Aneurysmal bone cyst concomitant with fibrous dysplasia in the frontal bone

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ABSTRACT

يعد كيس أم الدم العظمي الثانوي الذي يظهر في خلل التنسج الليفي العظمي مرضاً نادراً جداً وخصوصاً عندما يوجد في عظام الجمجمة وبالتحديد عظمة الجبين. نقدم في هذا التقرير الحالة الثالثة في العالم والأولى في الشرق الأوسط من هذا النوع لمريض ذكر عمره 15 عاماً. لقد كان هذا الشاب يشكو من صداع وانتشاء بالإضافة إلى بعض الصور التخيلية الغير محددة. وقد تم علاجها عن طريق الاستئصال الجراحي الكامل لهذا الكيس.

Secondary aneurysmal bone cyst in fibrous dysplasia is exceedingly rare, especially in the skull and particularly in the frontal bone. We present a case of aneurysmal bone cyst concomitant with fibrous dysplasia in the frontal bone in a 15-year-old male patient presenting with headache and euphoria with an uncharacteristic imaging appearance and treated successfully by total resection.

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Aneurysmal bone cysts (ABCs) are uncommon lesions of the bone, most commonly found in the second decade with a female to male ratio of 2:1. These lesions are benign and non-neoplastic in nature and consist of cystic cavities filled with blood. The ABCs can be primary or secondary associated with a preexisting abnormality that can be confused with malignant degeneration. Increasingly more musculoskeletal pathologists regard the ABC as a secondary vascular phenomenon superimposed on a preexisting lesion.[1] It is suggested that an ABC signifies pathophysiological change, rather than a true neoplasm.[1] Only 2-6% of lesions develop in the cranial bone,[2] while secondary ABCs of the skull bone are extremely rare. We report a rare and interesting case of secondary ABC arising from fibrous dysplasia (FD) in the frontal bone and presenting clinically with euphoria, with imaging appearances misleading for malignant degeneration in FD.

Case Report. A 15-year-old male patient was medically free until 2 years ago when he started to complain of recurrent attacks of headache for which he was treated as a case of sinusitis at the family medicine clinics. One month ago, the attacks progressively increased in severity and frequency. He describes the headache of tension like, early morning, and relieved by analgesia. The family also reported behavioral changes as their son became euphoric. He was referred to a public hospital where brain CT scan (**Figure 1a**) without intravenous contrast (IV) was carried out and showed an expansile bony lesion in the diploic space of the right frontal bone with its extension mainly intracranially causing compression of the right frontal lobe. This lesion also showed a hypodense central region with a peripheral rim of soft tissue density. According to the CT scan findings, he was referred to the University of Jordan Hospital as a case of abscess. He was admitted via the Emergency Room to the Neurosurgery Department and on systemic review, his symptoms were not associated with nausea or vomiting, seizures or history of head trauma, but with a possible history of fever. His general physical and neurological examinations were normal with no head swelling, and laboratory results were all within normal limits. On MRI, the lesion of the right frontal bone showed on T1W images (**Figure 1b**) a hypointense central portion with a hyperintense peripheral thin rim and thick outer portion. On T1W images after IV contrast (Gadolinium) injection (**Figure 1c**), the peripheral thin rim showed ring enhancement, while neither the outer soft tissue component nor the central fluid signal part showed any enhancement. On T2W images (**Figure 1d**) the central region appeared hyperintense with septations and heterogeneous peripheral rim, but fluid-fluid appearance was not seen. He was referred for

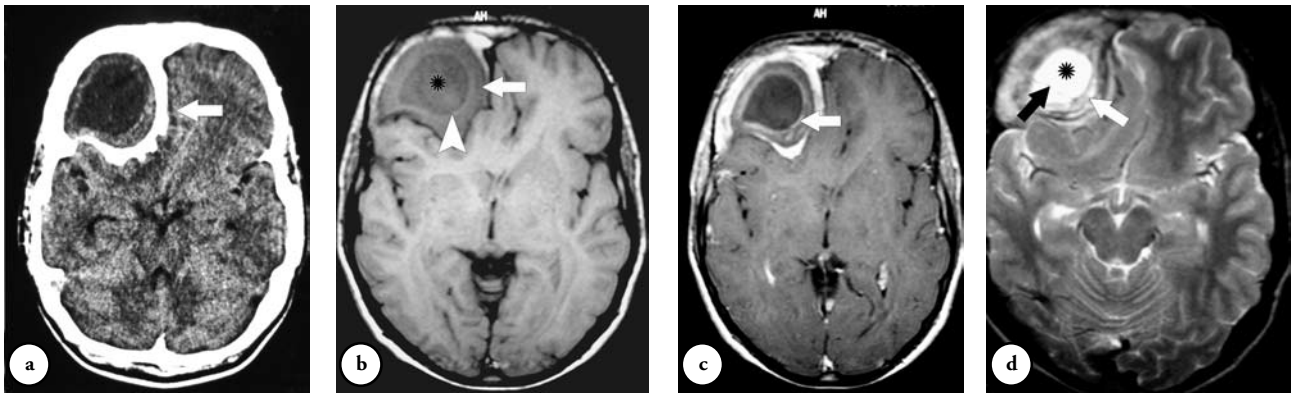

Figure 1 - Patient images showing **a)** axial CT scan without intravenous contrast shows a diploic expansile hypodense lesion in the right frontal bone. **b)** Axial T1W MRI shows the right frontal bony lesion with hypointense center (star) with hyperintense rim (arrow head) and the thick outer portion (arrow). **c)** Axial T1W MRI with intravenous contrast showing the enhancement of the rim (arrow). **d)** Axial T2W MRI showing central hyperintensity (star) with septation (black arrow) and heterogenous outer part (white arrow).

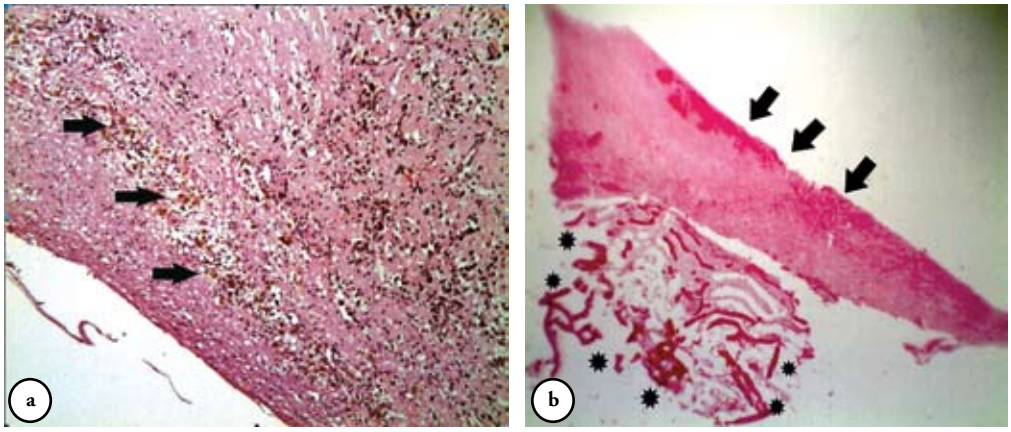

Figure 2 - Histopathological examination showing **a)** The cyst wall is densely collagenized, highly vascularized, and lacking cellular lining. There is infiltration by lymphocytes and plasma cells with foci of hemosiderin-laden macrophages (arrows). Some septae protrude into the lumen composed of fibrin and fibrous tissue. (40X). **b)** The area is indirect contact with the cyst wall (arrows), contains irregular curvilinear trabeculae of woven bone containing fibrous tissue in between (stars) (whole mount view).

neurosurgery as a case of ABC, and consequently he was treated by right frontal craniectomy, and total resection of the lesion was achieved with cranioplasty.

Histopathological examination of the specimen revealed a cyst wall with features consistent with ABC (**Figure 2a**). The area beneath the cyst wall shows typical morphology of FD (**Figure 2b**). External to this was normal lamellar bone trabeculae with normal intervening bone marrow. The appearances are of a composite bone lesion of both ABC and FD. He was symptom free in the postoperative period after 6 months of surgery and kept under close follow up.

Discussion. Fibrous dysplasia is an idiopathic condition in which normal bone is altered by abnormal fibro-osseous tissue causing distortion and overgrowth of the affected bone.[3] The clinical course of FD is unpredictable; although usually lesions tend to stabilize with the onset of puberty, others rarely (<1%) may undergo malignant transformation into osteosarcoma, fibrosarcoma, and chondrosarcoma.[1] The FD affects craniofacial structures in 25-30% of cases with a monostotic pattern, and 50% with polyostotic variety. The radiographic characteristics of FD, as described by Fries in 1957,[4] are pagetoid (56%), a mixture of dense and radiolucent areas of fibrosis; sclerotic (23%), massive homogenously dense; cystic (21%), and a spherical or ovoid lucency surrounded by a dense boundary. The radiological differential diagnosis of ABC includes, giant reparative granuloma, which has definite previous history of trauma, giant cell tumors, which are usually seen in the >30 years age group, and less commonly include a hemorrhagic cyst, and FD.[5] Jaffe in 1950 and again in 1962[6] referred to the possibility that an ABC might sometimes represent a secondary "blowout" in some preexisting bone lesion, like giant cell tumor

Table 1 - Reported cases of secondary aneurysmal bone cyst in fibrous dysplasia (FD) of the frontal bone.

Author/year	Age/ gender	Location/ size	Symptoms	CT findings	MRI findings	Treatment	Follow up	Remarks	FD since, monostotic/ polyostotic	Provisional diagnosis
Wojno and McCarthy, 1994[10]	F, 14	Left frontal, 5 cm	Expanding mass	Non homogenous cystic mass	Two components: solid and cystic	Surgical resection	Not available	Mass appeared after head trauma	Unknown	Not reported
Lin et al 2004[1]	M, 18	Left frontal, 7.3 cm	Left frontal bone mass with severe headache	*Initial CT:* an expansile sclerotic bone lesion, with a homogeneous ground-glass appearance at left frontal bone. *Follow up CT after one month:* several expansile cystic spaces, which bulged out, with cortical destruction beyond the frontal bone. Fluid-fluid levels inside the cysts	Cystic lesion with fluid-fluid level. Heterogenous SI on T1W and T2W images.	Surgical resection	Uneventful	Sudden rapid progressive enlargement of a left frontal bone mass with severe headache over 2 weeks	FD since 10 years, unknown	Malignant degeneration of FD
Present case	M, 15	Right frontal, 4.5 cm	Severe headache and euphoria	Expansile bony lesion, central hypodense central region with peripheral soft tissue density	Heterogeneous SI on T1W and T2W images, minimally enhanced after IV gadolinium	Surgical resection	After 6 months disease free	Recurrent attacks of headache over 2 years, treated as sinusitis	Unknown, monostotic	Abscess

(14.3%), chondroblastoma (14.6%), chondromyxoid fibroma (6.6%), non ossifying fibroma (3.3%), osteoblastoma (6.5%), fibrosarcoma (2%), malignant fibrous histiocytoma (6%), FD (2.4%),[7] and was recently reported with acute lymphoblastic leukemia.[8] The occurrence of FD and ABC is exceedingly rare, in a report of 57 ABCs associated with other osseous lesions; not a single case of FD was found.[9] In a review of 42 cases of FD, Martinez et al,[7] found only one case of secondary ABC with FD in a rib. While reviewing the English literature for secondary ABC and FD in the skull, Hadad et al[3] found only 6 cases and only one case was in the frontal bone,[10] while our publication survey revealed another similar case (**Table 1**).

An ABC in the frontal bone is seen in 15% of the ABCs of the cranium in children,[5] while FD involvement of the frontal bone is seen in 33%.[3,5] An ABC arising from FD is very rare especially within the skull.[1] Most present as painful swelling over a short period simulating malignant transformation, which occurs in 0.5% of the monostotic type, and 4% of patients with polyostotic FD.[1] Rapid enlargement of previously dormant lesions should not be misdiagnosed as malignant degeneration of FD, and the thought of secondary ABC with possible spontaneous hemorrhage should be kept in mind and dealt with accordingly.[3] In our patient, the FD lesion was of the monostotic variety and its growth was only intracranially and discovered only on histopathology.

Secondary ABC in FD should be taken into consideration when the classical fluid-fluid appearance is described in CT or MRI, optimally seen in sagittal and axial T2W sequences, appearances which were not seen in our patient. A CT is the gold standard technique for diagnosis and follow-up of FD because of its superior bony detail and accurate assessment of the extent of the lesion.[5] For ABC, MRI is the best with sensitivity of 77.8% and specificity of 66.7%, especially when characteristic features are present.[11]

In reviewing the literature, the treatment suggested for ABC with FD varied from close follow-up to total resection of lesions according to their size and location. Radiotherapy is contraindicated in the treatment of secondary ABC associated with FD due to an increased chance of malignant transformation.[12] Fortunately the lesion in our case was amenable for total resection.

In conclusion, the incidence of secondary ABC in FD in the frontal bone is exceptionally rare. We report a young adult who presented with euphoria and headache and was found histopathologically to have secondary ABC concomitant with FD, although CT and MRI findings were not classical for ABC or FD. The patient was successfully managed by total surgical resection of the lesion, which is the preferred method of treatment.

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