

Aneurysmal Bone Cyst of the Skull Base—Case Report

Cisto ósseo aneurismático da base do crânio—relato de caso

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Arq Bras Neurocir 2019;38:51–55.

Abstract

Introduction Aneurysmal bone cysts (ABCs) are pseudotumoral bone lesions of unknown etiology that are also hypervascularized, benign, and locally destructive. They are rare in the base of the skull. The present case report describes a case of aneurysmal bone cyst in the sella turcica.

Case Report The present study was developed at the department of neurosurgery of the Hospital Universitário Professor Alberto Antunes of the Universidade Federal de Alagoas (HUPAA-AL, in the Portuguese acronym), Maceió, state of Alagoas, Brazil, and is accompanied by a review of the literature from the PubMed database. A 17-year-old female patient with bitemporal hemianopia and intense left hemicranial headache associated with symptoms from the cranial nerves contained in the cavernous sinus. Neuroimaging evidenced a large lesion in the suprasellar region with calcification foci, sellar erosion, and extension to the cavernous sinus. The patient was submitted to a partial lesion resection and the histopathological analysis showed an aneurysmal bone cyst.

Conclusion A rare case of intracranial aneurysmal bone cyst, with the important differential diagnosis from pituitary adenoma.

Keywords

- aneurysmal bone cysts
- sella turcica

Resumo

Introdução Cistos ósseos aneurismáticos (COAs) são lesões ósseas pseudotumorais, de etiologia desconhecida, hipervascularizadas, benignas, localmente destrutivas. Cistos ósseos aneurismáticos são raros na topografia da base do crânio. O objetivo do presente relato de caso é descrever um caso de cisto ósseo aneurismático localizado na sela túrcica.

Relato de caso O caso foi acompanhado no Serviço de Neurocirurgia do Hospital Universitário Professor Alberto Antunes da Universidade Federal de Alagoas (HUPAA-AL), Maceió, AL, Brasil, e sua descrição foi feita conforme dados encontrados em revisão de literatura realizada por meio do banco de dados PubMed. Paciente do sexo feminino, 17 anos, com hemianopsia bitemporal e intensa cefaleia hemicraniana esquerda

received
January 28, 2016
accepted
April 21, 2016

DOI <https://doi.org/10.1055/s-0036-1584508>.
ISSN 0103-5355.

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Palavras-chave

- cistos ósseos aneurismáticos
- sela túrcica

associada à lesão dos nervos cranianos contidos no seio cavernoso. Exames de neuroimagem evidenciaram uma lesão extensa na região supraselar com focos de calcificação e erosão da sela túrcica, e com extensão para o seio cavernoso. Tratamento com ressecção parcial da lesão, a qual o exame histopatológico revelou ser cisto ósseo aneurismático.

Conclusão Caso raro de cisto ósseo aneurismático intracraniano, sendo importante o diagnóstico diferencial com adenoma hipofisário.

Introduction

Aneurysmal bone cysts (ABCs) are pseudotumoral bone lesions of unknown etiology that are also hypervascularized, benign, and locally destructive; in addition, they often grow progressively. The literature reports an incidence of 0.14 per 100,000 individuals, corresponding to ~ 1% of all bone tumors.¹ The long bones and the spine are the most commonly affected sites. Aneurysmal bone cysts are rare and more prevalent in children and in young people; they can evolve asymptotically, or rapidly cause pain and neurological symptoms. The present case report describes a case of ABCs unusually located in the sella turcica.

Material and Methods

The present case report was described by the authors from the Neurosurgery Service of the Hospital Universitário Professor Alberto Antunes of the Universidade Federal de Alagoas (HUPAA-AL), Maceió, state of Alagoas, Brazil. The informed consent form was signed by a legal representative of the patient. The literature review was performed at the PubMed database up to December 2015.

Case Report

The patient J. F. S., female, 17 years old, was admitted to the HUPAA-AL with a clinical history (~ 7 months) of mild to moderate hemicranial headache associated with visual field deficits. The condition worsened after 4 months, with severe left hemicranial headache. Upon admission, at the neurological examination, the patient was awake, a little confused, with no limb motor and/or sensory deficit, presenting left-side ptosis, anisocoric pupils (left > right), direct and consensual photomotor reflex absent on the left side and present on the right side, diplopia, fourth and sixth cranial nerve palsy on the left and right side, respectively, and bitemporal hemianopsia at the confrontation test, confirmed with a visual field test. After admission, the patient progressed with a stable clinical/neurological condition until she presented with an episode of psychomotor agitation associated with psychotic symptoms and delirium. She was submitted to a computed tomography (CT) of the head, which showed a sellar lesion with expansion and bilateral involvement of the cavernous sinus (►Fig. 1) and erosion of the upper portion of the clivus (►Fig. 2) and of the sellar floor (►Fig. 3). A magnetic resonance imaging (MRI) of the skull was not available during the hospitalization of the patient.

The results of routine laboratory tests, as well as of hormonal tests requested due to a suspected diagnosis of pituitary adenoma, were within the normal range.

A transsphenoidal approach aided by a surgical microscope was selected. The sellar and suprasellar lesion was partially resected. Maximal resection was not possible due to the location of the lesion and to the occurrence of major bleeding during the procedure.

In the postoperative period, the patient maintained the same clinical picture, with no signs of cerebrospinal fluid (CSF) fistula and/or incisional bleeding. The results of the anatomopathological study revealed a fibrovascular tissue composed of fusiform and epithelioid cells (giant cells) with fibrous tissue struts and hematic content consistent with an aneurysmal bone cyst (►Figs. 4 and 5).

The patient was lost at follow-up and the treatment outcome is unknown.



Fig. 1 Contrast-enhanced, axial head computed tomography. Expansive process at the base of the skull with sellar and parasellar regions involvement, cavernous sinuses invasion and apical clival and clinoid erosion; this lesion presents mixed density, predominantly with peripheral hyperdensity and central hypodensity.

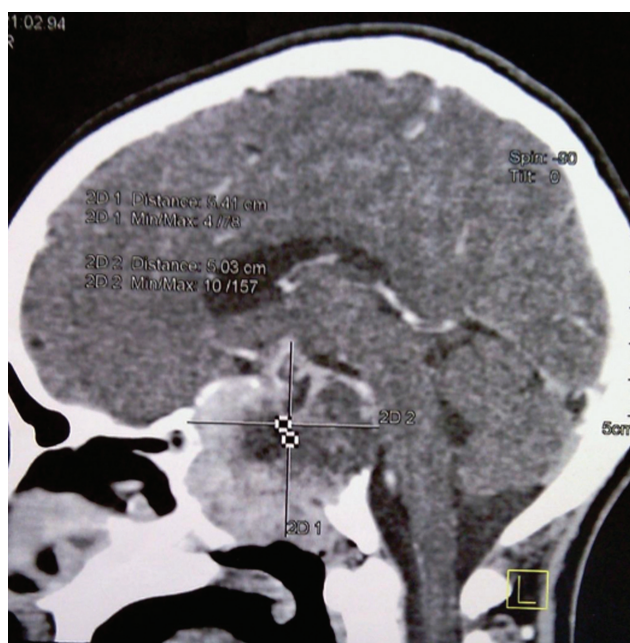


Fig. 2 Contrast-enhanced, sagittal head computed tomography. Expansive process of massive volume eroding the sella turcica and the base of the skull with inferior extension to the sphenoidal sinus and progressing to the clivus and cavum region.

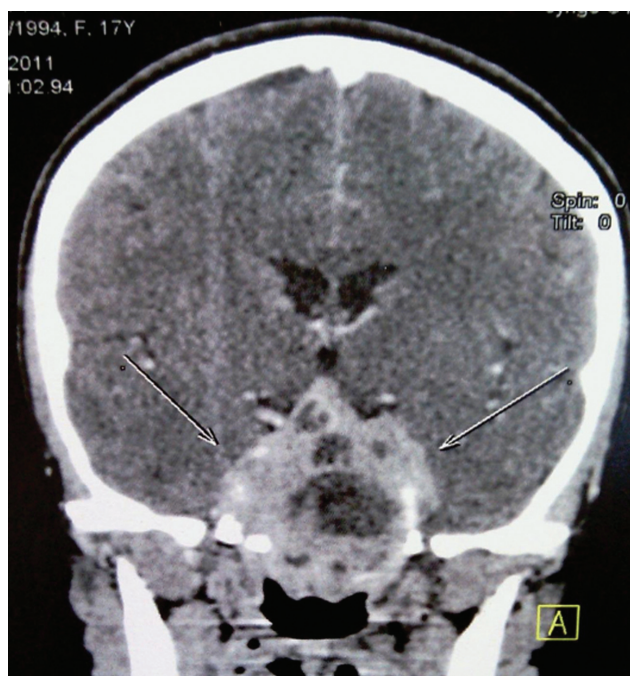


Fig. 3 Contrast-enhanced, coronal head computed tomography showing a tumor mass with invasive aspect, bilateral involvement of the cavernous sinuses and superior extension to the optic-chiasmatic cistern.

Literature Review

First described by Jaffe et al in 1942, ABCs are pseudotumoral bone lesions of unknown etiology that are hypervascularized, benign, locally destructive and grow progressively.² According to Lipman et al and to Biesecker et al, ABCs are non-neoplastic lesions composed of cystic cavities with

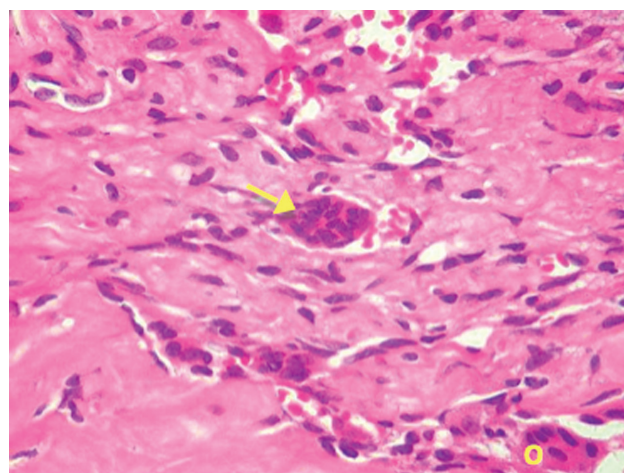


Fig. 4 This histological section (magnification 100x) shows an epithelioid cell (arrow), osteoclasts (O) and fusiform cells. Hematoxylin and eosin stain.

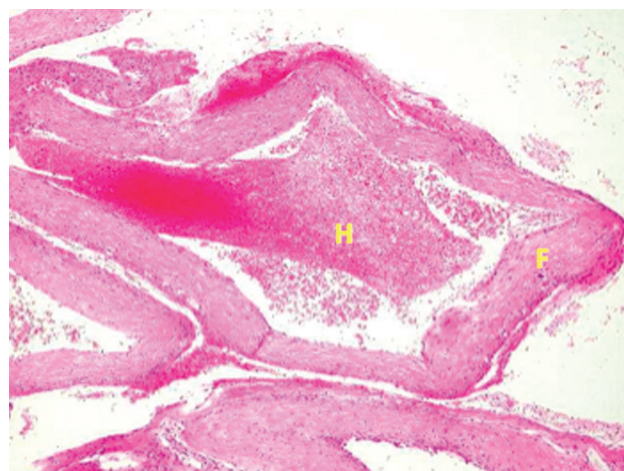


Fig. 5 This photograph shows the hematic content (H) within the fibrous septa (F). Magnification 40x. Hematoxylin and eosin stain.

fibrous walls and free blood circulation without endothelial delimitation.^{3,4} Some older studies suggested that the origin of the lesion was the hemorrhagic degeneration of preexisting bone lesions (including giant cell tumor, osteoblastoma, fibrous dysplasia and chondromyxoid fibroma).^{2,5,6} Isolated lesions are called primary lesions, whereas lesions arising from previous lesions are classified as secondary lesions.⁷ In the present case, the nature of the lesion, whether primary or secondary, could not be established.

Lesions are macroscopically described as lytic bone structures with a thinner, brownish and fragile cortex (hematopoietic content),⁸ and fibrous walls resembling soap bubbles.⁹ Under light microscopy and routine staining (hematoxylin and eosin), cystic structures with fibrous or osseous septations containing fragile, hemorrhagic vascular tissue are described.¹⁰ Some authors reported that the cyst is surrounded by hemosiderin, fibroblasts, giant cells, and stroma cells, but not by endothelium;^{3,11} however, O'Brien et al, in 1994, reported the presence of endothelium and giant cells.¹² Since routine staining is sufficient for the diagnosis, immunohistochemical analysis is not required.^{7,12}

Aneurysmal bone cysts are more commonly found in young individuals, and ~ 80% of the cases are diagnosed during the first 2 decades of life,¹³ with a slight prevalence in female patients.^{14,15} The most common sites for ABCs are the tibia, the femur, the humerus, the spine, and the pelvis.^{5,6} Few studies show intracranial lesions, observed in only between 2 and 6% of all of the cases; the calvaria is the most common affected site in the skull.^{16,17} The bones most commonly affected in the skull are the temporal (22%), the occipital (20%), the frontal (14%), the parietal (11%), the orbit (11%), the sinus (5%), the sphenoidal sinus, and the ethmoid bones (3%).^{18–20} Only 15 cases located in the ethmoid bone were described up to 2014,^{21–32} and 15 cases located in the sphenoid bone^{26,30,33} were described up to 2015.

In some cases, the diagnosis of aneurysmal bone cyst is difficult because the radiological lesions are similar to other benign or malignant conditions, especially in unusual sites such as the base of the skull.^{20,34}

In the present case, an important differential diagnosis to be considered at the neurological examination would be hypothalamic-pituitary-axis lesions. However, the radiological aspect of the lesion was a massive cystic formation with a solid portion spontaneously denser than the cerebral parenchyma and sellar bone erosion,³⁵ not corresponding to the description of the neuroimaging aspect of most pituitary macroadenomas. These lesions are homogeneous, and their density is similar to the parenchyma, except in cases with apoplexy and posthemorrhagic cystic cavities formation, which become similar to the one reported here.

Another differential diagnosis regarding imaging and topography would be craniopharyngioma, whose formation is similar to a cystic cavity; however, the aneurysmal bone cyst has thicker and irregular walls, formed by fibrous septations,¹⁰ while the craniopharyngioma is a thin-walled cystic formation with calcification at the sellar base.

Treatment is performed by lesion curettage, followed by the injection of bone substitutes such as polymethylmethacrylate. Until the 1990s, the procedure included only lesion curettage; however, it was found that grafting reduced the recurrence rate from 26 to 17%, as mentioned by Mankin et al in 2005. These authors reported a recurrence rate in their own series of ~ 22%,³⁶ but other investigators found values ranging from 5 to 40%,³⁷ regardless of the approach.

Currently, the treatment of choice, whenever possible, is the complete lesion resection preceded or not by embolization.⁷ Unfortunately, in the present case, this was impossible due to the location at the base of the skull and to the occurrence of a massive intraoperative bleeding. Some authors recommend radiotherapy, although its recurrence rate is > 30%,⁷ while others contraindicate it due to the risk of sarcomatous degeneration.²⁴ There are also reports of interferon α -2a use in lesions considered unresectable.²⁹

Conclusion

The present case report brings to light the knowledge of aneurysmal bone cysts in unusual topographies, such as the base of the skull, requiring its differential diagnosis with

typically hypothalamic-pituitary-axis lesions, such as pituitary adenoma and craniopharyngioma. Although the treatment of choice is complete resection, it is not feasible at this location.

Conflicts of Interest

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