

# Aneurysmal bone cyst secondary to fibrous dysplasia of calvaria: case report

## Abstract

We report a case of fibrous dysplasia with the development of secondary aneurysmal bone cyst of skull, a rare entity, with approximately 20 reports described in the literature. Despite particularly rare, it is valuable for the radiologist to comprehend this hypothesis and specially the diagnostic reasoning to narrow the differential diagnosis of similar lesions.

**Keywords:** fibrous dysplasia of bone, bone cysts, aneurysmal, skull, tomography, x-ray computed

Volume 6 Issue 2 - 2019

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**Received:** March 09, 2019 | **Published:** March 19, 2019

## Introduction

Fibrous dysplasia (FD) is a developmental disease in which the normal bone marrow is replaced by fibrous tissue due to fibroblast proliferation. It represents 2.5% of all bone tumors and 7% of benign bone tumors and arises mostly in adolescents and young adult patients.<sup>1-3</sup>

Aneurysmal bone cyst (ABC) is a rare benign, expansive and lytic bone lesion, constituted by irregular lacunas containing varying density hematic content.<sup>1</sup> ABCs occur mostly in long bones and the spinal column, and craniofacial involvement is a very rare occurrence. ABCs are either primary or secondary; secondary ABCs develop over preceding bone pathologies such as giant cell tumor, osteoblastoma, angioma and chondroblastoma and less commonly including FD, ossifying fibroma and osteosarcoma.<sup>3</sup> Reports of secondary ABC occurring in craniofacial FDs are exceptionally rare in the literature, accounting for about 20 cases.<sup>3</sup> However, it is a hypothesis that adds value to the differential diagnosis of the radiologist.

## Case report

A 55-year-old man presented with a history of falling from his own height today, with mild traumatic brain injury (TBI) at the occipital region. He reported bone prominence since childhood (without previous investigation), with a hump after TBI. No neurological complaints.

The radiograph (Figure 1) showed a predominantly lytic lesion in the interest region, relatively circumscribed, with no suspected periosteal reaction. The investigation was continued with computed tomography (CT).

CT (Figure 2) exhibited a predominantly expansive lytic insufflative lesion, measuring approximately 4.5 x 2.2 x 4.2cm, located in the left and right parietal and occipital bones, associated with adjacent bone textural alteration with "ground glass" appearance (compatible with fibrous dysplasia). In its interior, we visualize liquid levels. It determines changes in the external contours of the cranial vault as well as compression of the adjacent brain parenchyma, but there was no change in the brain attenuation coefficients or significant deviation of the midline structures, due to chronic accommodation.

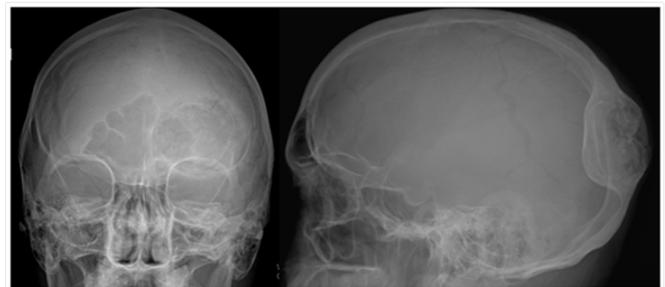


Figure 1 Lateral radiograph.

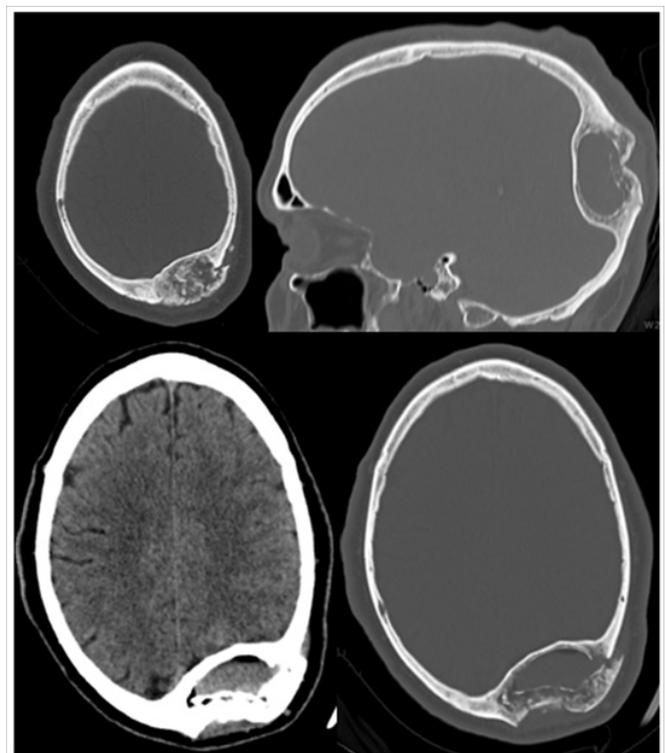


Figure 2 CT.

There was also pathological comminutive fracture (due to local greater susceptibility in the trauma scenario determined by the lesion) of the outer table at the lesion, with preservation of the inner table; as well as increasing volume and densification of adjacent soft tissue. There was no sign of additional bone lesions except the calvarial one at the radiographs performed by the patient. The hypothesis of ABC associated to FD of calvaria was raised. The patient was referred to the local referral service. Histopathological findings later corroborated the hypothesis of FD combined with ABC.

## Discussion

ABC secondary to FD is an extremely rare entity, first described at calvaria by Branch et al.<sup>4</sup> The first diseases known to be associated with ABC were osteoclastoma, osteosarcoma, osteoblastoma and Hemangioma.<sup>4,5</sup>

FD can present as classic “ground glass” pattern (mostly seen but not isolated), associated with diploic bone non-aggressive remodeling and thickening; although it may manifest as mixed areas of sclerosis and lucencies or homogeneously sclerotic areas. On magnetic resonance imaging (MRI) varies depending on the degree of lucencies versus sclerosis. Although FD is a benign disease, there may be malignant transformation in 0.5% of monostotic FDs and in 4% of polyostotic FDs.<sup>1,5</sup>

Fluid-fluid level containing lesions have some differential diagnosis, that must be shortened by considering age, location, benign x malignant features and radiographic appearance, as well as concomitant lesions like in our case of ABC secondary to FD. In ABC these levels they are mostly due to blood content of variable age.<sup>1</sup>

The differential diagnosis of an osteolytic skull lesion comprises hemangioma, epidermoid cyst, and eosinophilic granuloma. Skull

hemangioma is generally a solitary lytic diploic lesion. Hemorrhagic transformation may be a complication (seen on MRI), which would make differentiation with aneurysmal bone cyst more difficult. Epidermoid cyst involves both the inner and outer tables of the calvarial bone and is well-defined lesions that lack central trabeculae and have a sclerotic rim. On the other hand, eosinophilic granulomas are round or oval nonsclerotic skull lesion with sharply defined margins.<sup>1,5</sup>

## Acknowledgments

None.

## Conflicts of interest

The author declares that there is no conflicts of interest.

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