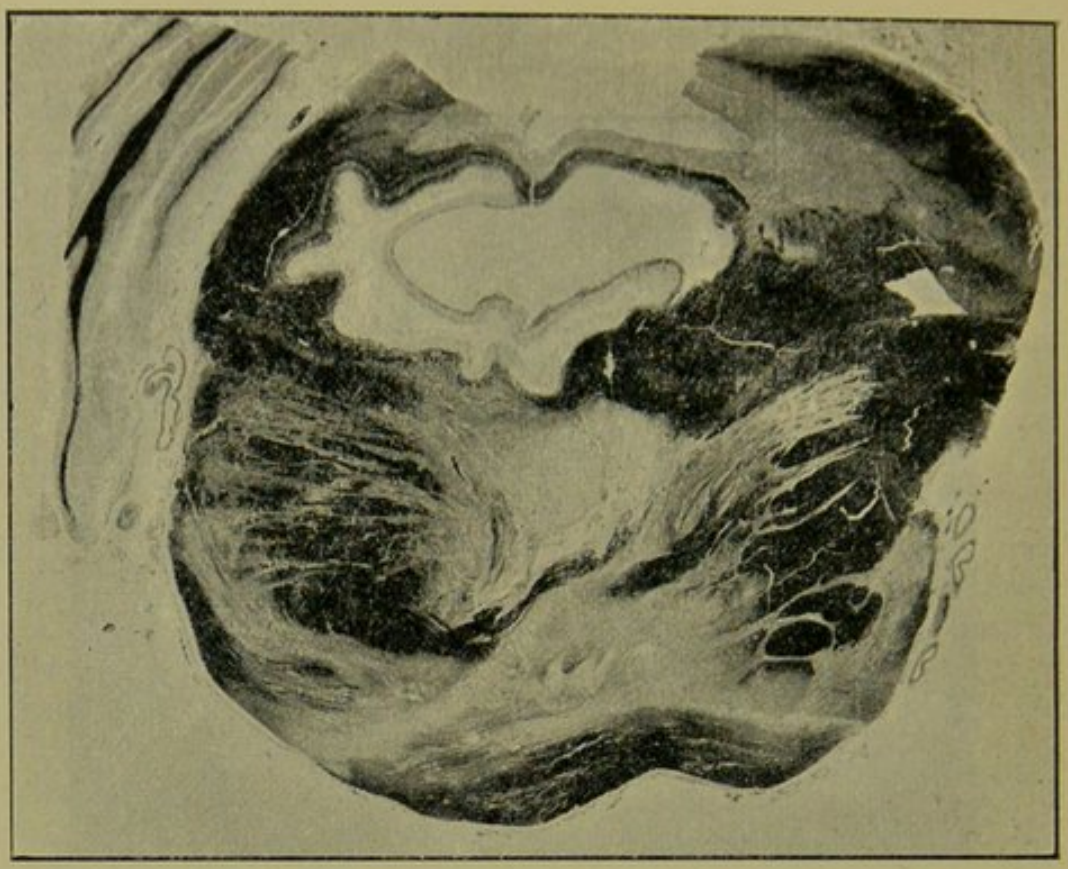
CASE II.—Vermis of cerebellum removed, exposing cyst in fifth ventricle.

FIG. 6.



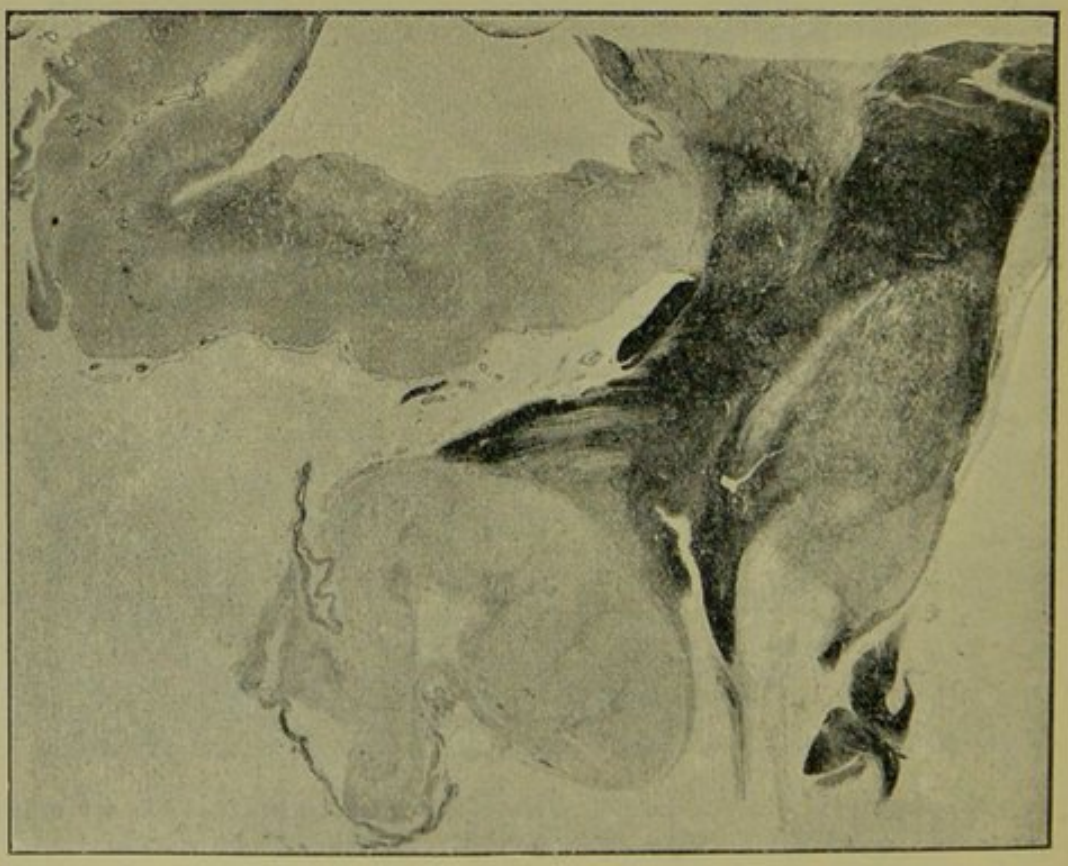
CASE II.—Weigert-Pal. Level of the fifth nuclei. Showing relation of cyst to floor of ventricle and thick layer of medullated nerve fibres in cyst wall.

FIG. 7.



CASE II.—Level of the posterior corpora quadrigemina. The cyst is beneath and distinct from the aqueduct of Sylvius, the roof of which is torn across. Cyst wall has collapsed on one side. Gliomatous infiltration of pons.

FIG. 8.



CASE II.—Weigert-Pal. Showing crus cerebri, optic tract and chasm; basal ganglia, internal capsule, and a portion of temporal lobe. Cleft in crural tumor representing termination of cyst.

irregular outline, and fixed. Left pupil is normal in size, with sluggish reactions. Other cranial nerves are negative.

Note, July 12, 1902. Emaciation is most extreme. The left hand and arm are in extreme flexion contracture, the hand resting on the acromion; left leg shows extension contracture. The left side of the body is cooler than the right. He hears and attempts to respond to some questions, but the response is inarticulate and indistinguishable. During the past four months there have been no paroxysms of forced laughter. He can tell the number of fingers held before either eye. The patient leads a purely vegetative existence; never speaks; sleeps most of the time; points to the mouth to indicate desire for food or drink. He recognizes members of the family and calls them by name. He drinks large quantities of water, and for three months has passed urine and feces in the bed, and if not watched will carry excrement to his mouth. The movements of the tongue and palate are slow and feeble, but there are no evidences of cranial nerve palsies other than those mentioned, with the exception of the motor portion of the right trigeminus nerve, the jaw deviating toward the right side. Pain impressions are equally keen on both sides of the body; no albumin or sugar in the urine.

Autopsy Cadaver is in a state of extreme emaciation; sacral and trochanteric decubitus; tubercular focus in left apex; central pneumonic area in the left lower lobe. Chronic cystitis; otherwise the examination of the thoracic and abdominal contents was negative. There was incomplete union of the osteoplastic flap at the site of the operation. The dura mater over this region is thickened and adherent and the inferior surface of the right temporal lobe shows old superficial foci of softening, with pial thickening. The sinuses of the dura and the vessels of Willis are normal; moderate internal hydrocephalus.

Description of Crural Tumor and Cyst. Springing from the right crus cerebri is a firm, nodular, grayish-white mass filling up the interval between the cerebral peduncles. The outer side of the growth is in close relation with the right optic tract; the inner side encroaches on the third nerve. The right third nerve is compressed and atrophic. The basilar surface of the pons and medulla oblongata with their respective cranial nerves are normal in appearance.

On splitting the cerebellum to afford inspection of the interior of the fourth ventricle, a cyst is disclosed springing from the anterior portion of its floor, to which it is firmly attached. (Fig. 5.) The posterior orifice of the aqueduct of Sylvius is free. The cyst is flattened and measures one and a quarter inches in its long and about five-eighths of an inch in its short diameter. The cyst wall is tough and dense and the surface is wrinkled. The interior is smooth and the only contents a clear fluid. The ependyma of the ventricle is dotted with numerous granulations. The cyst wall, at its attachment to the ventricular wall, dips down into the substance

of the pons, through which it passes beneath the aqueduct and appears on the inferior aspect of the right crus cerebri, where it stands in immediate relation to the tumor. The growth is firmly adherent to the superior wall of the cyst, which is collapsed in this region.

Microscopic Examination. A series of sections was prepared from various levels of the medulla, pons, and crus cerebri, including the cyst, according to the Weigert-Pal method with contrast stain. The medulla shows atrophy of the right pyramidal tract; the right spinal trigeminus root is normal. The central canal in the lower portion is obliterated and Helweg's tracts are degenerated. In the pons at the level of the fifth nucleus the cyst wall becomes firmly attached to it, forming an integral part of the floor of the ventricle. Numerous bundles of nerve fibres pass into and encircle the cyst wall, which contains a rich supply of medullated nerve fibres and glia cells. (Fig. 6.) The fasciculi long. postici are contained in the left wall of the cyst at its junction with the floor. The right fifth nerve root is atrophic. At this level no evidence of tumor infiltration is seen.

On a plane with the trochlearis nucleus the cyst is enclosed within the substance of the pons below the aqueduct, with which it has no communication, and a little to the right of the median line. (Fig. 7.) The configuration of the surrounding parts has been somewhat distorted; the important structures are, however, easily discernible—*i. e.*, the median and lateral fillet, the roots of the cerebral trigeminus and the trochlearis, and the trochlearis conjunctivum. The latter is displaced toward the left side. The dense bundles of the fasciculi long. postici are distributed over the superior portion of the cyst wall. At this level, below the lemnisci, infiltrating gliomatous tissue is already apparent.

Sections through the right crus cerebri include the thalamus, the lenticular nucleus, optic tract, and chiasm, and the inferior convolutions of the temporal lobe. (Fig. 8.) The normal outline of the crus is obliterated and is replaced by a tumor mass. On the under surface of and attached to the growth is the collapsed cyst wall. In the wall of the cyst medullated nerve fibres are still demonstrable. The wall is composed of a coarse, wavy glia tissue in which the fibrillar elements preponderate, and containing a sparse network of medullated nerve fibres. At several points lining the interior distinct areas of columnar epithelial cells are seen resembling the ependymal lining of the ventricles.

The tumor proliferation begins in the outer layers of the cyst wall and is here firm in texture, the spindle type predominating, forming interlacing waves and bands; numerous giant cells. In the periphery of the growth the proliferation is more typically gliomatous, merging gradually into normal areas. The vascular supply is rich.

The *cyst wall* may be roughly divided into two layers, the inner

composed of coarse glia and an external rich in medullary nerve fibres. The interior lacks a continuous epithelial lining, but a few remnants still persist. These consist of columnar cells, flat cells, and stratified layers of cells, the latter suggesting an epithelial metaplasia. No ganglion cells were observed.

Remarks. The occurrence in the brain of congenital cysts, while rare, is recognized in all classifications of the subject. They originate in offshoots of the primary cerebral vesicles. Bruns mentions, in connection with gliomata of the pons, the occurrence in the brain stem of minute cysts lined by ependyma and evidently separations from the primary neural tube. Stroebe demonstrated in gliomata of the brain minute cysts lined by columnar epithelium, probably sprouts of the primary cerebral vesicles.

In spinal cord pathology the relation of central gliosis, gliomata, and syringomyelia to developmental defects and embryonal rests of the central canal is fairly well established. In brief, abnormal offshoots and diverticula of the primary neural tube in any portion of its course may furnish the incentive to morbid proliferation.

Cysts of the size, location, and nature just described are unique in my pathological experience. A parasitic origin could be definitely excluded by the nature of the histological findings.

In favor of a congenital origin are:

1. The cyst wall throughout was composed of medullated nerve fibres and glia.
2. The intimate relations of the cyst wall with the adjacent nerve structures as it traverses the brain stem; the cyst is not intercalated, but forms an integral part of the pons.
3. Remnants of an epithelial lining.
4. The associated neoplasms.
5. The absence of cerebral symptoms preceding the development of tumor.

The gross anatomical relations of the cysts in my cases suggest an origin from that portion of the medullary tube engaged in the formation of the second and third primary cerebral vesicles.

The developmental changes in these vesicles, which form the mid-brain and hind-brain, are especially conspicuous and complicated. This portion of the fetal brain is wedged in between the head and neck bend, flaring out laterally to form the expansions of the floor of the fourth ventricle, corresponding to the lateral recesses.

The evaginations of the *Rauten lippen*, which develop subsequently, enfolding and covering in the ventral and dorsal zones of His, further complicate matters and would favor the occurrence of developmental defect. The frequency of gliomata in this region is well known.

In conclusion, I would emphasize the importance of subjecting the so-called old, sterile, *parasitic* cysts in the neighborhood of the ventricular cavity, to serial study and the Weigert method before excluding a congenital origin.