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CASE REPORT

Endoscopic Removal of Clival Chordoma

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ABSTRACT

Clivus chordomas are locally aggressive benign, with primary bone tumor arising from remnant of notochord anywhere from the coccyx to the base of the skull, in either a midline or paramedian position. The tumors show rare presentation and relatively common recurrence rate. Intracranial chordomas usually arise from the region around the clivus and account for about a third of all chordomas. Clival chordomas rarely metastasize and become symptomatic after local invasion to cranial nerves and skull base structure. Diagnosis is based on computed tomography scan/magnetic resonance imaging findings and histopathological examination of soft tissue mass. Complete surgical resection is the primary line of management with or without radiotherapy. We are presenting a case of clival chordoma of a 35-year-old female who presented with diminished vision and headache since 6 months. After evaluation, the patient was treated with endoscopic endonasal transsphenoidal resection of the tumor. After surgery, the patient showed relief from headache and slight improvement in vision loss.

Keywords: Clival chordoma, Endonasal, Endoscopic, Transsphenoid approach.

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INTRODUCTION

The clivus is the unpaired basilar portion of the occipital bone extending obliquely from the anterior aspect of the foramen magnum up to the basisphenoid bone just inferior to the dorsum sella. Hence, clival chordomas are midline structures projecting posteriorly and compressing the nearby structures.[1] Hence, spheno-occipital chordomas are also called as "clival chordomas" and the most common presenting symptoms are headache (50%), visual complaints, facial pain, numbness or paresthesia (50%), and diplopia (42%). Other nonspecific symptoms of chordomas are dysphagia and dysarthria, ataxia, extreme weakness, and hoarseness.

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Chordomas are rare, slow-growing benign tumors of the bone that account for 4% of all primary bone tumors.[2] These tumors arise from the remnants of primitive notochord, which extend from the Rathke's pouch at skull base to the coccyx. The cranial and caudal end of the spine are the most commonly affected areas due to nests of notochordal cells behind the clivus and the sacrococcygeal regions, while the remainder of the notochord regresses during fetal life.

Incidence is less than 0.1/100,000 of population.[3] Mean age of presentation is 47 years (8–80 years). Incidence is slightly more in male. Sacral-coccygeal chordomas (50–60%) are the commonest type of presentation, followed by skull base or spheno-occipital chordomas (25–35%) and the cervical vertebral chordomas (approximately 10%).[4]

Chordomas of skull base usually occur in patients of 20 to 40 years of age, whereas sacrococcygeal chordomas are typically seen in an older age group of around 50 years.[2,3] These are locally aggressive and erode the surrounding bones and rarely metastasize to distant sites over a period of time.[5]

CASE REPORT

A 35-year-old female patient presented to our ear, nose, and throat outpatient department with complaint of headache for the past 7 years and left side blindness with right side diminished vision, which was progressively increasing in the last 6 months. Headache was spontaneous, continuous, dull aching, holocranial, slowly progressive in severity, and was not associated with vomiting. She developed spontaneous, painless, progressive diminished vision in the left eye. She gave no significant past medical or surgical history.

Detailed history was taken and general examination was done. Bilateral diminished vision, more on the left side, was present. Bilateral fundus examination was normal. Visual evoked potential was done, which showed nonrecordable waveform on left side and normal P100 latency with borderline reduced amplitude in the right side. No other significant clinical finding was present.

On radiological examination, computed tomography (CT) paranasal sinuses (PNS) (Fig. 1) and magnetic resonance imaging (MRI) (Figs 2A and B) showed large altered signal intensity mass lesion occupied bilateral sphenoid and posterior ethmoid air cells. No obvious bony erosion of anterior, midcranial fossa, or intracranial

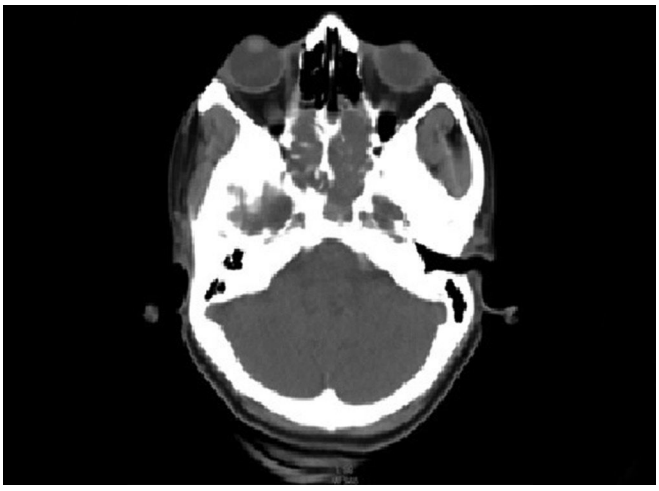


Fig. 1: Contrast-enhanced CT brain with PNS in axial section showing large destructive lytic soft tissue mass involving body of sphenoid extending into middle cranial fossa posteriorly and anteriorly into bilateral ethmoid sinuses

extension was seen. Pituitary gland appears normal and was pushed posterosuperiorly. Mass lesion causes bony remodeling of posterior end of bilateral medial orbital wall and bulging it laterally (more on left side). Lesions appear to be compressing bilateral optic nerve. These findings were suggestive of clival chordoma.

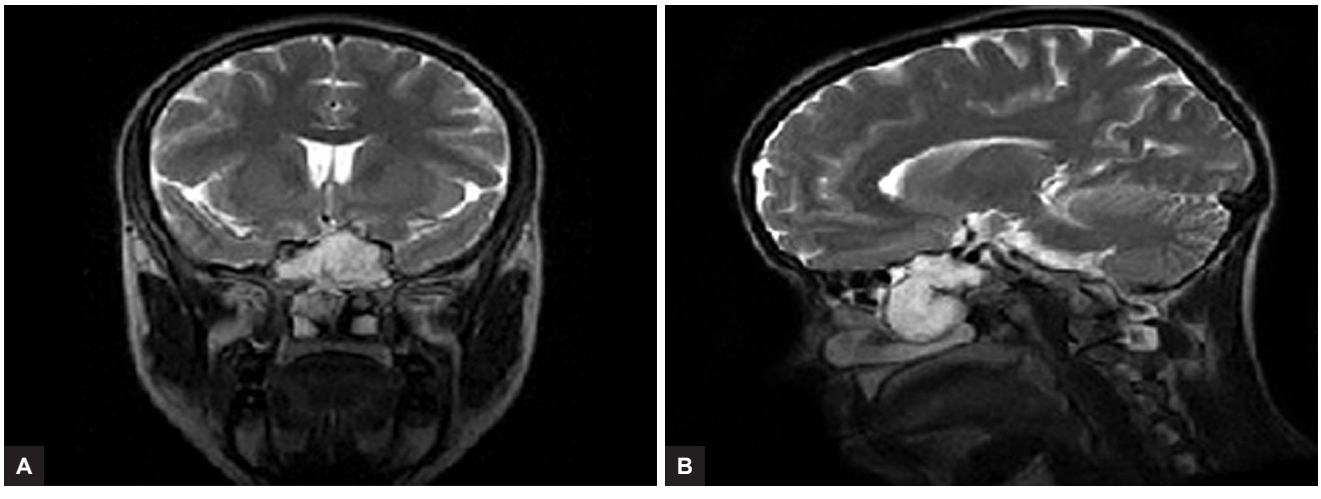
After complete evaluation, endoscopic endonasal transsphenoidal removal of the tumor mass from the clivus under general anesthesia was done. During surgery, the tumor was approached through the sphenoid sinus and the tumor was totally removed in small pieces by curetting and chiseling out along with pterygoid plate. Dura and optic nerve was exposed, which was found to be intact. The residual space and defect in dural plate was closed with fat graft taken from the abdominal wall and Hadad-Bassagasteguy flap[6] to prevent cerebrospinal fluid leak in case if it happens and to protect the intracranial tissue. The surgery was uneventful. No neurological deficits were seen after the surgery. Patient recovered well in the postoperative period with complete relief from headache and slight improvement in vision, and was discharged after 10 days of surgery. Postoperative CT PNS was done and no abnormality was detected. Histopathological report of the surgical specimen shows physaliferous cells separated by fibrous and myxoid stroma, suggestive of chordoma (Fig. 3).

DISCUSSION

In the 1980s, only 3 cases of clival chordoma were reported, whereas there were 8 cases in the 1990s, 27 cases in the 2000s, and an additional 9 from 2011 to 2013.[7] This increasing number of clival chordoma reporting may be due to increasing number of literature on this disease, modern diagnosing modalities like CT scan and MRI, modern surgical interventions with the introduction of endoscopic endonasal approaches (EEAs) to the clivus, and increasing longevity of patients.

Clinical presentation of clival chordomas varies according to the location and size of the tumors.[3] Symptoms are due to the mass effect on adjacent structures.[2] They present with cranial nerve palsies presenting as visual disturbances or loss of vision, orbitofrontal headache, loss of facial sensation, voice change and difficulty in swallowing, neck pain, and intracranial hemorrhages.[8-10] But they can also remain silent for a long duration as they are slow growing and can pose difficulty and delay in diagnosis of the disease.

The CT scan and MRI play a very important role in the diagnosis and evaluation of the clival chordoma: CT scan helps to detect any bony erosion by the tumor and the degree of ossification within the tumor, whereas MRI helps in analyzing the brainstem involvement better than



Figs 2A and B: T2-weighted image of MRI brain with PNS showing benign looking mass correlating with the CT findings. Also optic nerves were compressed, which was more in left side

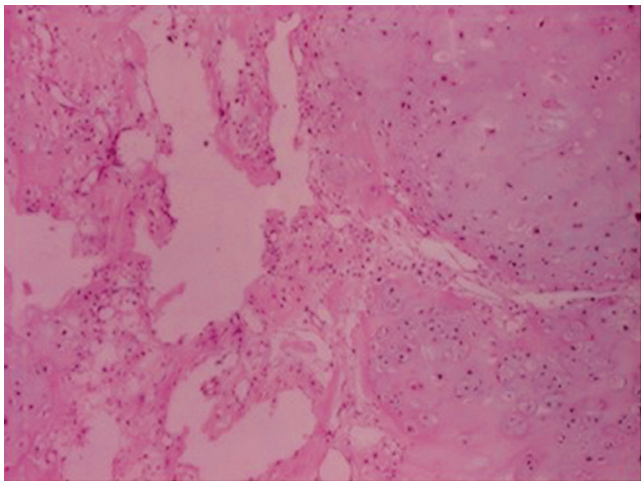


Fig. 3: Histopathological slide of the patient

the CT,[2,11,12] and on MRI examination, they are typically isointense to hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging and typically show heterogeneous enhancement, which helps to distinguish it from meningiomas and schwannomas.[2,12]

On histopathological analysis of the surgical specimen, clival chordomas show fluid and mucoid substance with hemorrhagic and necrotic areas within the tumor. In some cases, calcification and bone sequestrations can also be found.[1,10,13-15]

Immunohistochemical staining shows positive staining for S100, Vimentin, Cytokeratin, carcinoembryonic antigen, and epithelial membrane antigen (positivity only in chondroid differentiation).

Radical resection with or without radiotherapy of clival chordomas has been recommended by many researchers for better results,[16] and radiotherapy can be given to recurrent cases.[2,17] Approach to the clival region has always remained a challenge for the surgeons over a period of time. Transbasal, orbitozygomatic, subtemporal, transcondylar, transmaxillary, and extended transsphenoidal techniques have been proposed in the past.[16,18,19] With the development of nasal endoscopic surgery, EEA has been developed as it takes advantage of the natural sinus route and may provide a minimally invasive approach for these midline tumors.[1,5,9,20] This EEA shows almost equivalent results to extensive approaches.[21,22] The complication of CSF leak can be managed with local flap repair techniques. Hence, this approach should be in the surgery of choice for the management of clival chordomas.[20-22]

Conventional radiotherapy with high-energy photons at a dose of 60 to 65 Gy may offer temporary benefit in disease control in patients with inadequate surgery or recurrence or as exclusive treatment for inoperable disease.[22]

Prognosis of the tumor is typically poor, due to the locally aggressive nature of these tumors, with the 10-year survival approximately 40%.[2,17,22]

CONCLUSION

Endoscopic endonasal surgery is the safe and reliable approach of clival chordoma tumor resection. It is a minimally invasive procedure and should be considered as a preferential approach to conventional surgical method.

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