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CONGENITAL ORBITAL SARCOMA OF ENDOTHELIAL ORIGIN IN AN INFANT. OPERATION AND PRESERVATION OF GLOBE.*

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The literature of orbital sarcoma, especially in young subjects, is extensive, but congenital orbital sarcoma of endothelial origin is certainly not common. In a careful study of the literature on the subject I have not found a case, hence I report this one in the belief that it is unique. A great diversity of opinion exists on the subject of endothelioma, as regards not only its exact place in the oncologic system, but also its clinical and anatomical significance, leaving many important and numerous questions still unsettled. In many instances it is difficult to decide, from the structure of the tumor alone, whether it is an endothelioma, sarcoma, or adenoma. Some prefer to class them as sarcomata, while others describe them as embryonal adenosarcomata. The most striking feature common to this class of neoplasms is the combination of cellular and connective tissue elements, sometimes one predominating and again the other.

Of the clinical history of this case, all that is essential may be summarized as follows:

Examination: The patient, a male infant, was eight weeks old when first seen on September 12, 1902. The parents were healthy, and the mother, a primipara, said gestation and labor had been normal. At birth a swelling at the inner part of the left upper lid, producing a slight protrusion of the

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left eye, was noticed, and had about the same appearance then as it had when brought in for examination. The infant was well nourished, and physical examination showed no disease of the other organs. On examination, there was found beneath the upper lid of the left eye, between the internal angular process and the supraorbital notch of the frontal bone, occupying the inner wall of the orbit, a growth, the subconjunctival surface of which was about the size and shape of the end of a large peanut. The tumor was smooth, tense, not nodulated, and when steady pressure was made upon it no pulsation nor recession was felt. The integument of the upper lid was of natural color and unattached, but, owing to the presence of the neoplasm, the lids were not tightly approximated when closed. The left eyeball slightly protruded, and was displaced downward and outward. Globe movements were limited. There was no œdema of the lids and no injection of the ocular conjunctiva. The pupil was dilated and the iris reacted slowly to light. Ophthalmoscopic examination was negative.

Operation: Under chloroform anæsthesia, the bulbar conjunctiva was opened directly over the external surface of the tumor, the eyeball itself being left *in situ*, and carefully freed from its attachments for some distance. The growth was found to be firmly adherent to the sclera, and was separated by gentle dissection. The neoplasm was found to be encapsulated and a needle was introduced into the sac, but nothing could be withdrawn. This precaution was taken, with the idea that the mass might prove to be an encephalocele. In order to facilitate its removal, the internal rectus and superior oblique muscles were severed. The sac was then opened, and the contents, which resembled brain tissue, both in color and consistency, removed with a small, dull curette. After the sac had been emptied, the portion adher-

ing to the inner wall of the orbit was explored for a possible opening into the cranial vault, but none could be found. A ligature was tied around the proximal or attached end and the sac cut away beyond it. The severed muscles were approximated and sutured, and the opening in the bulbar conjunctiva closed. Two weeks after the operation, the little patient was taken to his home, in Wisconsin, with the eye slightly converged, but otherwise normal. I am informed that the child is healthy, and that the only evidence of an operation having been performed is the slight convergence which still persists. (September 15, 1903.)

Macroscopical Examination: The tumor was enclosed in a fibrous capsule, and measured approximately 10mm. in thickness, and about 2 cm. in length. It was grayish-red in color, friable in consistency, resembling normal brain tissue, and similar in shape to a sausage. It had no pedicle, but was attached at its proximal end by numerous bands directly to the periosteum of the inner wall of the orbit.

Histological Examination: The tumor was preserved in alcohol and imbedded in celloidin and paraffin. Sections were stained in hematoxylin and eosin, safranin, Van Gieson's stain, thionin, and Unna's polychrome methyl blue.

Viewed as a whole, with a low power (Fig. 1), the tumor is composed of a large number of cell masses, bounded by more or less complete anastomosing bands of fibrous tissue, so that an alveolar arrangement is produced. The alveoli, however, differ widely in size and in proximity to each other, and are not perfectly solid. On more careful examination it is clearly seen that the alveolar masses are formed by a somewhat complete double row of cells (Fig. 2), the long axes of which lie in line with the radii of the alveoli. The center of the alveolus within this stockade of border or covering cells is

occupied by similar cells, which are not regularly arranged nor so closely packed together. These cells, as a rule, are not firmly connected with the lining ones, but lie free in the open spaces. More commonly, however, the alveolar structure is not so manifest, for there are large areas in which the proliferating cells are so closely packed together that the alveolar arrangement is lost and only a narrow

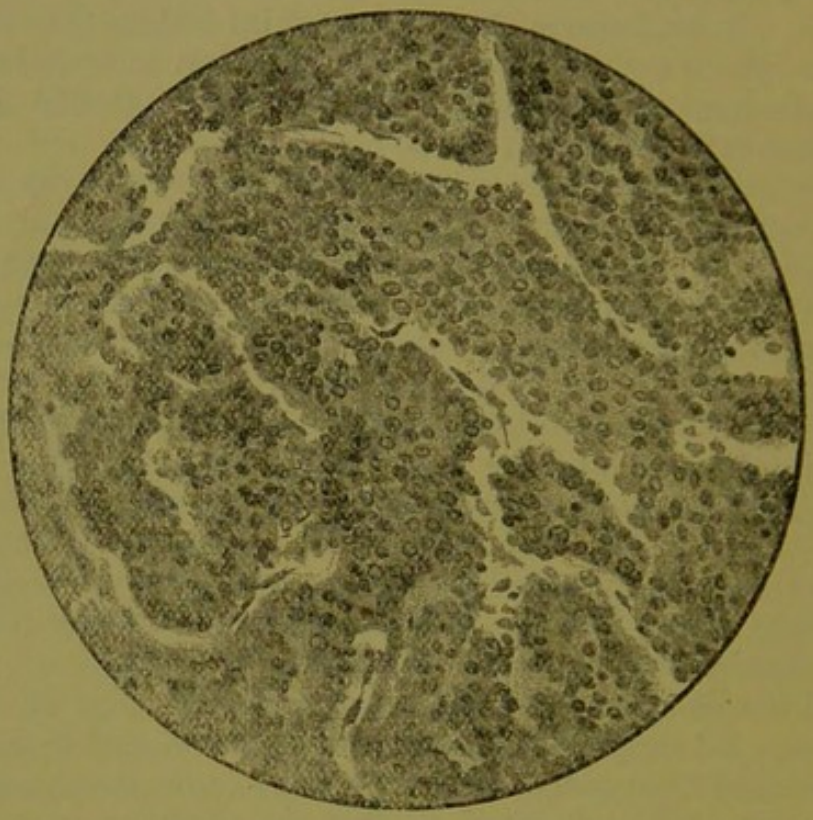

Fig. 1. Shows the alveolar arrangement and the large number of cell masses bounded by anastomosing fibrous tissue.

streak of stroma is seen to traverse its center. This probably does not indicate a marked deviation from an alveolar structure, which the tumor certainly possesses, but rather a packing together or a coalescence of smaller alveoli, which has occurred in the course of its growth. Coming to the cells proper of the tumor, one observes a striking monotony of appear-

ance. The cells in general are cuboidal, but when they are not closely packed together are decidedly cylindrical. The nucleus of the former is round, while that of the latter is oval and often elongated. All of the nuclei are vesicular, and possess a considerable amount of granular chromatin scattered in small masses evenly within them. One or more deeply-stained nuclei can usually be demonstrated.

Fig. 2. An alveolus under high power, showing its formation of a double row of cells.

The cells possess, in general, a comparatively small granular body of protoplasm, except the cylindrical cells, which have a more extensive cytoplasm. Here and there a cell will take a heavy stain, the nucleus being large, and presenting every appearance of being in a state of mitotic division. An attempt at bringing out karyokinetic figures proved unsuccess-

ful. This was probably due to the alcoholic fixation or to the slow growth of the tumor, as shown by the clinical course. In some of the alveolar spaces the cells have undergone degeneration. The cellular territories just described are bounded by tracts of connective tissue which, although not abundant, really form the structural framework of the tumor. This basement membrane, varying in thickness, forms a network with wide and irregular meshes in which the cellular elements lie. With Unna's polychrome methyl blue the connective tissue element stands out conspicuously, and while the cells within the alveolar spaces have but few fibers between them, it can readily be seen that at the circumference the covering cells are supported by bands of interstitial tissue carrying numerous blood-vessels. The stroma consists of loosely arranged fibers, with long, spindle-shaped or oval nuclei. Between these fibers are gaps and spaces which are occupied by red blood cells and occasionally by a few of the tumor cells. Within the stroma are seen vessels lined by a single layer of cells, identical with the tumor cells of the alveoli, representing a process of lymphangiectasis in which, as yet, no cell proliferation has occurred. Whether these be blood or lymph vessels, they seem to be conclusive proof that the growth is of mesodermic origin. The alveolar masses described above represent a proliferation of either blood or lymph vessel endothelium, and where they have coalesced, this proliferation has broken through its normal boundaries, in consequence of which a general invasion of the cells has occurred.

Those portions of the neoplasm that contain the typical and well-marked alveolar structure lined with the double row of cells, which are probably dilated lymph vessels (lymphactasia), might suggest adenoma were it not that the development of these tubular structures forms an almost complete and

uninterrupted picture from the beginning of proliferation in the small gaps or lymph channels up to the apparently atypic and irregular masses of cell material. From the foregoing, it seems probable that the tumor is a congenital alveolar sarcoma of endothelial origin.

Histogenesis: The probable histogenesis of the growth is as follows: Early during the invagination of the mesodermic portion of the primary optic vesicle in the formation of the primary optic cup, a diverticulum grew forward from it, and later on became cut off from the parent structure. The imprisoned mesodermic tissue surrounded by mesenchyma began to proliferate, leading to the structure which the neoplasm shows.

Remarks: In the classification of these tumors, although their exact place is still *sub judice*, their embryonic origin is a fundamental consideration. In the simplest embryonic form of cell growth, all cells are alike, and, according to Thiersch and Waldeyer, if endothelium is of mesodermic origin and as such belongs to the connective tissue group, then endothelioma must belong to the class of sarcomatous tumors. A fuller discussion of this question does not come within the scope of this paper.

In a tumor which is encapsulated and slow-growing, there is always some question as to malignancy. The accepted criterion of such tendency, namely, unbridled growth of embryonal cells and their recurrence after removal and tendency to form metastases, is sometimes at fault as to each and every detail. Embryonal-celled tumors are sometimes not malignant, as, witness teratomata, while, on the other hand, mature-cell tumors are sometimes malignant, as witness fibroids. In time, back of this criterion, which is really not cause but effect, will some time be found an etiological factor that will become the test of malignancy. Nevertheless, at the pres-

ent time any tumor composed of embryonal connective tissue cells and not inflammatory in origin must be called a sarcoma, whether encapsulated and slow-growing or the reverse.

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LITERATURE.

Burkhardt, *Beiträge zur klinischen Chirurgie*, October, 1902.

Minot, *Science*, March 29, 1901.

Volkmann, *Deutsche Zeitschrift für Chirurgie*, Bd. XLI.

Glockner, *Zeitschrift für Heilkunde*, Bd. XVIII, 1897

Lubarsch, "Ergebnisse d. Allg. Path.," Jahrg. I-II.

Marchand, Vert. der Deutschen Path. Gesellschaft, Bd. III, 1900.

Birsch-Hirschfeld, *Beiträge zur Pathologischen Anatomie*, Bd. XXIV, 1898.

Beard, *Central. f. Allg. Path. und Path. Anat.*, Bd. XIV, July, 1903.

Warthin, *Archives of Ophthalmology*, Vol. XXX, 1901.

Ribbert, "Lehrbuch," 1901.

Barbot, L., "Etude sur le Sarcoma de l'Orbite," Paris, 1877.

Veasey, *Medicine*, 1902.

Borrman, *Virchow's Archiv*, Bd. 151.

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