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Case Report

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Clivus chordoma: a surgical challenge using the endonasal endoscopic technique

Key message

The aim of this article is to report a case of an endoscopic endonasal approach to a chordoma in the clivus region. This rare, slow-growing and locally aggressive tumor had already been surgically approached 12 years ago. With the advance of the endoscopic endonasal surgery, it is possible to access this critical and very important region, which is close to the central nervous system and very important nervous and vascular structures.

Abstract

Introduction: Chordomas are rare tumors derived from embryonic remnants of the notochord. They commonly affect the axial skeleton, including the skull base and sacral region, posing challenges in treatment due to their aggressive nature and high recurrence rates.

Case presentation: Patient GKHP, a 43-year-old female, presented with a recurrent clivus chordoma following initial surgery in 2010. Endoscopic endonasal resection was performed in July 2022, resulting in a cerebrospinal fluid fistula postoperatively. Reconstruction utilized various materials, requiring a second surgical intervention and antibiotic treatment.

Discussion and conclusion: Chordomas represent a small percentage of malignant bone tumors, originating from primitive notochord remnants. While surgical excision remains the primary treatment, challenges persist due to tumor location and invasiveness. Endoscopic techniques offer promise in managing complex cases, although proximity to vital structures presents ongoing challenges. A multidisciplinary approach, including surgery and radiotherapy, remains essential in achieving successful outcomes for patients with chordomas.

Keywords: chordoma, skull base, clivus, endoscopic surgery

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Background

Chordomas are rare, slow-growing but locally aggressive tumors derived from embryonic remnants of the notochord. They have high recurrence rates which can lead to a locally uncontrolled disease and mortality. Malignization occurs in 2 to 8% of the cases. The most affected regions are the axial skeleton, including the sphenooccipital region, the skull base and the sacral region. The chordoma of the skull base, especially in the clivus region, affects a delicate region surrounded by important structures such as the encephalon, nerve structures and noble vascular structures.[1–3]

Patients are usually affected by vague symptoms such as headache, neck pain, diplopia and cranial nerve involvement, which delay the diagnosis. Imaging tests such as MRI will show the tumor and its dural extension, while CT scans are better at showing bone lesions. PET will be useful for staging, monitoring the progress and the response to the treatment.[4]

There are no randomized clinical treatment protocols, but the literature supports the consensus that the maximum surgical treatment (i.e. with the greatest possible resection of the tumor) combined with radiotherapy is the best therapeutic modality to date, since due to the impaired location, the total resection is unfeasible in most cases.[5–7]

Case presentation

Patient female, 43 years old, presented with a recurrence of a clivus chordoma whose first surgery was carried out in 2010. The patient presented with headache as the sole symptom, and the follow-up magnetic resonance imaging revealed recurrence of the previously operated lesion. The patient underwent an endoscopic endo-nasal resection with neuro navigation in July 2022, a procedure which intraoperatively generated a cerebrospinal fluid fistula with a high output, a high pressure and a difficult reconstruction of the large defect. For the reconstruction, a Duradry dural substitute (Technodry, Belo Horizonte MG), fat, a free septal cartilage graft, a naso-septal flap and an Adherus dural sealant (Hyperbranch Inc, Durham, North Carolina) were used. This was followed by a rigorous nasal hemostasis, hemostatic materials and a bilateral finger nasal packing. After 11 days, the patient presented with rhinoliquorrhea and on the 12th post-operative day she underwent a new surgical approach. In the second procedure, a large free double-layer fascia lata graft, fat, a septal cartilage, a nasoseptal flap and an Adherus and a Tissucol dural sealant were used for the closure. The hemostasis and the tamponade followed in the same way. In the second procedure, a lumbar drain was placed. The cerebrospinal fluid culture revealed *S. aureus*, which was treated with Oxacillin for 14 days. The patient was discharged in good conditions with no further complications and was referred for a radiotherapy and an otolaryngological and neurosurgical follow-up (Figure 1-5).

Discussion

Chordomas account for 1 to 4% of all malignant bone tumors and originate from embryonic remnants of the primitive notochord.

They can affect the skull base and the axial skeleton. These tumors are mostly found in the sacrum and more than a third are intracranial, preferentially affecting the clivus region.[8–10] They are rare, have a strong tendency to a local invasion and a recurrence can cause different levels of neurological deficits, but rarely cause metastases. The treatment is traditionally surgical and with the advent of the endoscopic surgery, increasingly larger and more complex lesions can be treated. The surgery can be challenging because the dorsal region of the clivus corresponds to the brainstem and the vertebrobasilar system. With trained and experienced surgical teams, it is possible to achieve a good success rate with less morbidity using the endoscopic technique with neuro navigation over the conventional technique. Radiotherapy has also played an important role in these neoplasms, which are typically refractory to chemotherapy.[11–13]

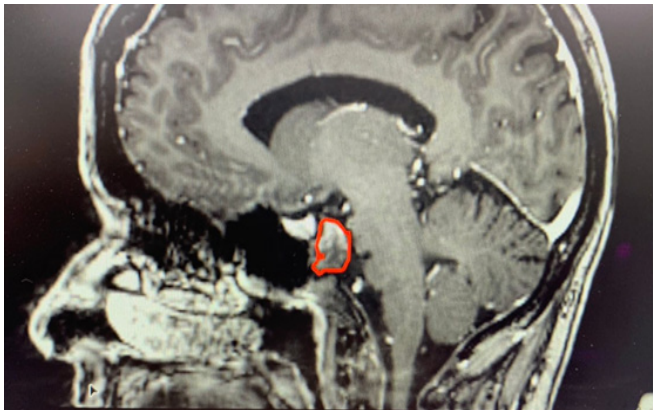

Figure 1 Magnetic Resonance Imaging (MRI) in sagittal section revealing the tumor.

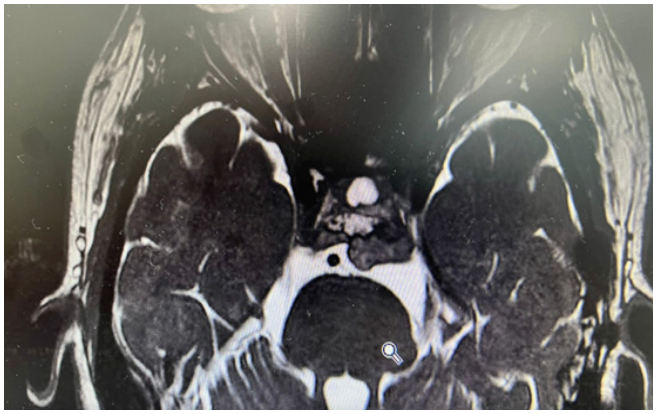

Figure 2 Magnetic Resonance in axial section demonstrating tumoral area.

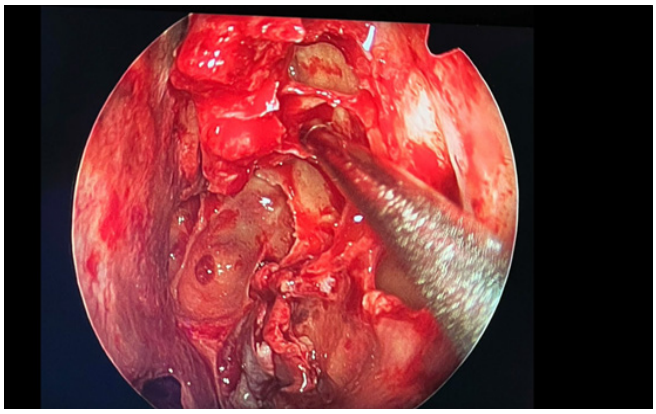

Figure 3 Clivus region displaying bone erosion.

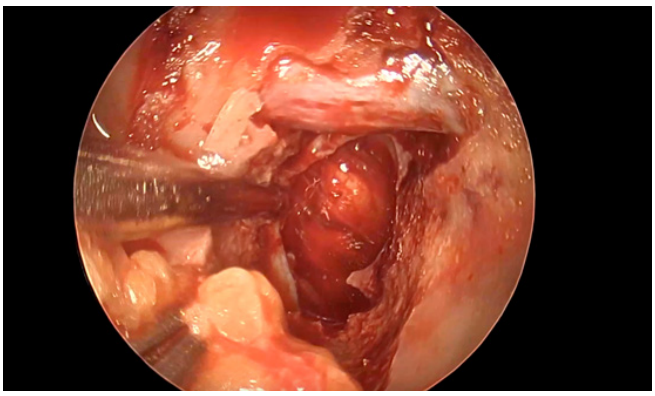

Figure 4 The skull base exposed after tumor resection.

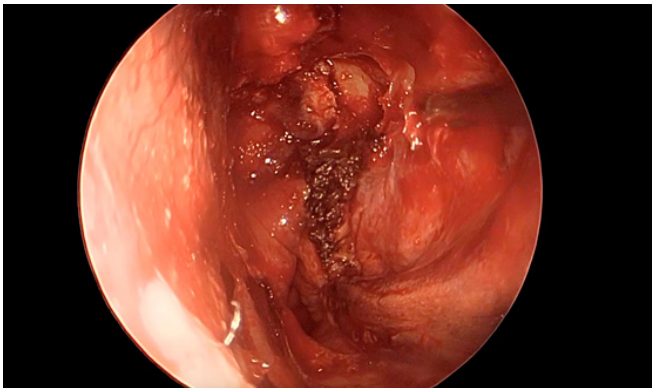

Figure 5 Skull base after reconstruction in multiple layers.

Conclusion

With the advent of the endoscopic endonasal surgery, more and more tumors and different anatomical sites can be treated with a lower morbidity and higher success rates. Clivus chordoma becomes a challenge even in the hands of experienced teams due to its proximity to noble structures and the high cerebrospinal fluid pressure, which makes it difficult to allocate free grafts and flaps for a reconstruction of the skull base defect. A good preoperative radiological assessment, the use of appropriate techniques and a neuro navigation device facilitate a good surgical excision, which in most cases will be complemented by radiotherapy.

Ethics approval and consent to participate

Not applicable.

Consent for publication

A copy of the consent form is available for review by the Editor of this journal.

Authorship contribution

LBP wrote the protocol, TVS collected the data, PRN analyzed and interpreted data, CSC made the literature review and wrote the main text, all authors read and approved the final manuscript.

Availability of data and materials

The data sets used and/or analysed during the present study are available from the corresponding author upon reasonable request. The name of the patient whose case report was analysed has not been disclosed in the article, the remaining data sets are available in the present study.

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None.

Conflicts of interest

The authors declare that there are no conflicts of interest.

References

1. Volpe NJ, Liebsch NJ, Munzenrider JE, et al. Neuro-ophthalmologic findings in chordoma and chondrosarcoma of the skull base. *Am J Ophthalmol*. 1993;115(1):97–104.
2. Leong JL, Batra PS, Citardi MJ. CT-MR image fusion for the management of skull base lesions. *Otolaryngol Head Neck Surg*. 2006;134(5):868–876.
3. Olson JT, Wenger DE, Rose PS, et al. Chordoma: 18F-FDG PET/CT and MRI imaging features. *Skeletal Radiol*. 2021;50(8):1657–1666.
4. Heffelfinger MJ, Dahlin DC, MacCarty CS, et al. Chordomas and cartilaginous tumors at the skull base. *Cancer*. 1973;32(2):410–420.
5. Tenny S, Varacallo M. Chordoma. In: bone tumors: diagnosis, treatment and prognosis, 2nd Ed, WB Saunders, Philadelphia. 1991;599.
6. Perzin KH, Pushparaj N. Nonepithelial tumors of the nasal cavity, paranasal sinuses, and nasopharynx. A clinicopathologic study. XIV: Chordomas. *Cancer*. 1986;57(4):784–796.
7. Harbour JW, Lawton MT, Criscuolo GR, et al. Clivus chordoma: a report of 12 recent cases and review of the literature. *Skull Base Surg*. 1991;1(4):200–206.
8. Rich TA, Schiller A, Suit HD, et al. Clinical and pathologic review of 48 cases of chordoma. *Cancer*. 1985;56(1):182–187.
9. Sen CN, Sekhar LN, Schramm VL, et al. Chordoma and chondrosarcoma of the cranial base: an 8-year experience. *Neurosurgery*. 1989;25(6):931–940.
10. Stacchiotti S, Sommer J, Chordoma Global Consensus Group. Building a global consensus approach to chordoma: a position paper from the medical and patient community. *Lancet Oncol*. 2015;16(2):e71–e83.
11. Fernandez-Miranda JC, Gardner PA, Snyderman CH, et al. Clival chordomas: a pathological, surgical, and radio therapeutic review. *Head Neck*. 2014;36(6):892–906.
12. Koutourousiou M, Gardner PA, Tormenti MJ, et al. Endoscopic endonasal approach for resection of cranial base chordomas: outcomes and learning curve. *Neurosurgery*. 2012;71(3):614–624.
13. Cannizzaro D, Tropeano MP, Milani D, et al. Microsurgical versus endoscopic trans-sphenoidal approaches for clivus chordoma: a pooled and meta-analysis. *Neurosurg Rev*. 2021;44(3):1217–1225.