Aneurysmal bone cyst concomitant with fibrous dysplasia in the frontal bone

Azmy M. Hadidy, MD, FRCR, Ahmad F. Tamimi, MD, PhD, Luma M. Fayad, MD, FRCPath, Huda A. Al-Jadiry, MD, HSCM.

ABSTRACT

Aneurysmal bone cyst concomitant with fibrous dysplasia in the frontal bone 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 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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From the Departments of Radiology (Hadidy, Al-Jadiry), Neurosurgery (Tamimi), and Pathology (Fayad), The University of Jordan Hospital, Amman, Jordan.

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Address correspondence and reprint request to: Dr. Azmy M. Hadidy, Radiology Department, The University of Jordan Hospital, Queen Rania Street, PO Box 13046, Amman 11942, Jordan. Tel. +962 788707000. Fax: +962 (6) 5353388. E-mail: amshadidy@yahoo.com

Neurysmal 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
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. 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. 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, 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. Jaffe in 1950 and again in 1962 referred to the possibility that an ABC might sometimes represent a secondary “blowout” in some preexisting bone lesion, like giant cell tumor.
(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%), and was recently reported with acute lymphoblastic leukemia. 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. In a review of 42 cases of FD, Martinez et al. 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. found only 6 cases and only one case was in the frontal bone, 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, while FD involvement of the frontal bone is seen in 33%. An ABC arising from FD is very rare especially within the skull. 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. Rapid enlargement of previously dormant lesions should not be misdiagnosed as malignant degeneration of FD, and the thought of secondary ABC 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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References


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

<table>
<thead>
<tr>
<th>Author/year</th>
<th>Age/ gender</th>
<th>Location/ size</th>
<th>Symptoms</th>
<th>CT findings</th>
<th>MRI findings</th>
<th>Treatment</th>
<th>Follow up</th>
<th>Remarks</th>
<th>Provisional diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wujia and McCarthy, 1994</td>
<td>E, 14</td>
<td>Left frontal, 5 cm</td>
<td>Expanding mass</td>
<td>Non homogenous cystic mass</td>
<td>Two component solid and cystic</td>
<td>Surgical resection</td>
<td>Not available</td>
<td>Mass appeared after head trauma</td>
<td>Unknown</td>
</tr>
<tr>
<td>Lin et al 2004</td>
<td>M, 14</td>
<td>Left frontal, 7.5 cm</td>
<td>Left frontal bone mass with severe headache</td>
<td>Initial CT: an expansile sclerotic bone lesion, with a homogenous 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</td>
<td>Cystic lesion with fluid-fluid level. Heterogenous SI on T1W and T2W images.</td>
<td>Surgical resection</td>
<td>Uneventful</td>
<td>Sudden rapid progressive enlargement of a left frontal bone mass with severe headache over 2 weeks</td>
<td>FD since 10 years, unknown</td>
</tr>
<tr>
<td>Present case</td>
<td>M, 15</td>
<td>Right frontal, 4.5 cm</td>
<td>Severe headache and euphoria</td>
<td>Expandable bony lesion, central hypodense central region with peripheral soft tissue density</td>
<td>Heterogenous SI on T1W and T2W images, minimally enhanced after IV gadolinium</td>
<td>Surgical resection</td>
<td>After 6 months disease free</td>
<td>Recurrent attacks of headache over 2 years, treated as sinusitis</td>
<td>Unknown, monostotic</td>
</tr>
</tbody>
</table>

For ABC, MRI is the best with sensitivity of 77.8% and specificity of 66.7%, especially when characteristic features are present.

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. 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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References

Aneurysmal bone cyst in fibrous dysplasia ... Hadidy et al


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