Chondroid chordoma

Case report

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A case of the very rare chondroid variant of chordomas at the base of the skull is presented. The characteristic presentation, radiological and computerized tomographic appearance, findings at surgery, pathology, and treatment of this lesion are discussed. Neurosurgeons should be aware of this variant, due to its predilection for occurrence at the base of the skull and its more favorable prognosis compared with that of typical chordoma.

KEY WORDS • chondroid chordoma • chordoma • brain tumor • base of the skull

CHORDOMA is a rare tumor appearing in close relation to the axial skeleton and believed to arise from notochordal remnants. Although they are histologically benign and slow-growing, chordomas may be considered as possessing malignant potential due to their occurrence in critical locations, their locally aggressive behavior, and their high recurrence rates.14

The histological resemblance of chondroma and chondroid variants of other neoplasms has been appreciated since Virchow's study15 and has contributed to the difficulty in differentiating the two classes of neoplasia. Both Stewart and Morin12 and Mabrey8 have reviewed these historical aspects, and their respective works should be consulted for further details. The entity known as chondroid chordoma was first described by Heffelfinger, et al.,1 in 1973. This variant must be differentiated from both chondrosarcomas and chordomas, since it has a predilection for occurrence at the base of the skull,4,6,11 a proclivity to invade bone and entwine itself around vital structures,12 and a more prolonged and benign course.4,6,11,12 The current case report is presented in order to provide neurosurgeons and others who deal with tumors at the base of the skull an appreciation of these subtle but exceedingly important differences.

Case Report

This 26-year-old woman was evaluated for chronic intermittent generalized headache, which had become more persistent in recent months and which was associated with early morning nausea and ptosis of the left eye. Her general health was good.

Examination. The patient was a pleasant, cooperative, alert, and oriented woman who complained of headache. Her pupils were equal and reactive. Her funduscopic examination was benign. A mild ptosis of the left eye was noted, but there was no other cranial nerve deficit. The remainder of her neurological examination was unremarkable.

Plain skull films showed mild erosion of the clivus, with thinning of the dorsum sellae and no intracranial calcifications (Fig. 1). Computerized tomography of the head, with and without contrast material, revealed a large enhancing tumor mass in the left parasellar and retrosellar regions, with suprasellar extension into the medial aspect of the left middle fossa (Fig. 2 left). Cerebral angiography revealed minimal vascularity associated with the tumor identified on the CT scan (Fig. 3).

Operation. The tumor mass was approached through a left temporal craniotomy, with particular attention given to preserving the venous drainage of the temporal lobe. As the temporal lobe was elevated, the tumor was identified in the medial aspect of the left middle fossa. The tentorium and middle fossa dura were stretched over the mass. With the aid of the carbon dioxide surgical laser, a limited incision was made through the tentorium and middle fossa dura overlying the mass. Elements of the left trigem-
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Fig. 1. Plain skull film showing thinning of the dorsum sellae.

Fig. 2. Computerized tomography scans with contrast enhancement. Left: Preoperative scan revealing an enhancing tumor mass in the left para- and retrosellar regions, with suprasellar extension into the left middle fossa. Right: Postoperative scan revealing cast from the tumor and near total removal of the tumor mass.

The tumor mass appeared to be a chordoma, showing an admixture of vacuolated cells with a mucinous matrix and chondroid elements (Fig. 4).

Postoperative Course. Following surgery, there was numbness of the left side of the face in all divisions of the trigeminal nerve, and ptosis of the left eye. These abnormalities progressively cleared over a period of 7 to 10 days, with good wound healing and no other neurological deficits. A postoperative CT scan of the head with contrast enhancement several days after surgery revealed nearly total removal of the tumor mass (Fig. 2 right).
Fig. 4. Photomicrographs of the tumor specimen. Left: Physaliferous or "bubble-bearing" cells are seen (arrows). Alcian blue & PAS, × 230. Right: Section showing the junction of cartilaginous and chordomatous tissue. Note the transition from chordoma (A), to a chondroid tissue (B), to a more mature cartilage (C). Alcian blue & PAS, × 115.

Discussion

The incidence of chordoma is extremely low, especially if one accepts Ribbert's 10 autopsy finding of a 2% incidence of notochordal ectopia in the basiocciput. 12 The chondroid variant is even more rare, with an incidence of around 15% of recently reported cases of chordoma. 6,11 Of those chordomas found at the base of the skull, almost one-third were diagnosed as the chondroid variant, with 88% of all chondroid chordomas occurring in this area.

In contrast to the 3:2 male predominance reported for all chordomas, 12 there is a slight female predilection in the chondroid variant. 5 Although chordomas at the base of the skull were found more often in patients between the third and sixth decades or in the fifth decade, 4 73% of patients with the chondroid variant presented before the age of 40 years.

Among patients with either typical chordomas or the chondroid variants located at the base of the skull, there were apparently no differences in signs and symptoms or in duration prior to treatment. 6 The symptoms present at the time of diagnosis could generally be attributed to the size and location of the tumor. From 60% to 90% of patients presented with diplopia, representing a loss of function of the oculomotor or abducens nerves. Progressively severe frontal or occipital headaches were slightly less common; between 50% and 75% of patients presented with this complaint. 4,6 Papilledema was a decidedly uncommon finding. The duration of symptoms was usually 6 months to 3 years, 6 with one study reporting a mean duration of 1 year. 4

The classic radiographic finding in chordomas at the base of the skull was an expansile osteolytic lesion, 6 with bone destruction present in 75% of cases, 13 most commonly in the clivus and sella turcica. 14 The presence of calcification is important, since chondroid chordomas were most likely to calcify. Arteriograms typically show avascularity. 12 The CT scan provides a graphic representation of the tumor and its accompanying bone erosion, and demonstrates any associated hydrocephalus with relatively little increased risk to the patient. It does not, however, provide the information regarding vascularity that is demonstrated by angiography.

Based upon our review, gross examination of the tumor tissue provides little to aid in the differentiation of the chondroid variant from the typical chordoma. Chordomas are characteristically not encapsulated, gelatinous, lobulated, and grayish in color, with areas of hemorrhagic necrosis. 1,6,12 The presence of flecks of bone in the outer, advancing margins of the tumor may aid in its identification, since calcification was much more likely in the chondroid variants at the base of the skull. 6 Histopathological examination most often reveals an admixture of the physaliphorous (blister-bearing) or vacuolated cells, a mucinous matrix, and a tendency for the tumor cells to grow in cords typical of chordomas and cartilaginous or chondroid elements. This admixture may range from small scattered areas of hyaline cartilage in a chordoma.
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background to the reverse in which the chondroid component dominates. Areas in which chordomatosus and chondroid elements merge should be identified if possible. Crawford has advocated the use of phoshotungstic acid hematoxylin (PTAH) and reticulin stains to facilitate histological screening of these tumors. A chondroid matrix was thought to stain well with PTAH and reticulin stains, whereas a chondroid matrix did not. Batsakis and Kittleson stated that these stains were not helpful in the differentiation of these tumors, and this has since been confirmed. Furthermore, although the ultrastructure of the typical chordoma was described by Friedmann, in 1962, the ultrastructural features of the chondroid variants which would aid in differentiation have not been delineated.

Extensive surgery combined with high doses of radiation has been advocated as the management of choice for chordomas. Heffelfinger, et al., observed that patients with the chondroid variants usually fare better than those with typical chordoma, and found a statistically significant difference in the survival times of the two groups. The tumor is only moderately radiosensitive, and a review of the efficacy of radiation therapy as a primary modality revealed that doses of irradiation of 6000 to 8000 rads provided only a 26% 5-year cure rate, with doses of greater than 8000 rads providing only a 6% cure rate at 5 years. In contrast, when irradiation was combined with surgery, median survival increased from 17.4 years in patients treated with surgery alone to 24.9 years in patients receiving combined therapy.

In view of these high doses of radiation, the limited radiosensitivity of the tumor, and the typical proximity of the tumor to the brain stem and other vital structures, decisions to use adjuvant radiation must be made on an individual basis.

Conclusions

The neurosurgeon should be aware of this chondroid variant for a number of reasons. First, it has an apparent predilection for occurrence at the base of the skull. Second, while there are no distinguishing characteristics in the clinical presentation, careful attention to the radiographic presentation may aid in its identification. Finally, its markedly improved prognosis when compared to typical chordomas at the base of the skull should become a consideration in the management of patients presenting with the chondroid variant.

In summary, there are no pathognomonic features of chondroid chordoma. The presence of calcification on plain radiographs of the skull suggests this variant. The diagnosis is further aided by demonstrating the histopathological admixture of cartilaginous and chordomatosus tissue biopsied at surgery. Treatment should consist of as complete a surgical removal as possible, with or without subsequent adjuvant high-dose radiation.

References


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