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Removal of Clival Chordoma in an Adolescent Thorough Combned Pterional Transsylvian and Anterior Temporal Approach

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ABSTRACT

Extensive and aggressive surgical removal is treatment of choice for patients who have chordomas of the cranial base. Well-developed microsurgical techniques, as well as good surgical judgment learned from experience are essential to avoid major morbidity. A 14-year-old female presented with progressive headaches and diplopia for three months. Cranial magnetic resonance imaging demonstrated a well-circumscribed mass in the clival region. The mass was totally excised via right combined pterional transsylvian and anterior temporal approach (+ orbitozygomatic osteotomy). The tumor was located extradurally. The resected tumor had the typical histological and immunohistochemical characteristics of chordoma. No radiation therapy or chemotherapy was administered.

KEY WORDS: Chordoma, Clivus, Combined approach, Transsylvian approach, Anterior temporal approach.

INTRODUCTION

Chordomas are midline tumors of the central nervous system that arise from remnants of the primitive notochord 6,9. While the majority of chordomas occur in the sacral region, more than one third arise in the skull base (5,6). The vast majority of these tumors are extradural and associated with extensive bone destruction (5).

Intracranial chordomas usually occur in adults and are extremely rare in children. Fewer than %5 of chordomas are diagnosed in patients 20 years of age or younger.2 Long survival rates have been associated with more extensive tumor removal. Complete removal of clivus chordoma is extremely difficult to achieve, even with extensive surgery, because a few tumor cells are frequently left behind in the bone and dura. Adequate exposure of the lesion is therefore important. Many surgical approaches to the clival regions such as anterior basal, lateral basal or combined have been described (1,11). In the literature, some modifications have been described for the traditional approaches to improve the visualization of the tumor, facilitating its total removal, thus decreasing although not eliminating the morbidity due to brain retraction. In our case, we chose combined pterional transsylvian and anterior temporal approach and right orbitozygomatic osteotomy (OZO).

We present a rare case of chordoma in an adolescent patient with a large mass located in the clival region. In this case, we performed combined pterional transsylvian using the anterior temporal approach and OZO.

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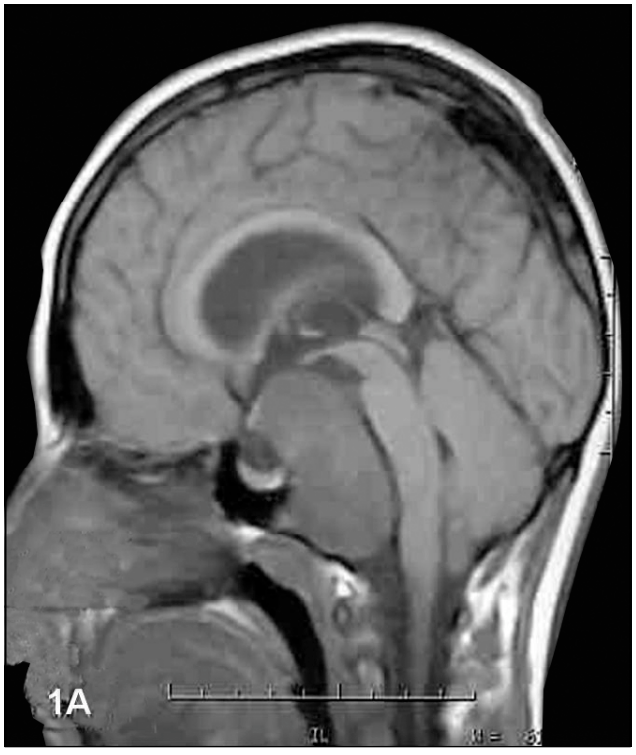
CASE REPORT

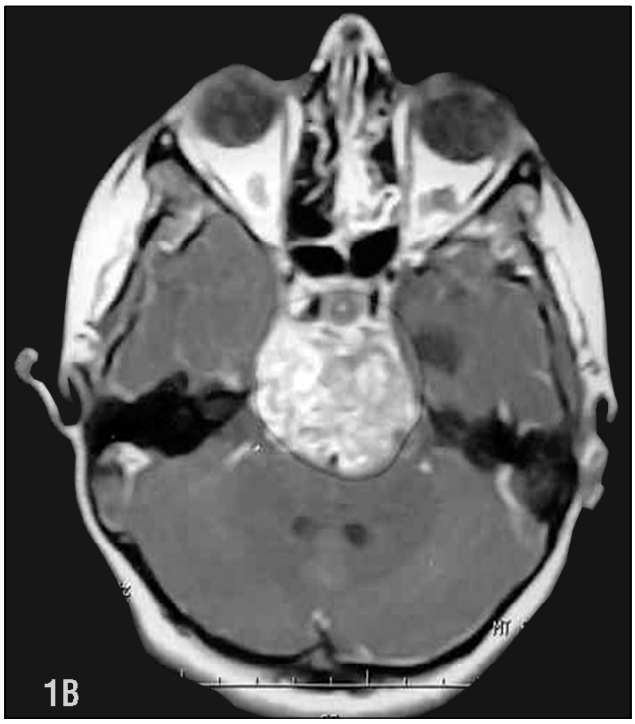
History. A 14-year-old female presented with progressive headaches and diplopia for three months.

Neurological Examination. The remainder of the neurological examination was nonfocal. Fundoscopy revealed bilateral optic atrophy and macular oedema.

Radiological Findings. Cranial magnetic resonance (MR) imaging demonstrated a well-circumscribed mass in the clival region. The mass was iso-intense on T1-weighted and showed a high-intensity area on T2-weighted MR images, and was heterogeneously enhanced by the intravenous infusion of gadolinium-diethylenetriamine penta-acetic acid (Figure 1). The tumor was compressing the brainstem from the medulla oblongata to the midbrain and was not contiguous with the clivus. The basilar artery was observed to be encased and displaced posteriorly. Vertebral angiography showed that the basilar artery was displaced posterolaterally to the left side and the right vertebral artery was stretched. However, the tumor was avascular (Figure 2).

Operation. A right fronto-temporo-parieto-sphenoidal (large pterional) free bone flap was elevated. A right unilateral orbitozygomatic osteotomy (OZO) from the supraorbital notch to the





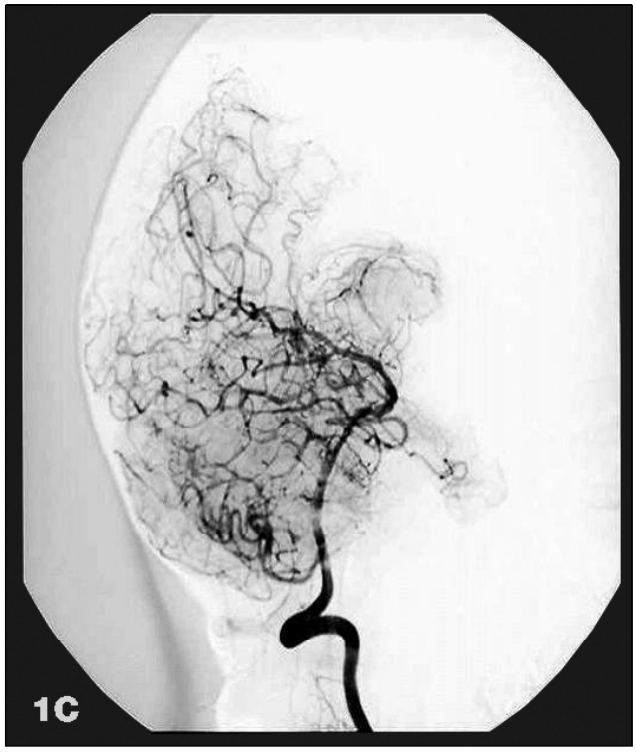


Figure 1: A (Left). Presurgical MR images, sagittal view, showing a mass within the clival region. The mass was iso-intensite on T1-weighted MR images. **B** (Middle): Presurgical MR images, axial view. The mass was heterogeneous enhanced by the intravenous infusion of gadolinium-diethylenetriamine penta-acetic acid. **C** (Right): Presurgical vertebral angiography showed that the basilar artery was displaced posterior and the tumor was avascular.

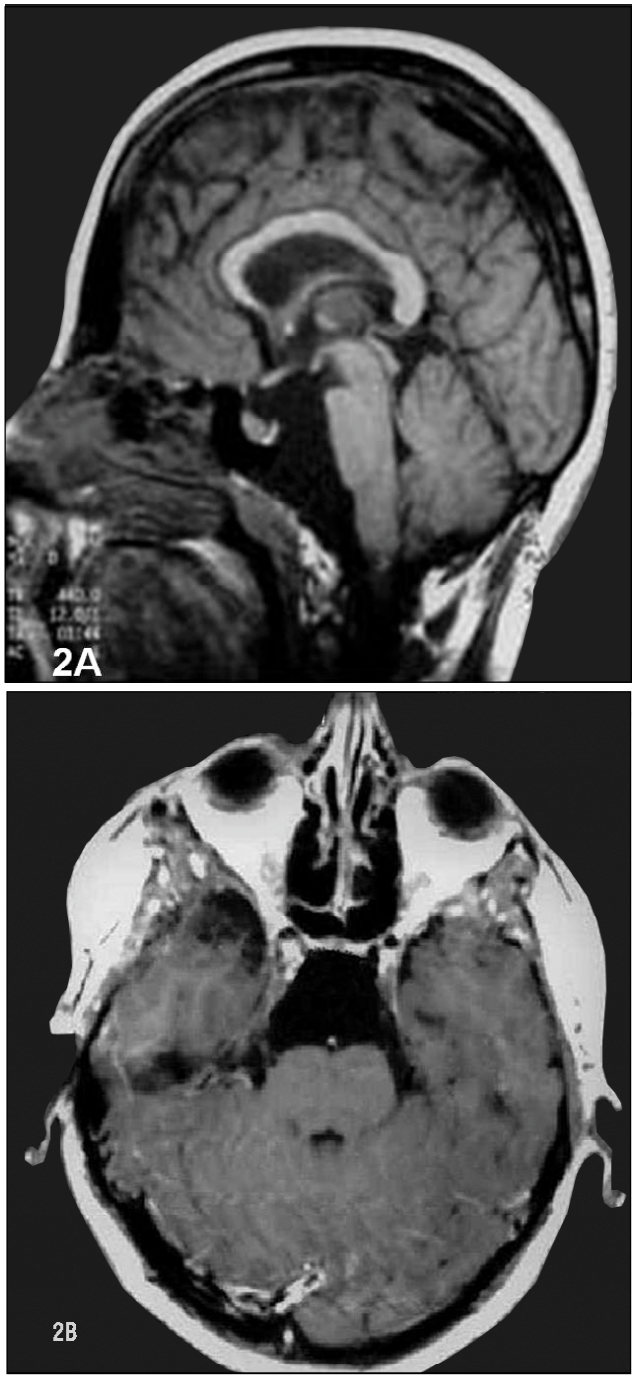


Figure 2: A: MR images, sagittal and axial view. **B**: Early postsurgical T (inf/1/weighted) image after intravenous infusion of gadolinium-DTPA demonstrating no evidence of residual tumor mass.

inferior end of the lateral wall and from the roof of the orbit for about 3 cm posteriorly was carried out separately. The basal squamous temporal bone and the superficial part of the sphenoid ridge were rongeured away. Further resection of the sphenoid ridge was performed to the base of the anterior clinoid using a high-speed drill. The dura was incised in a semicircular fashion centered on the pterional area. The sylvian fissure was split; the carotid and other basal cisterns were opened in the usual manner; sufficient cerebrospinal fluid was drained to obtain good brain relaxation. Polar temporal bridging veins were identified, coagulated and cut away. The temporal lobe was released and taken laterally. The clival region was reached by working in the opticocarotid distance and oculomotor nerve-carotid distance. A soft, friable, grayish mass, invading the dura like a patch, was seen in the clival region. The tumor, which was found to compress the brainstem, was clearly distinguished from the brainstem. The tumor mass was dissected from the neurovascular structure meticulously and aspirated by means of a suction tool, and the tumor tissue was dissected away from related nerves and arteries. The lesion was totally removed with the right combined pterional transsylvian and anterior temporal approach (+OZO).

Pathological Findings. The tumor had a myxomatous appearance on hematoxylin and eosin staining. The tumor tissue was characterized histologically by lobules composed of typical physaliphorous cells with abundantly vacuolated cytoplasm. These cells were embedded in an acellular mucoid matrix. There were many blood vessels between the lobules, surrounded by prominent lymphocytic infiltration. Mitotic figures and cytological atypia were not observed. The resected tumor had the typical histological and immunohistochemical characteristics of chordoma. S-100 protein and cytokeratin were both strong and diffuse positive.

Postoperative Course: The patient complained of diplopia due to right abducent nerve palsy, but this improved rapidly in the a few weeks. Cranial MR images after surgery revealed no evidence of residual tumor mass (Figure 3). The patient was discharged on the 10th postoperative day. No radiation therapy or chemotherapy was administered. After three years, the tumor recurred in the same area and the patient was operated again. The patient has been followed for two years since the second operation without any recurrence.

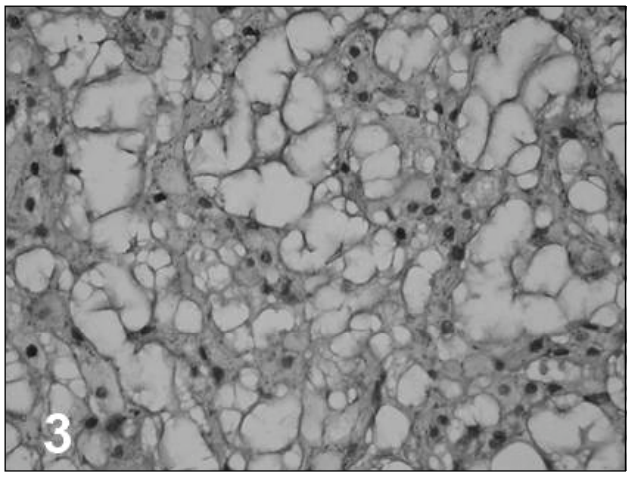


Figure 3: Histopathological photomicrograph. The tumour has a histological pattern typical of chordoma with cords of even, rounded cells set in a myxoid matrix. Typical physaliphorous cells are present.

DISCUSSION

Chordomas account for 1% of intracranial tumors and 4% of all primary bone tumors.4 They are relatively rare benign tumors that arise from embryonic remnants of the primitive notochord, a primitive cell line around which the skull base and the vertebral column develop. They may occur at any age but are usually seen in adults, with a peak prevalence in the 4th decade of life (1). Chordomas in children and adolescents are extremely uncommon. Of all chordomas, only 4.7 - 6.7% are in people of 20 years old or younger.8,12 The chordomas in young patients are classified as those in patients younger than 5 years of age and between 5 and 20 years of age (2). The reason for this is the difference between the spread of the lesion, treatment, histopathology, metastases and prognosis in the two groups. The prevalence of atypical histological findings with aggressive behavior is greater in patients younger than 5 years of age. The incidence of metastases is higher in children less than 5 years of age (57.9%) than older patients (8.5%).2 Chondroid variants were not found in patients younger than 5 years of age, whereas %17 of patients older than 5 years of age had a chondroid component to their tumors.2 There was no correlation between radical resection and improved prognosis for younger patients with atypical histological findings.2

Surgical removal is a very effective treatment for intracranial chordomas.3 Chordomas are thought to be essentially extradural in nature and are generally associated with bone destruction. Bone involvement by chordomas is characteristic and osseous destruction has often been described as a hallmark of these tumors (10). Dural invasion by extradural chordomas occurs late in their course, with very aggressive tumors, and in recurrent tumors when the dura was opened (14). Long survival rates have been associated with more extensive tumor removal. In skull base chordomas, the radical removal rate is 43.5%, subtotal removal rate 47.8% and partial removal rate 8.7% (1). Chordomas are basically midline tumors, so anterior midline approaches have generally been preferred. These include the transbasal, extended frontal, transseptal–transsphenoidal, facial translocation, transmaxillary, midfacial degloving, transoral, mandible-splitting transoral, transcervical–transclival, and anterior cervical approaches. Sometimes two or more skull base procedures may be necessary to achieve a radical removal (1,11). The main factors in selection of the approach are the anatomical limits reached, tumor compartment (intradural or extradural), localization, and size (1).

The pterional approach was first described by Yasargil.13 This approach is used for the treatment of vascular and tumor lesions by its own or in combination with other approaches. In general, the problem with the pterional approach is that the upward visualization of the surgeon is limited by the basal structures such as the orbital roof, sphenoidal ridge, frontal and temporal lobes, ICA, temporal muscle around the working space. This approach also requires the surgeon to work on a deep, narrow field between the optic nerve and the carotid artery or between the carotid artery and the oculomotor nerve. The OZO combined pterional transsylvian and anterior temporal approach is an effort to eliminate the disadvantages that are inherent to these two approaches while having the advantages of both approaches, and providing a much wider and shorter route. In the OZO combined pterional trans-sylvian and anterior temporal approach, brain retraction is minimal; furthermore, it is well known that posterior retraction of the temporal pole, as used in the combined approach, is much better tolerated than the temporal lobe elevation required by the subtemporal approach.7 This approach provides a much wider and shorter operative field making the multidirectional viewing of the tumor possible, the

dissection easier, and the exposure more generous. We often use a combined pterional transsylvian and anterior temporal approach for aneuryms of the upper basilar complex, purely posterior fossa lesions (epidermoid cyst vs.), and middle and posterior fossa component of tumors in our clinic. We prefer a wide cisternal/subarachnoid space dissection in order to get a good brain relaxation while using this combined approach. We certainly do proximal and distal sylvian cistern, chiasmatic cistern and carotid cistern dissection. With this technique, we get abundant cerebrospinal fluid drainage so that posterior and lateral retraction of temporal lobe and anterior retraction of frontal pole are easily obtained. On some MR images of our case, we obtained that the posterior margin of the tumor was irregular and it might have been dural invasion. We therefore chose the combined approach. In the case we present here, a total infratentorial mass was completely resected with a supratentorial approach by using OZO combined with pterional transsylvian and anterior temporal approach. During the surgery, we observed invasion of dura in a punctate fashion. From the edge of the tentorium to the anterior part of the brainstem and the basion, all clivus visualization was genereously provided. This combined approach can give exposure with minimal retraction for tumors anterior to the brain stem, towards the middle fossa, and to basion and all clivus.

In conclusion, we present a rare case of chordoma in an adolescent patient with a large mass located in the clival region. Treatment of skull base chordomas is very difficult and complex. The most important problems in surgical resection of a clival lesion is reaching the lesion and resecting it radically. Our case presented here is a lesion which had settled extradurally, involving the whole clival region and and placed totally infratentorially. A totally infratentorial lesion was completely resected with a supratentorial approach, by using physiological distances without any problem.

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