Intracranial chordoma in infancy

Case report

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The authors describe a case in which a large clival chordoma became symptomatic in a 10-month-old infant, and was surgically excised and irradiated at 21 months. Two years later there is no evidence of recurrence.

KEY WORDS • chordoma • clivus • brain scan

Clival chordomas are uncommon at any age. By 1967 less than a dozen cases had been reported in children who had not yet reached puberty. We believe the case presented here is in the youngest patient reported to have this problem.

Case Report

A previously healthy, 10-month-old boy developed a right sixth nerve paresis. By 16 months he also had developed left sixth and third nerve pareses, and at 21 months he stopped walking and became unduly irritable. We first saw him when he was 23 months old.

Examination. The patient had an enlarged head, with a 54 cm circumference. The anterior fontanel was open and tense. He was totally blind, with fixed pupils and bilateral optic atrophy. The legs were spastic, the deep tendon reflexes were hyperactive particularly on the left, and the plantar response was extensor on the right. Plain skull films showed widely separated sutures. The brain scan uptake was increased in the left parasellar area and in the posterior fossa (Fig. 1 left). An air encephalogram showed hydrocephalus with posterior displacement of the fourth ventricle and elevation of the left temporal horn. Carotid and vertebral angiography confirmed the presence of a large mass in the left anterior, middle, and posterior fossae (Fig. 2 left and right).

Operations. At frontotemporal craniotomy, an enormous extradural tumor was found underneath the temporal lobe and enveloping the optic nerves. The bulk of the tumor in the middle fossa was removed except for some that had invaded the cavernous sinus. One week after operation, because the hydrocephalus was progressing, a right ventriculoperitoneal shunt was inserted. The child's skull was irradiated with 5264 rads bilaterally. One year later he required burr hole drainage of a large subdural hematoma over the right cerebral hemisphere.

Now, at 4 years of age, the spasticity has gone from the legs and his gait is normal. His head circumference remains unchanged at 53 cm, but he is still blind and his pupils react only sluggishly to light.
Histological Examination. The surgical specimen consisted of 3 gm of soft gray-red lobulated gelatinous tissue. Sections stained with hematoxylin and eosin showed sheets of cells with no architectural arrangement and no fibrous tissue septae. Most of the cells were large, with well-defined margins and small, dark nuclei. The cytoplasm was finely granular in some but physaliphorous in others. No lakes of mucinous material could be found. Many cells had more than one nucleus, and a moderate degree of variation in cellular size and shape was evident (Fig. 3). Staining reactions with alcian blue, periodic acid-Schiff (PAS), and mucicarmine indicated the presence of an intracellular mucopolysaccharide. Extracellular material was scanty, but similar.
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Under the electron microscope the majority of the cells were characteristically physaliphorous. Their cytoplasm contained numerous smooth-walled vesicles of various sizes, some of which held amorphous granular material (Fig. 4). In some cells the Golgi apparatus was prominent and associated with numerous coated vesicles. Abundant glycogen, clumps of granulofibrillar material, and free ribosomes were scattered through the cytoplasm. Bundles of fibrils could be identified clearly. Dilated cisternae of the endoplasmic reticulum were prominent. Many apparently normal mitochondria were surrounded by one or two rows of neatly arranged rough endoplasmic reticulum. Interdigitating plasma membranes and numerous pinocytotic vesicles were seen. Extracellular granulofibrillar material similar to that seen intracellularly was also present. Only a few stellate cells were found.

Fig. 3. Light micrograph of tumor shows cellular pleomorphism; note the cell at lower left with two nuclei. The cytoplasm is finely granular and some cells contain small vacuoles. H & E, × 394.

Fig. 4. Electron micrograph of physaliphorous cells shows clumps of granulofibrillar material (GF), Golgi apparatus (G) at upper border, and smooth-walled vesicles (V) throughout the cytoplasm. × 35,000.
Discussion

Chordomas or tumors of notochordal remnants can occur at any level of the craniovertebral axis. Although early reviews stated they were more common in the sacrococcygeal region than at other sites, recent reports indicate that they are just as common inside the cranium. Here, they account for less than 1% of all intracranial tumors; in all locations chordomas are twice as common in males as in females.

What initiates growth of these tumors is unknown, but the higher incidence at the poles of the vertebral axis may be related to the complex anatomical developments which occur in these regions during the embryological period.

Although histologically benign and characteristically slow growing, chordomas can metastasize widely. Intracranial chordomas tend to produce symptoms in later life, but those in the spheno-occipital region are often discovered before the patient reaches 20 years of age. Even in this site, however, they are rare before puberty.

The similarity of the clinical and roentgenographic manifestations of intracranial chordomas and other cranial neoplasms makes correct preoperative diagnosis difficult. However, chordoma should be considered in the differential diagnosis of all midline tumors at the base of the brain.

Our patient first displayed signs of neurological disease at 10 months of age, but the tumor may have been present long before that. It may even have been congenital. Since chordomas are usually discovered late in life they are presumed to be slow growing. In this infantile case the chordoma was extensive. Despite the absence of stellate cells, the diagnosis was made clear by the presence of the physaliphorous cells, with an ultrastructure resembling that described in other reports. Usually, intracranial chordomas do not respond well to surgical excision or radiation. Total removal of the tumor, particularly such a large one, is difficult. Nevertheless, our patient seems to have had a good short-term response to radiotherapy.

Brain scans (Fig. 1 right) and pneumoencephalograms at 24 and 26 months respectively after treatment show no evidence of residual tumor, and aside from blindness, the child is well.

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References


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