Endoscopic Endonasal Approach for Clival Chordomas – Case Report and Review of Literature

RESUMO
Cordomas do clivo são lesões agressivas que se originam de remanescentes da notocorda primitiva, apresentando diagnóstico e conduta desafiadoras. A ressecção radical da lesão tem sido recomendada para melhor prognóstico e o tratamento cirúrgico constitui desafio ao neurocirurgião. Vias de acesso transbasal, orbitozigomática, subtemporal, transcondilar, transmaxilar e transesfenoidal extensiva têm sido opções no tratamento cirúrgico. Relatamos caso de cordoma de clivus tratado por via endoscópica endonasal transesfenoidal.

Palavras-chave: Base de crânio, cordoma de clivus, acesso endoscópico endonasal, transesfenoidal.

ABSTRACT
Chordomas of the clivus are aggressive lesions which arise from the remnants of the primitive notochord and pose unique diagnostic and management challenges. Radical resection of chordomas has been recommended for better outcomes and their surgical treatment has been challenging to neurosurgeons for many years. Transbasal, orbitozygomatic, subtemporal, transcondylar, transmaxillary, and extended transsphenoidal techniques have been proposed. We report a case of clival chordoma treated by endonasal endoscopic transsphenoidal approach.

Key words: Skull base, clival chordoma, endonasal endoscopic approach, transsphenoidal.

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INTRODUCTION

Chordomas of the clivus are aggressive lesions which arise from the remnants of the primitive notochord and pose unique diagnostic and management challenges. They account for 1% of all intracranial tumours and present with cranial nerve palsies, orbito-frontal headache, visual disturbances and intracranial hemorrhages. Under the microscope, chordoma cells appear to be benign, but due to their location, invasive nature, and recurrence rate, these tumours are considered as malignant.

About one-third of chordomas are found around the clivus, but they occur with equal frequency at the skull base, the vertebrae and the sacrococcygeal area towards the bottom of the spine. Symptoms of these tumours vary with their location and size.

Radical resection of chordomas has been recommended for better outcomes and their surgical treatment has been challenging to neurosurgeons for many years. Transbasal, orbitozygomatic, subtemporal, transcondylar, transmaxillary, and extended transsphenoidal techniques have been proposed.

We report a case of a clival chordoma treated by endonasal endoscopic transsphenoidal approach.

CASE REPORT

A 47 year old man was admitted with a 8 months’history of bifrontal mild headache, dull aching in character, without nausea or vomiting, and relief with common analgesics. Eight weeks prior to admission, he noticed blurring of vision in the left eye and occasional diplopia. General physical examination was normal. He had minimal left abducens palsy. MRI and CT studies (Fig. 1) demonstrated a large destructive soft tissue mass arising from the clivus and extending both anteriorly into the sphenoid sinus and posteriorly into the prepontine cistern, where it indents the pons and displaces the basilar artery leftwards. The mass is isodense to the brain on CT, and on MR shows a low T1 and a high T2 signal, with high contrast enhancement.

We chose an endoscopic endonasal transsphenoidal direct approach to the clival region (Fig. 2). Our main preoperative concern was the possible occurrence of CSF leakage. To minimize that risk, we kept the dural opening as small as possible. A large abdominal fat graft was placed at the dura, during closure. The anesthesiologist performed Valsalva maneuvers to help confirming watertight closure. The patient was discharged 5 days after the procedure without complications. A three-months follow-up MRI showed total tumor resection and the patient recovered from his visual symptoms.
Chordomas are rare tumours believed to arise from remnants of the notochord. About 45-50% of them are reported to arise at the sacrococcygeal region; 35-40% at the sphenosquamosal region; the remaining 15% of these tumours arise along other sites of the spine. The typical age range of patients presenting with these tumours is 30-50 years old for intracranial tumours and 40-60 years old for sacrococcygeal lesions. Intracranial chordomas classically arise in the midline in the region of the clivus. Patients can present with headache, nausea, vomiting, and cranial nerve palsies. These tumours are normally very locally aggressive, often showing extensive surrounding bone erosion. Distant metastasis is an unusual occurrence, although there is a report of a chordoma of the clivus presenting with cutaneous manifestations due to metastasis.

On imaging, chordomas are well-defined extra-axial masses that enhance with contrast on CT. The tumours may demonstrate focal calcifications. On MRI, these tumors are typically iso- to hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging and typically show heterogeneous enhancement. This heterogeneous pattern of enhancement helps distinguish chordomas from other common tumours of the skull base such as meningiomas and schwannomas that normally have a more homogenous pattern of enhancement.

Pathologically, chordomas appear grossly gray to tan in color with gelatinous texture and are often lobulated and encapsulated. They can be divided into chondroid and nonchondroid subtypes on basis of the evidence of chondroid differentiation within the tumour. Histologically, they show a pattern of cells with foamy cytoplasm in a mucinous matrix (the characteristic physaliphorous cells). Immunohistochemical staining will often demonstrate positive staining for cytokeratin and epithelial membrane antigen (EMA) with variable staining for vimentin (in cases of chordomas with chondroid differentiation) and S-100. The epithelial phenotype, expressed in positive staining for cytokeratin and EMA, is important in differentiating chordomas with chondroid features from chondrosarcomas.

Surgical treatment of chordomas has been challenging to neurosurgeons for many years. Transbasal, orbitozygomatic, subtemporal, transcordylin, transmaxillary, and extended transphenoidal techniques have been suggested. However, no particular surgical protocols are described. When complete resection of the tumour is not achieved, radiation treatment is recommended.

The endoscopic endonasal transclival approach uses a minimal access gateway, but with obtainable maximal visualization to the clival region. In addition, neuronavigation aids in localizing the lesion and surrounding vital structures, estimating depth through this long surgical approach providing a secure tool for the surgeon’s understanding of anatomical relationships. Another important technique in this approach is the use of angled endoscopes: 30 and 45-degree endoscopes help in providing a wider view of the exposed skull base, and extend the operative region without expanding the surgical approach. By moving the lens and the light source so close to the field, angled endoscopes extend the straight microscopic transphenoidal view into a more extensive operative vista, which turn feasible the extended approaches to the skull base.

Postoperative CSF leak—the often-feared complication—was reported in 16.7% of cases in the published series. Clearly, the degree of intradural invasion of the tumour has a major impact on the risk of this complication.

Surgical management of clival chordomas is undergoing a paradigm shift as endoscopic endonasal approaches become more widely practiced. It is difficult to directly compare these approaches with open microsurgical approaches specifically for chordomas, as the rare epidemiology of the disease does not provide significant case numbers. However, in examining the limited published data and adding our own experience, it is clear that the early short-term results of endoscopic endonasal transclival resection, combined with radiotherapy, provides an encouraging option for higher rates of tumour control and progression-free survival, with limited morbidity for selected cases of moderate-sized midline tumours.

### REFERENCES


