

CASE REPORT

Removal of a low clival chordoma in a teenager by dorsolateral suboccipital transcondylar approach

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ABSTRACT

Chordomas are rare midline tumors of the central nervous system which arise from the remnants of the primitive notochord and have unique diagnostic and management challenges. Although recommended treatment for chordoma is radical resection, this may require extended skull base approaches. We report a case of low clival chordoma in a teenage patient which was successfully treated by dorsolateral suboccipital transcondylar approach.

Key words: Clivus, dorsolateral suboccipital transcondylar approach, low clival chordoma

Introduction

Chordomas mainly occur in the sacrococcygeal area (50%) and in the basisphenoidal region (35-40%) where they typically involve the clivus.^[1] Most of the remainder is related to vertebral bodies.

They account for approximately 1% of all intracranial tumors. Complete removal of the clivus chordoma is extremely difficult to achieve, even with radical surgery, because a few tumor cells are frequently left behind in the bone and dura. Therefore, adequate exposure of the lesion is important. Various extended skull base approaches particularly the trans-oral approach has been used to improve the visualization of the tumor, facilitating its complete removal, thus reducing the morbidity due to brain retraction. In our case report, we present a rare case of a chordoma in low clival region in an 18-year-old treated by dorsolateral suboccipital transcondylar approach.

Case Report

An 18-year-old male was presented in January 2012 with an aching type recurrent diffuse headache, which is not relieved by simple analgesics. It was associated with dizziness and

recurrent attacks of loss of consciousness. General physical examination was normal. The initial neuroimaging was unable to detect any lesions in the brain.

One month after, a follow-up magnetic resonance imaging (MRI) was able to detect a solid and cystic mass lesion arising from lower clivus causing external compression of the brain stem and obliteration of the fourth ventricle. There was cerebellar tonsillar ectopia with added compression [Figures 1 and 2]. A trans-oral biopsy was done. Histology report revealed cells with moderately pleomorphic round-oval nuclei arranged in a myxoid stroma. Epithelial membrane antigen and cytokeratin were strongly positive. S-100 protein was focally positive.

While waiting for surgery, in September 2012, he presented with neck pain during head extension and progressive weakness of the right upper and lower limbs. He had slurring of speech, recurrent dysphagia for solids and liquids. He noticed blurring of vision in the right eye. Physical examination revealed right-sided tongue wasting, lower motor neuron type IX and XII nerve palsy. There was upper motor neuron type weakness in the right upper and lower limbs.

In February 2013, tumor was excised by dorsolateral suboccipital transcondylar approach with the guidance of neuronavigation. Patient was positioned sitting and an inverted U incision was made in the left occipital region starting at the mastoid process, proceeding under the superior nuchal line and continuing down the midline to the C6 spinous process. A left posterior fossa craniotomy was done with excision of the left posterior part of the C1 vertebra. Foramen magnum was opened and left-sided vertebral artery was identified and preserved. Dura was hitched and opened. Left cerebellum was retracted, and chordoma which was extending along the brainstem and the anterior extradural extension of clival region was excised.

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After the surgery, patient developed a mild headache, cerebrospinal fluid rhinorrhea, and numbness in left lower limb which resolved after a few days. Cranial computed tomography (CT) after surgery revealed a pneumocephalus and a seroma at the surgical site [Figures 3 and 4].

Discussion

Clivus is formed by the union of the basiocciput and basisphenoid. Basiocciput develops from the fusion of four occipital sclerotomes. Incidentally, this junction also represents the cephalic end of the notochord. Therefore, cellular remnants of the primitive notochord are seen in the cephalic and caudal regions and chordomas are seen in these two regions.^[2] Chordomas are slow growing, but due to their location, an invasive nature, and recurrence rate, these tumors are considered as malignant. Symptoms of these tumors vary with their location and size. Most commonly, the patient presents with headache, diplopia secondary to VI cranial nerve

palsy, and changes in vision. The patient may present with symptoms of multiple lower cranial nerve palsy. Patients with large tumors may present with long tract signs and ataxia.

On CT scan, chordomas appear as well-defined extra-axial masses that enhance with contrast. CT is superior to MRI in demonstrating the tumoral calcification and associated bone destruction. On T1-weighted MRI, these tumors are typically iso-to hypodense and on T2-weighted imaging hyperdense and show typical heterogeneous enhancement.^[2] In general, MRI is better for defining the anatomy of the surrounding structures and tumor extension into the nasopharynx and cavernous sinus. Histologically, tumor appears as pleomorphic cells with dark nuclei and vacuolated or granular cytoplasm set within an abundant myxoid matrix. Neoplastic cells are arranged in epithelial cords separated by mucinous material, which is a classic feature of chordomas. On immunohistochemistry, the cells are positive for S-100 protein and epithelial membrane antigen.

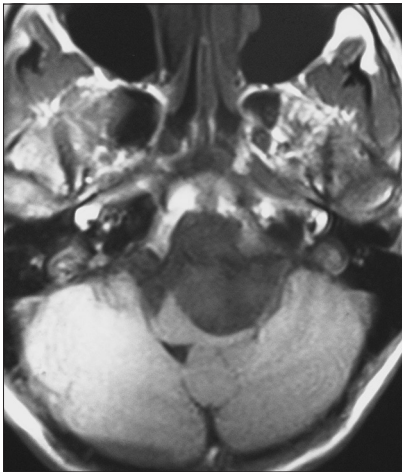


Figure 1: Presurgical magnetic resonance imaging images, axial view, showing a mass within the clival region

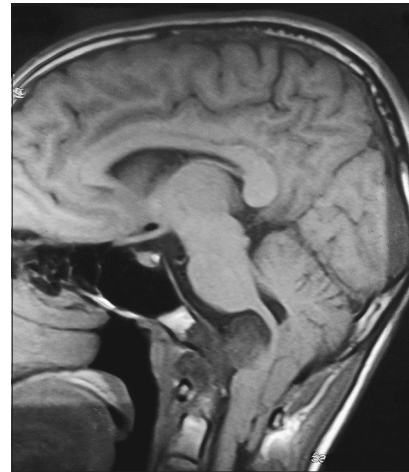


Figure 2: Presurgical magnetic resonance imaging images, sagittal view, showing a mass within the lower clival region

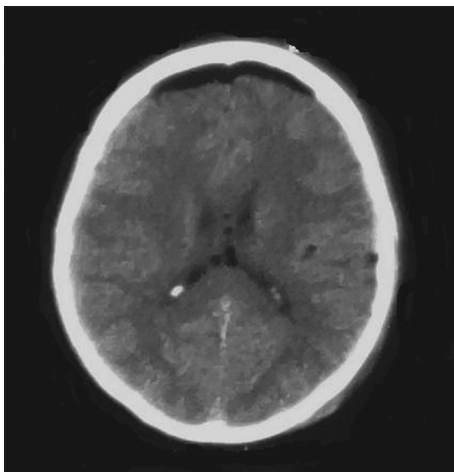


Figure 3: Postsurgical computed tomography images, axial view, showing pneumocephalus

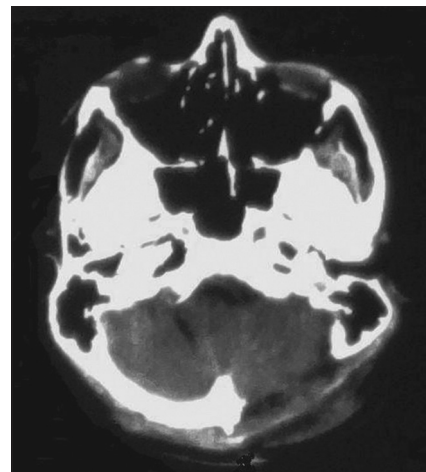


Figure 4: Postsurgical computed tomography images, axial view, showing seroma collection at the surgical site

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.^[3] The chordomas in young patients can be classified as those between 5 and 20 years of age and those who are younger than 5 years of age.^[1] This is due to the considerable difference between the clinical presentations, spread of the lesion, treatment, histopathology, metastases, and prognosis in the two groups. Chordomas are essentially extradural and associated typically with an extradural extension and bone destruction. Dural invasion by extradural chordomas occurs late in their course, with very aggressive tumors, and in recurrent tumors when the dura was opened.^[4] Patients with chordomas have a life expectancy of < 10 years after diagnosis.^[5] Long survival rates have been associated with more extensive tumor removal.^[6,7]

Chordomas are midline tumors. Therefore, trans-oral approaches have generally been preferred.^[8] The main factors in the selection of the approach are the anatomical limits reached, tumor compartment (intradural or extradural), localization, and size.^[6] In our case, we chose dorsolateral suboccipital transcondylar approach. This approach has been useful in lesions related to anterior rim of the foramen magnum, the lateral clivus, and the front of the lower brain stem and for access to the vertebrobasillar circulation. Two types of incisions are used to expose the lateral mass of the atlas and the vertebral artery, the inverted hockey stick incision, and the paramedian incision. Both are capable of exposing the ipsilateral occipital bone and the spinous processes and the posterior laminar arches of C1 and C2 depending on the inferior extent of the lesion.^[9]

Currently, there are new surgical approaches like endoscopic surgery in the management of clival chordomas, which not only provides direct surgical access but also an excellent

visualization of the clivus and surrounding structures. However, no particular surgical protocols are described. When complete resection of the tumor is not achieved, radiation treatment is recommended.^[2,10]

References

1. Gay E, Sekhar LN, Rubinstein E, Wright DC, Sen C, Janecka IP, *et al.* Chordomas and chondrosarcomas of the cranial base: Results and follow-up of 60 patients. *Neurosurgery* 1995;36:887-96.
2. Oot RF, Melville GE, New PF, Austin-Seymour M, Munzenrider J, Pile-Spellman J, *et al.* The role of MR and CT in evaluating clival chordomas and chondrosarcomas. *AJR Am J Roentgenol* 1988;151:567-75.
3. Heffelfinger MJ, Dahlin DC, MacCarty CS, Beabout JW. Chordomas and cartilaginous tumors at the skull base. *Cancer* 1973;32:410-20.
4. Uttley D, Moore A, Archer DJ. Surgical management of midline skull-base tumors: A new approach. *J Neurosurg* 1989;71:705-10.
5. St Martin M, Levine SC. Chordomas of the skull base: Manifestations and management. *Curr Opin Otolaryngol Head Neck Surg* 2003;11:324-7.
6. al-Mefty O, Borba LA. Skull base chordomas: A management challenge. *J Neurosurg* 1997;86:182-9.
7. Crockard HA, Steel T, Plowman N, Singh A, Crossman J, Revesz T, *et al.* A multidisciplinary team approach to skull base chordomas. *J Neurosurg* 2001;95:175-83.
8. Sekhar LN, Sen CN. Anterior and lateral basal approaches to the clivus. *Contemp Neurosurg* 1989;1:1-7.
9. Lang DA, Neil-Dwyer G, Iannotti F. The suboccipital transcondylar approach to the clivus and cranio-cervical junction for ventrally placed pathology at and above the foramen magnum. *Acta Neurochir (Wien)* 1993;125:132-7.
10. Borba LA, Al-Mefty O, Mrak RE, Suen J. Cranial chordomas in children and adolescents. *J Neurosurg* 1996;84:584-91.

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