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A Rare Case of Clival Hemangioma Simulating Chordoma

Um caso raro de hemangioma clival simulando cordoma

Gustavo Rassier Isolan[1] Alessandra Locatelli Smith[2] Camila Meotti[2] João Paulo Mota Telles[3] Nícollas Nunes Rabelo[3] Eberval Gadelha Figueiredo[3]

[1] The Center for Advanced Neurology and Neurosurgery, CEANNE, Porto Alegre, RS, Brazil
[2] Hospital de Clínicas de Porto Alegre, Porto Alegre, RS, Brazil
[3] Department of Neurosurgery, University of São Paulo, São Paulo, SP, Brazil

Address for correspondence Eberval Gadelha Figueiredo, MD, PhD, Department of Neurosurgery, University of São Paulo, Rua Eneas Aguiar, 255, São Paulo, Brazil. Zip code: 05403-010 (e-mail: ebgadelha@yahoo.com).

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Abstract

Primary intraosseous hemangiomas are rare, benign, vascular malformations that account for ~1% of all primary bone neoplasias. A 59-year-old female patient with unknown comorbidities had a history of headache, visual impairment and dizziness that led to the diagnosis of a clivus tumor. Two resections were attempted through transcranial and transnasal transsphenoidal approach in the last two years in another hospital. The initial MRI scan showed an expansive lesion with T2 hyperintense signal and diffuse, heterogenic contrast enhancement. Clival chordoma was the main diagnostic hypothesis done. A CT scan was performed to evaluate the extent of clival invasion, the sinus anatomy, and the clival destruction - all simulating clival chordoma. The interdisciplinary tumor board decided to proceed with endoscopic endonasal tumor resection. There were no postoperative complications and the histopathological analysis revealed a primary intraosseous haemangioma. Skull base intraosseous hemangiomas are rare entities, with a limited number of case reports found after literature reviews, especially in the clival region. The clinical pattern and imaging characteristics can vary widely according to the tumor extension and development, simulating some other common tumors found at this topography. We present a case report of a clival intraosseous hemangioma presenting as an isolated abducens paresis with a positive outcome after intranasal endoscopic resection after two years of follow-up.

Keywords
- chordoma
- hemangioma
- neurosurgery
- tumor

Resumo

Os hemangiomas intraósseos primários são malformações vasculares benignas raras que representam cerca de ~1% de todas as neoplasias ósseas primárias. Uma paciente de 59 anos do sexo feminino com comorbidades desconhecidas história de cefaleia deficiência visual e tontura que levou ao diagnóstico de tumor de clivus. Duas ressecç ões foram tentadas por via transcraniana e transesfenoidal transnasal nos dois anos anteriores ao presente estudo em outro hospital. A ressonância magnética inicial

Palavras-chave
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mostrou lesão expansiva com sinal hiperintenso em T2 e realce difuso e heterogêneo pelo contraste. Cordoma clival foi a principal hipótese diagnóstica. Uma tomografia computadorizada foi realizada para avaliar a extensão da invasão clival a anatomia do seio e a destruiç ão clival - todas simulando cordoma clival. A comissão interdisciplinar do tumor decidiu prosseguir coma ressecç ão endoscópica do tumor endonasal. Não houve complicaç ões pós-operatórias e a análise histopatológica revelou hemangioma intraósseo primário. Os hemangiomas intraósseos da base do crânio são entidades raras com número limitado de relatos de casos encontrados após revisões da literatura principalmente na região clival. O padrão clínico e as características de imagem podem variar amplamente de acordo com a extensão e desenvolvimento do tumor simulando alguns outros tumores comuns encontrados nesta topografia. Apresentamos um relato de caso de hemangioma clival intraósseo apresentando-se como uma paresia isolada do abducente com evoluç ão positive após ressecç ão endoscópica intranasal e dois anos de acompanhamento.

Introduction

Primary intraosseous hemangiomas are benign, vascular malformations that account for ~1% of all primary bone neoplasias. These tumors are commonly found in the calvarium and vertebral bones but are distinctly unusual in the skull base, where they represent 0.2% of bony neoplasias and 10% of benign skull tumors.[1] We present a case of primary intraosseous hemangioma of the clivus simulating a chordoma and a review of the pertinent literature.

Case Report

A 59-year-old female patient with unknown comorbidities had a history of headache, visual impairment and dizziness that led to the diagnosis of a clivus tumor. Two resections were attempted through transcranial and transnasal transsphenoidal approach in the last two years in another hospital. She reported no neurological symptoms after those interventions. Months later presented with a new isolated right abducens paresis at the emergency service of a tertiary hospital. The patient was awake, fully oriented and pupils were equal, reactive to light and accommodation. There were no signs of meningeal irritation, no other cranial nerves affected. The remaining examination showed no abnormalities except for right abducens paresis.

The initial MRI scan (►**Fig. 1**) showed a $3.6 \times 3.4 \times 3.4$ cm expansive lesion with T2 hyperintense signal and diffuse, heterogenic contrast enhancement. The geometric center was sitting at the basal portion of the sphenoidal bone (clivus), with no cavernous sinus invasion. The mass expanded within the clivus infiltrating the sphenoidal sinus anteriorly and the pre-pontine cistern and clinoid processes posteriorly, where it touched the ventral portion of the pons and basilar artery. Cranially, the lesion expanded through the pituitary gland and anteriorly to the clinoid process. The cavernous sinus, internal carotid and petrous apex were involved, including the optic canal.

Clival chordoma was the main diagnostic hypothesis done. A CT scan was performed to evaluate the extent of clival invasion, the sinus anatomy, and the clival destruction - all simulating clival chordoma. The interdisciplinary tumor board decided to proceed with endoscopic endonasal tumor resection. There were no postoperative complications and the histopathological analysis revealed a primary intraosseous haemangioma (►**Fig. 2**).

The right abducens paresis regressed after the first year of follow up. A new MRI scan performed 6 months after surgery showed near-total resection. The patient is currently in the second year of follow up with no neurological symptoms and the small enhancement at the lower clivus remains stable.

Discussion

Primary intraosseous hemangiomas (PIH) of the skull base are extremely rare tumors. They appear typically in the parietal and frontal bones of the calvarium, whereas location in the craniofacial bones is less common (zygoma, maxilla, mandible, and vomer).[2] The particular clinical evolution and especially the risks inherent to the surgical approach make the management of skull base intraosseous hemangiomas significantly different from those arising in cranial vault bones. Furthermore, an additional challenge in preoperative diagnosis is due to the lack of cases reported in the literature and to the fact that this type of lesion can simulate other

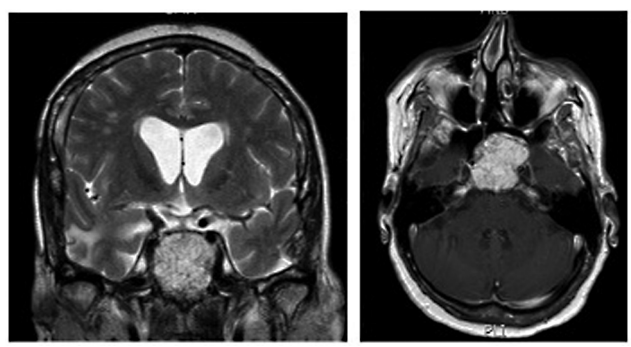

Fig. 1 T2 coronal and T1-weigthed axial images revealing a hyperintense lesion with diffuse enhancement after gadolinium injection.

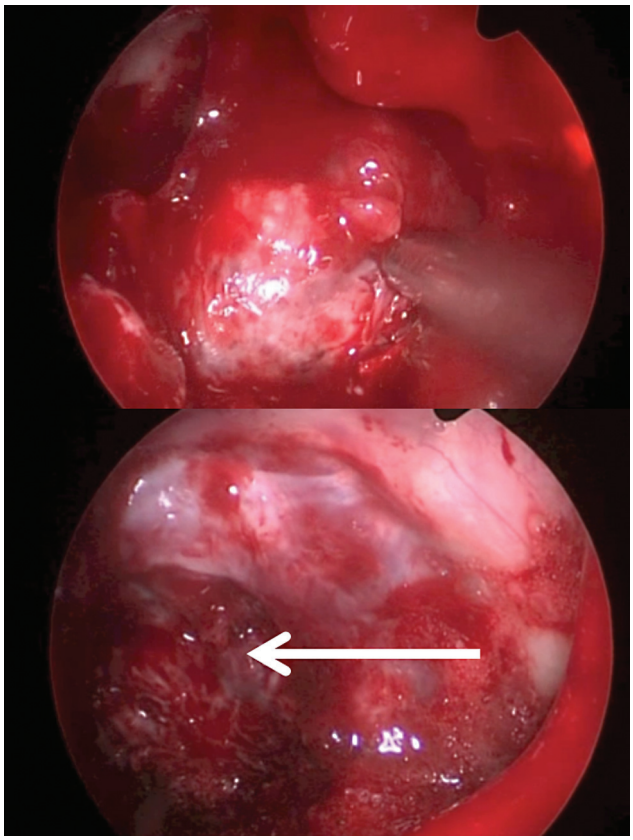

Fig. 2 Intraoperative endoscopic view before (above) and after (below) tumor resection.

common lesions of the skull base (i. e., clival chordoma, pituitary adenoma).[3]

The few published cases of clival PIH tell us that while these tumors can present with progressive cranial nerve deficits, they usually expand extracranially with minimal symptomatic manifestations. Neurological deficits are unusual. However, when intracranial extension is present, they tend to grow very slowly, leading to headaches, visual impairment or other cranial nerve deficits. Additionally, skull base intraosseous hemangiomas may involve neighboring structures, including cavernous sinus and carotid artery.[4] Interestingly, our patient presented with an isolated right abducens paresis - the same pattern of presentation that was recently described by Serrano et al.[5] In their case, the paresis remitted during the early postoperative follow-up, suggesting that opening the tumor cavity relieved the intratumoral pressure and, therefore, improved its compressive effect on the abducens nerve. In our case, however, the paresis improved after almost one year of postoperative follow-up, an outcome reported in no other case throughout the literature.

PIH are usually solitary tumors most commonly found in women between the second and fifth decades. Histologically, they can be classified as cavernous or capillary hemangiomas. Cavernous ones are composed of large dilated blood vessels separated by fibrous tissue, whereas capillary ones lack fibrous septa and have smaller vascular lumens. Calvarial hemangiomas are usually of the cavernous type, whereas vertebral hemangiomas are most frequently of the capillary type.[6]

Concerning the radiological features, there is little difference between the vertebral, commonly found hemangiomas and the skull base ones. The imaging study of choice is the MRI scan, but the characteristics can change in both T1 and T2-weighted sequences according to the amount of venous flow and fatty transformation within the tumor. Lesions with more significant fat content present high signal intensity on T1-weighted images, whereas larger lesions tend to have lower signal intensity. On T2-weighted images, high signal intensity may be caused by the pooling of blood or slow-flowing blood.[2] Cavernous hemangiomas typically enhance after administrations of gadolinium contrast.[7] As in our case, the radiographic findings are nonspecific and can mimic various common lesions of the skull base. Consequently, the diagnosis is most often made after surgical resection and histopathological analysis. In terms of surgical planning, the CT scan is more helpful than the MRI, for the first can evaluate more accurately where the extent of the lesion, the nasal and sinus endoscopic possibility approach on the bone windows.

Reports of PIH show they have a slow growth tendency, but they do not seem to show involution. Therefore, symptomatic tumors should be removed “en bloc” when possible, because the recurrence after this approach is uncommon.[8,9] In our case, the tumor was resected progressively through transnasal and extradural approaches and had no identified adherence to the meninges. Macroscopically, the gross tumor removal was achieved, even though the MRI after surgery may suggest some residual tumor. Once the transnasal approach was selected, a pedicled nasoseptal vascularized flap was used over the dura mater due to the risk of cerebrospinal fluid (CSF) fistula, even though this event was not observed during surgery. The patient remained with no signs or symptoms of CSF leak in the postoperative period.

Relevant differential diagnoses of clivus lesions include chordoma, chondroma, chondrosarcoma, osteosarcoma, osteoblastoma and metastasis. These lesions have differential imaging features and also more aggressive clinical presentations - particularly chondrosarcomas, osteosarcomas and metastases. (2). Finally, the definitive diagnosis is often obtained after biopsy and histological examination of the resection specimen by an experienced pathologist aware of the histopathological features found in PIH tumors. The choice of the surgical technique is of particular importance – in our case, conventional pterional approaches that had been attempted in two institutions were unsuccessful in removing the macroscopic lesion.

In terms of adjuvant therapy for PIH, some authors have suggested that fractionated radiotherapy can be offered as an alternative in patients with partial resection, as well as in high-risk elderly patients.[3] However, as these tumors usually grow slowly, a conservative follow-up of residual lesions is often more reasonable than to submit a patient to radiation and its well-known side effects.[10,11] Moreover, the proximity of the lesions to the cavernous sinus - of which the present case is an excellent example - increases the risk of radiation-

induced cranial nerve deficit and hypopituitarism. Once we obtained complete resection in our patient, no adjuvant treatment was prescribed and no neurological symptoms or deficits have been related during the post-operative period.

Conclusion

Skull base intraosseous hemangiomas are rare entities, with a limited number of case reports found after literature reviews, especially in the clival region. The clinical pattern and imaging characteristics can vary widely according to the tumor extension and development, simulating some other common tumors found at this topography. We present a case report of a clival intraosseous hemangioma presenting as an isolated abducens paresis with a positive outcome after intranasal endoscopic resection after two years of follow-up. Additionally, we discuss key imaging and clinical particularities of this rare type of tumor.

Note

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