SPHENO-OCCIPITAL CHORDOMA

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Intracranial neoplasms originating from the vestiges of the notochord in the basisphenoid have been recognized since the time of Virchow, but they are decidedly uncommon. Mabrey (1), in a painstaking review of the literature through 1934, found only 47 cases of intracranial chordoma recorded. Since the appearance of his article approximately 24 additional cases have been reported. The subject of the present paper is one of a series of neoplasms of the central nervous system previously studied by the author (2). This tumor furnishes confirmation of former histological observations and also a few unusual features of interest.

REPORT OF CASE

The patient was a white woman thirty-five years of age, whose history up to her last illness showed nothing relevant. About two years before admission to the Boston City Hospital she began to be troubled with headache. This gradually increased in severity, and was complicated in the course of time first by fainting attacks and subsequently by increasing ataxia. For these disorders the patient was admitted for observation on the Neurological Service. Localizing symptoms and signs of increased intracranial pressure were not then apparent, and for a while she was regarded as a victim of one of the protean forms of multiple disseminated sclerosis. As time passed, however, no remission appeared, and the intracranial manifestations tended progressively to dominate the picture. It was decided, therefore, to explore the posterior fossa on the chance that a removable tumor might be found.

Accordingly a suboccipital craniotomy was done. The floor of the fourth ventricle was found crowded up against the cerebellum, and the brain stem rotated some 30 degrees to the right. No tumor was visible, and since it was evident that the tumor, if any, was inoperable, and the patient's condition was precarious, operation was limited to suboccipital decompression. She did not regain consciousness and died the following day.

Gross Pathological Observations: A complete autopsy was performed seventeen hours post mortem. The thoracic and abdominal viscera were essentially negative.

On exposure of the brain the pia-arachnoid appeared to be pressed nearly dry, and the convolutions over the vertex markedly flattened. The cerebellar hemispheres were superficially traumatized and softened, and herniated into the suboccipital bony defect. As at operation, the brain stem was found elevated and rotated to the right. On lifting the brain from the base of the skull, this rotation was seen to be due to displacement by a roughly cylindrical mass, slightly lumpy, 4 by 2 cm., pushing up vertically from the clivus and dorsum sellae.

On section of the brain, the lateral and third ventricles were found to be distended with clear fluid. The anterior portion of the fourth ventricle was obliterated by pressure. A sharply demarcated spherical mass of tumor, 1.5 cm. in diameter, was buried in the white matter of the brain stem at the junction of the peduncles and the pons. This mass on close inspection was seen to have been connected with the main spheno-occipital tumor by a slender pedicle that had been unwittingly broken off during removal of the brain from the skull.

The main tumor on the clivus was extradural, and its intracranial projection was completely covered by thinned-out dura. The tumor tissue was of uniform soft custard-like
consistence, gray to yellowish, with a few small spots of hemorrhage and ill defined soft, translucent mucoid areas.

The basal portion of the sphenoid and the adjacent occipital bone were found to have been eroded by the tumor, and the posterior clinoid processes were separated from their bony attachment. The pituitary fossa was not encroached upon, but the posterior wall of the sphenoidal sinus was almost completely destroyed. A rounded, soft mass of tumor tissue 1 cm. in diameter and smoothly covered with mucous membrane projected into the cavity of the sinus.

**Histologic Observations:** The tumor tissue was fixed in Zenker's fluid, 10 per cent formalin and 95 per cent alcohol. Appropriate portions were stained with eosin-methylene blue, phosphotungstic acid hematoxylin, Masson's trichrome stain, Foot's impregnation method for reticulum, orcein for elastic tissue, mucicarmine and thionin, Best's carmine for glycogen, and Sudan III for lipoids.

The tumor is composed of cords, masses, and concentric nests of cells embedded in a fairly abundant homogeneous or stringy mucoid matrix (Fig. 1). This matrix constitutes about one third of the total bulk of the tumor tissue. Occasionally ill defined nodules are seen formed of a peripheral zone of cells and a mucoid center into which project incomplete radial strands of tumor cells. These nodules are about 1 mm. in diameter.

The majority of tumor cells are of irregular polygonal shape with indistinct cell boundaries. Their vesicular nuclei are round or oval with a thin but heavily stained nuclear membrane enclosing a moderate number of diffusely scattered fine chromatin granules and one to three small round nucleoli. No mitotic figures were seen in any section examined. Vacuolated nuclei are very rare.

Multinucleated cells are fairly numerous, and are often syncytial masses of cytoplasm containing several nuclei of much larger than average size.

The cytoplasm of the tumor cells stains intensely with eosin. It usually appears lumpy or stringy, less commonly finely granular. In the majority of cells one or more apparently empty vacuoles are present in the cytoplasm. These vacuoles vary in size from small to
very large holes, the latter pushing the nucleus to one side and giving the cell a signet-ring appearance.

While the bulk of the tumor is composed of cords of epithelial-like cells, a gradual transition to small areas of spindle cells sometimes occurs. These latter areas are formed of closely crowded and compressed cords in which cells are more or less flattened, but still retain in part the cytoplasmic vacuoles seen in the epithelioid cells. These flat cells may also be distinguished from fibroblasts by the presence of tangled webs of fibrils in and upon the cell body in addition to the coarse longitudinal fibrils.

![Fig. 2. Physaliphorous Type of Tumor Cell, Showing the Peripheral Clear Zone Traversed by Radiating Fibrils. Phosphotungstic Acid Hematoxylin](image)

![Fig. 3. Normal Cell of Fish Notochord, Same Magnification as Fig. 2, Showing Radiating Fibrils. Phosphotungstic Acid Hematoxylin](image)

Scattered cells of the classic physaliphorous form are seen. These present a central cytoplasmic mass of stellate shape, often binucleated, from which radiate many fine fibrils crossing a peripheral clear zone to blend with the sharply defined cell wall (Fig. 2). The arrangement of analogous fibrils is shown with almost diagrammatic clarity in the normal notochordal cells of a fish (Catostomus) (Fig. 3). These fibrils are stained dark blue with phosphotungstic acid hematoxylin, in sharp contrast to the brownish red mucoid matrix.

The fibrils so clearly visible in this tumor (Fig. 4) seem to originate in the less differentiated cells as a condensation upon the surface of the cytoplasm of an intricate web of delicate intracellular fibrils. The coarser fibrils are wound about cells and often pass over the...
surface of adjacent cells. In cell nests the fibrils are frequently gathered into loose skeins in the center, and may stream thence out over attached cell cords. The fibrils are coarsest in spindle-cell areas, and in their most extreme development they form a dense parallel feltwork in which only scattered cell nuclei can be seen. The fibrils do not extend for any distance free into the mucoid matrix, and are everywhere sharply distinguished from it by their deep blue color. They usually remain within the condensed layer of mucoid material on the surface of the cell cords.

Alezais and Peyron (3) in 1920 made the first observation of fibrils in a chordoma, and Peyron subsequently published drawings showing the fibrils (4). The essential portion of the description by Alezais and Peyron is as follows: "The intracellular vacuoles rarely remain discrete, but fuse secondarily, leaving within the cell body several partitions of variable thickness. On close examination there is visible within these partitions a network of anastomosing fibrils staining black with iron hematoxylin and rose with the fuchsin of the trichrome. These are attached to and blend with the cell membrane." As vacuolization progresses cell boundaries disappear and there is formed a syncytial arrangement having "strands of chromophilic cytoplasm of filamentous structure within which are differentiated distinct fibrils of variable course, straight or spiral. This arrangement constitutes a special network of remarkable fineness, reminiscent of the histogenesis of the neuroglia. The fibrils form here and there curious arborizations in the form of a bouquet; ordinarily they are simply fused or interlaced. Their architecture is very similar to that of the glio-ependymal fibrils of Mallory."

The fibrils so described are apparently of the same nature as those in the present tumor. In this tumor also they are stainable by iron hematoxylin and the trichrome method, but are much more brilliantly and selectively shown by phosphotungstic acid hematoxylin. Their absence from the recorded descriptions of chordomas may in good part be due to the almost invariable employment of formalin fixation and hematoxylin-eosin stain, a method not suitable for their demonstration. They are not, however, constantly present, as is proved by the cases of Hass (5) and Furlow (6), in which the tissue was Zenker-fixed and properly stained. The chordomas on record vary considerably in degree of maturity and cellularity, and it seems probable that these fibrils, like fibroglia, may be abundant and stainable only at an intermediate stage of cell development.
True stroma in this tumor is practically absent. The cell masses float in a mucoid material that varies considerably from field to field in density and abundance. It is often vacuolated along the line of contact with cell cords. In this tumor at least the specific stains for mucin stained the matrix only weakly and in no very convincing manner, though mucin in simultaneously treated control sections of appendix stained clearly. The intracellular vacuoles, reported in some tumors to have contained mucin, are in this case uniformly empty to all stains save that for glycogen.

Glycogen is present in large amount, even though the tissue was fixed seventeen hours post mortem. Best's carmine method was controlled by digesting parallel sections for an hour in saliva before staining. By this means all but a few scattered granules of stainable material was dissolved out. With low power the glycogen appears to be fairly uniformly distributed in cells and in vacuoles in the matrix. High magnification, however, shows some cells to be crowded with granular masses, while adjacent cells are empty. Vacuoles in both cells and matrix usually contain many granules dusted over their inner surfaces. The very few vacuolated nuclei that can be located do not contain glycogen.

Vessels are scanty and irregular in shape, with imperfectly formed walls. About them is a sparse infiltration of lymphocytes and plasma cells. Occasional large patches of hemorrhage are present in the vicinity of the vessels, and in the adjacent tumor are scattered pigment-filled monocytes. In the walls of these few vessels and in the dura capsule is seen the only demonstrable elastic tissue. Foot’s impregnation for reticulum and collagen shows connective tissue likewise to be limited to the sparse vessels and the capsule. By this method the mucin is colored brown, while the many cellular fibrils are quite invisible. The tumor contains no material stainable with Sudan III.

Beneath the dural capsule are a few spicules of bone being destroyed by the advancing tumor. At some points the bone disappears by surface erosion leaving a sharply defined edge. In other fields cords of tumor cells seem to force their way into the bone canaliculi and distend them. Simultaneously the ground substance of the bone gradually loses its dense and deeply staining character, blends with the mucoid tumor matrix and is lost (Fig. 5).

**Summary**

The clinical history and post-mortem findings in a case of spheno-occipital chordoma occurring in a thirty-five-year-old female have been described.
Microscopically the tumor presented the usual features of a cellular and locally malignant chordoma. In addition there were present many cellular fibrils staining like fibroglia, which apparently have been described but once previously.

References