

ENDOSCOPIC RESECTION OF CLIVAL CHORDOMA. A CASE REPORT

Resección endoscópica de cordoma del clivus. Descripción de un caso

Inês GAMBÔA¹; Dítza VILHENA¹; Mário RESENDE²; Delfim DUARTE¹; Gustavo LOPES¹

¹ Department of Otolaryngology – Hospital Pedro Hispano ULS Matosinhos. Portugal. ² Department of Otolaryngology – Centro Hospitalar Vila Nova de Gaia. Espinho. Portugal.

Correspondence: inesfariagsilva@gmail.com

Reception date: December 4, 2017

Date of Acceptance: December 16, 2017

Publication date: December 19, 2017

Date of publication of the issue: June 1, 2019

Conflict of interest: The authors declare no conflicts of interest

Images: The authors declare that the images have been obtained with the permission of the patients

Rights policy and self-archive: the self-archive of the post-print version (SHERPA / RoMEO) is allowed

License CC BY-NC-ND. Creative Commons Attribution-Noncommercial-NoDerivate Works 4.0 International

University of Salamanca. Its commercialization is subject to the permission of the publisher

SUMMARY: Introduction: Chordoma is a rare malignant tumor that arises from remnants cells of the primitive notochord, which are located at caudal and cephalic ends of the vertebral column. It represents 2 to 5 % of all primary bone tumors. Description: We report the case of a patient with a clival chordoma, asymptomatic, diagnosed as an accidental finding in a paranasal sinus. imaging study. Discussion: The imaging findings were suggestive of a potentially malignant lesion given the underlying bone lysis. Once the diagnosis is histological, biopsy of clival suspicious lesions should be promptly carried out. In this case report, the surgical approach and the postoperative follow-up are presented.

KEYWORDS: chordoma; clivus; endoscopic resection; malignant tumor.

RESUMEN: Introducción: El cordoma es un tumor maligno raro que surge de las células remanentes de la notocorda primitiva, que se localizan en los extremos caudal y cefálico de la columna vertebral. Representa del 2 al 5% de todos los tumores óseos primarios. Descripción: Presentamos el caso de un paciente con un cordoma del clivus, asintomático, diagnosticado como hallazgo accidental en estudio de imagen de los senos paranasales. Discusión: los hallazgos de imagen fueron sugestivos de una lesión potencialmente maligna dada la lisis ósea subyacente. Dado que el diagnóstico es histológico, la biopsia de una lesión clival

ENDOSCOPIC RESECTION OF CLIVAL CHORDOMA. A CASE REPORT
GAMBÔA I ET AL

sospechosa debe llevarse a cabo con prontitud. En este caso clínico, se presentan el abordaje quirúrgico y el seguimiento postoperatorio.

PALABRAS CLAVE: cordoma; clivus; resección endoscópica; tumor maligno.

INTRODUCTION

Chordoma is a rare malignant tumor, accounting for about 4% of all malignant primary bone tumors [1]. It originates from remnant cells of primitive notochord, primary involving the caudal and cephalic ends of the vertebral column [2]. The most frequent locations are the sacrococcygeal and clival regions, and presents as a soft tissue mass with slow growth and progressive invasion, leading to regional bone erosion [3]. It is usually diagnosed only when the tumor causes symptoms by compression or invasion of adjacent structures [3]. Local recurrence is not uncommon, and the recommended treatment is surgical resection followed by radiotherapy [4]. We report the case of a patient with a clival chordoma, diagnosed as an accidental finding in a paranasal sinuses imaging study. The surgical approach and the postoperative follow-up are presented.

DESCRIPTION

A male patient, 30 years old, without relevant past medical history, presented at the Otolaryngology department with symptoms of long-term nasal obstruction and frontal weight sensation. There was no history of rhinorrhea, epistaxis, nasal pruritus, headache or ocular changes. On nasal endoscopy, a deviated nasal septum causing nasal obstruction and enlarged inferior turbinates were apparent. As he complained of frontal weight sensation, rhinosinusitis was suspected, and so a Computed Tomography (CT) of paranasal sinuses was requested to investigate sinus pathology. CT scan showed a deviated nasal septum with a right bone spur, and a 10mm soft tissue mass in the right posterior wall of the sphenoid sinus with underlying bone erosion (Figure 1, A and B). To clarify the etiology of the lesion, a Magnetic Resonance Imaging (MRI) was requested, which confirmed a mass in the clivus, with no contrast enhancement, and without invasion of intracranial structures (Figure 1, C and D). The patient underwent surgical intervention for

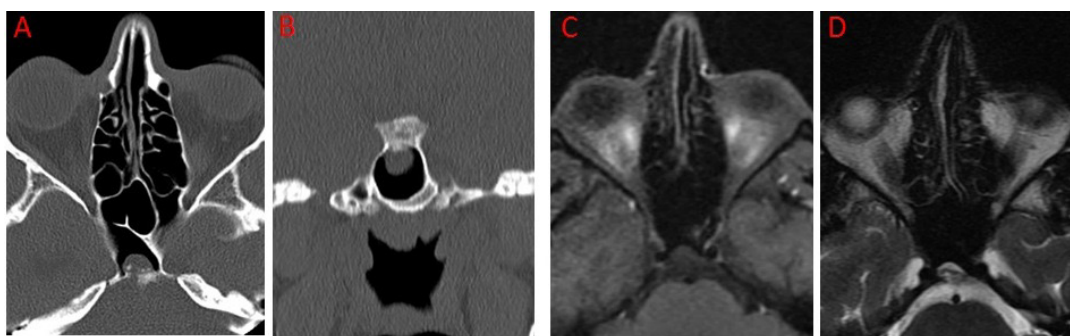


Figure 1. A and B: CT scan of the paranasal sinuses (A axial, B coronal) - Nasal septum deviation; soft tissue mass with rounded contours in the posterolateral wall of the right sphenoid sinus, with areas of regional bone erosion. C and D: MRI of the paranasal sinuses, axial sections (C-T1 with contrast; D-T2) - Soft tissue mass in the posterior wall of the sphenoid sinus/clivus, emerging from the pre-pontic cistern; It does not show markedly contrast enhancement; There are no images suggesting meningoencephalic or vascular malformative lesions.

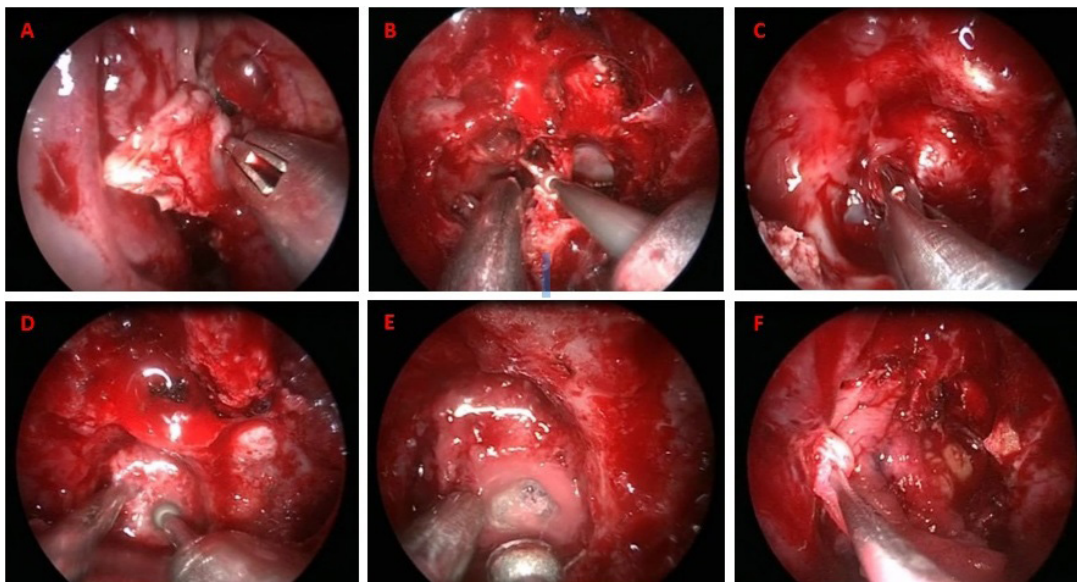


Figure 2. Surgical procedure - After insertion of a 0° endoscope, the posterior third of the nasal septum was resected (A), for sphenoid exposure (B). The sphenoid sinus was entered, and inflammatory mucosa and residual chordoma in the clivus region was removed (C). The bone erosion region was drilled (D) until duramater exposure (E). Closure was carried out with a flap of contra-lateral medium turbinate, abdominal fat and Duraform®(F).

septal deviation and nasal obstruction septoplasty and inferior turbinoplasty and at the same surgical time, an endoscopic transsphenoidal biopsy of the clival lesion was performed. The histopathological result was compatible with chordoma. There was no evidence of regional or distant metastatic disease. After presentation and discussion of the case at a multidisciplinary team and reevaluation with MRI, the patient was proposed for surgical treatment endoscopic transnasal resection of the clival chordoma.

Surgical approach. After insertion of a 0° endoscope, the posterior third of the nasal septum and the sphenoid septum were resected for clivus exposure. Nasoseptal flap was not collected because of the possibility of flap inviability due to previous septoplasty. The sphenoid sinus was entered, and inflammatory mucosa and residual chordoma were removed. The bone erosion region was drilled until duramater exposure. A low volume liquor fistula occurred. Closure was carried out with a flap of

contralateral medium turbinate, abdominal fat and Duraform®. (Figure 2, A to F).

Postoperative follow-up. There were no interurrences in the immediate postoperative period. The patient was discharged after 3 days of hospitalization, and was later evaluated in routine consultations, with only minor nasal crusts. The histopathology of the residual tumor removed was compatible with chordoma. The patient underwent postoperative radiotherapy (RT), having a total of 60 Gy in 30 fractions for 3 months. MRI performed 6 months after RT, 1 year after surgery, showed signs of surgical intervention with sphenoid sinus filled with the flaps used intraoperatively, with no evidence of residual lesion (Figure 3).

DISCUSSION

Chordoma is a malignant neoplasm that should be included in the differential diagnosis of clival lesions, especially when there is regional bone



Figure 3. A- Preoperative MRI, sagittal cut, showing the initial clival lesion (arrow). B and C- MRI 1 year after surgery, where signs of surgical intervention are observed, with sphenoid sinus filled with intraoperative flaps used, without evidence of residual of lesion.

erosion [5]. It should be noted that about 40% of chordomas are in the clival and para-clival regions [6].

These tumors have a slow growth pattern, with propensity for local invasion. Generally, symptoms like headaches or cranial nerve palsies occur when the tumor reaches considerable dimensions, and so the diagnosis is made often in advanced stages [3]. Distant metastasis can occur in about 30% of chordomas, more frequently in non-clival chordomas. Most frequent sites include lung, bone, liver and lymph nodes [3].

Differential diagnoses include ecchordosis physaliphora and benign notochordal cell tumor (BNCT), both benign lesions derived from remnant cells of the primitive notochord and primarily involving the clival region [6,7]. BNCT may also undergo malignant transformation to classic chordoma. On MRI, these two benign tumors most often do not show contrast enhancement, which usually occurs in chordomas [6]. However, contrast enhancement is not pathognomonic of chordoma, and the differential diagnosis with these two benign entities can't be definitive only with image studies. As so, suspected clival lesions must be biopsied for a definitive diagnosis [6,7].

The recommended treatment for clivus chordomas is complete surgical resection and postoperative RT [4,9]. In the present case, tumor location and dimensions allowed an exclusively endoscopic approach. Complete resection of clivus lesions may be difficult since their anatomical location is near noble structures of the vascular and central nervous system [4]. With respect to the surgical technique used in this anatomic region, in recent years there has been an increasing use of transnasal endoscopic approaches instead of external approaches, with similar results in tumor removal but lower morbidity for the patient [9]. In fact, some authors even point out that in this anatomical location, the use of the endoscope improves tumor visualization allowing a more detailed recession [4]. Contraindications for an exclusively endoscopic approach include tumor extension to the middle fossa or cavernous sinus [4,10].

Postoperative RT is recommended, which according to the literature is effective in the control of minor residual lesions after surgery [4]. In the present case, the conventional RT modality was used, due to the unavailability of other methods in our country's institutions. There has been debate on the most effective RT modality (conventional, proton beam, intensity- modulated), but recent

studies didn't not show significant differences in disease control with the different modalities [4,9]. Despite this, many authors advocate the use of proton beam radiotherapy for the ability to deliver a higher dose of radiation without increase damage in surrounding structures [1,4].

In the present case, the clivus lesion was an accidental finding, and although with no contrast enhancement on MRI (which usually occurs in the chordoma) the underlying bone erosion has alerted us to the possibility of a malignant lesion. In the suspicion of a malignant lesion, it is preponderant to perform a surgical biopsy for confirmation, since the definitive diagnosis of chordoma is histological [6]. These tumors usually present in advanced stages, causing symptoms by invasion of neighboring structures by tumor growth [3]. In this case, the accidental finding of the lesion at an early stage allowed a timely treatment with good results one year after its conclusion.

CONCLUSIONS

Chordoma is one of the most frequent malignant tumors in the clivus region [3]. Differential diagnoses include ecchordosis physaliphora and BNCT, both benign lesions, that not always can be differentiated from chordoma in image studies [6]. As so, biopsy of suspected lesions in the clival region should be promptly performed to begin the adequate treatment. In the present case, the finding of the lesion at an early stage, without symptoms, allowed a timely treatment with surgery and post-operative RT, with good response one year after the surgical treatment. A long follow-up period is necessary because these tumors tend to recur [4].

ACKNOWLEDGMENTS

Dr. Mário Resende, for the collaboration.

REFERENCES

1. Kim JW, Suh CO, Hong CK, Kim EH, Lee IJ, Cho J, et al. Maximum surgical resection and adjuvant intensity-modulated radiotherapy with simultaneous integrated boost for skull base chordoma. *Acta Neurochir (Wien)*; 2016 DOI:10.1007/s00701-016-2909-y
2. Tan NC, Naidoo Y, Oue S, Alexander H, Robinson S, Wickremesekera A, et al. Endoscopic surgery of skull base chordomas. *J Neurol Surg B Skull Base*. 2012; 73:379-86.
3. Heery CR. Chordoma: The Quest for Better Treatment Options. *Oncol Ther*. 2016;4:35-51.
4. Shimony N, Gonen L, Shofty B, Abergel A, Fliss DM, Margalit N. Surgical resection of skull-base chordomas: experience in case selection for surgical approach according to anatomical compartments and review of the literature. *Acta Neurochir (Wien)*. 2016 Dec 6. DOI:10.1007/s00701-016-3032-9.
5. Yuh SJ, Woulfe J, Corsten MJ, Carrau RL, Prevedello DM, Kassam AB. Diagnostic imaging dilemma of a clival lesion and its clinical management implications. *J Neurol Surg B Skull Base*. 2014;75:177-82.
6. Golden LD, Small JE. Benign notochordal lesions of the posterior clivus: retrospective review of prevalence and imaging characteristics. *J Neuroimaging*. 2014;24(3):245-9.
7. Bolzoni-Villaret A, Stefini R, Fontanella M, Bottazzoli M, Turri Zanoni M, Pistochini A, et al. Transnasal endoscopic resection of symptomatic ecchordosis physaliphora. *Laryngoscope*. 2014 Jun;124(6):1325-8.
8. Labidi M, Watanabe K, Bouazza S, Bresson D, Bernat AL, George B, et al. Clivus chordomas: a systematic review and metaanalysis of contemporary surgical management. *J Neurosurg* 2016; 60:476-84.
9. Amit M, Na'ara S, Binenbaum Y, Billan S, Svirin G, Cohen JT, et al. Treatment and Outcome of Patients with Skull Base Chordoma: A Meta-analysis. *J Neurol Surg B Skull Base*. 2014; 75:383-90.
10. Koechlin NO, Simmen D, Briner HR, Reisch R. Combined transnasal and transcranial removal of a giant clival chordoma. *J Neurol Surg Rep*. 2014;75(1): e98-e102.

