Aneurysmal bone cyst with fibrous dysplasia of the parietal bone

Report of two cases

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The diagnostic and surgical aspects of aneurysmal bone cysts contiguous to fibrous dysplasia of the parietal bone are reported in two adolescents. While aneurysmal bone cyst and fibrous dysplasia are known to be associated, the association is rare and has not been reported previously in the calvaria.

KEY WORDS  •  aneurysmal bone cyst  •  fibrous dysplasia  •  skull tumor

Fibrous dysplasia of bone is a benign mesenchymal maldevelopment first described by Lichtenstein in 1938.19 It usually involves a single bone but multiple bone involvement has been reported. Fibrous dysplasia plus hyperpigmentation of the skin, endocrinopathy, and, in particular, premature sexual development in a female patient constitute the “McCune-Albright syndrome.”10 Aneurysmal bone cyst is also a non-neoplastic lesion of bone, but it can be differentiated from fibrous dysplasia of the bone by its venous pools, a trabecular bone matrix, and reparative bone changes at its border. Although numerous reports associate aneurysmal bone cyst with neoplastic and non-neoplastic lesions in other bones, this report is the first documentation of aneurysmal bone cysts contiguous to fibrous dysplasia in the calvaria.

Case Reports

Case 1

This 9-year-old girl, seemingly in good health, was referred by her pediatrician for assessment of skull lesions. Three years before this admission she had developed left frontal swelling after a fall. The swelling was assumed at first to be a hematoma, but it did not resolve over the next 2 years, at the end of which time a second skull lesion had appeared in the left parietal region. Skull roentgenograms showed changes characteristic of fibrous dysplasia of the frontal and parietal bones (Fig. 1); a skeletal survey excluded other lesions. Visual field testing was normal, and the patient was followed at regular intervals. One month before this admission, the left parietal lesion became tender and began to enlarge.

Examination. Physical examination showed a well-appearing 9-year-old girl with a tender 4 × 5-cm cystic lesion in the left parietal area of the skull (Fig. 2) and a firm nontender protuberance of the left frontotemporal skull; she had no stigmata of the McCune-Albright syndrome. Skull roentgenograms were interpreted as essentially unchanged from the previous ones, but in light of the clinical change in the lesion, further diagnostic procedures, including biopsy, were carried out. A radionuclide bone scan confirmed the presence of two skull lesions and the absence of other skeletal involvement. Cranial computerized tomography (CT) revealed a large area of bone lysis with erosion of the inner and outer tables and expansion of the diploic space in the left parietal area (Fig. 3), a soft-tissue mass with an abnormal rim of enhancement superficial to the skull lesion, and abnormality of the inferior frontal bone and the greater wing of the sphenoid on the same side. The destructive appearance of the left parietal lesion was interpreted as a possible sarcomatous degeneration of fibrous dysplasia.

Operation. A dark hemorrhagic 3 × 4-cm cystic lesion of the skull was removed at operation. It had
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FIG. 2. Case 1. Preoperative photograph with the scalp shaved illustrates the 4 × 5-cm cystic lesion protruding from the left parietal region of the skull.

FIG. 1. Case 1. Plain roentgenogram of the skull, anteroposterior view, showing increased opacity of the left frontal skull base and parietal bone (arrowheads), indicative of fibrous dysplasia.

completely eroded the inner and outer tables of bone, but there was no evidence of invasion of the dura. At the borders of the lesion, the skull was thickened, soft, and gritty in an area 1 to 2 cm around the cysts; this area was resected as much as possible. A methyl methacrylate plate was molded into the cranial defect. The postoperative course was complicated only by a persistent subgaleal fluid collection, which required percutaneous aspiration several days after the operation.

Pathological Examination. Microscopic studies of the resected tissue displayed regions of proliferating fibroblasts within a collagenous matrix and trabeculae of osteoid and bone consistent with fibrous dysplasia (Fig. 4 left). In addition, there were cavernous spaces lined by thin-walled septae often surrounded by multiple benign-appearing giant cells (Fig. 4 right). The diagnosis of fibrous dysplasia with aneurysmal bone cyst was confirmed by several consultants.

Case 2

This 19-year-old man was referred for evaluation of...
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Fig. 4. Case 1. Left: Photomicrograph of the skull lesion showing overgrowth of the benign fibrous tissue (F) comprising the fibrous dysplasia. \( \times 123 \). Right: Photomicrograph of the aneurysmal bone cyst contained in the same lesion showing two large endothelium-lined spaces (asterisks) containing red blood cells. The walls show osteoclastic giant cells (arrowheads). \( \times 164 \).

an expanding skull lesion. Fifteen years earlier he had developed a fullness in the right periorbital region; biopsy and roentgenographic study of the lesion were consistent with a diagnosis of fibrous dysplasia of the skull. Subsequent roentgenograms documented mild progression of the lesion throughout puberty, but there were no major cosmetic deformities or neurological sequelae requiring surgical decompression. Four weeks before this evaluation, the patient had noticed a nontender swelling over the right parietal area of his skull, which progressively increased in size.

Examination. There was mild but easily detectable facial asymmetry in the form of a protuberant right orbit. A nontender 6 \( \times \) 8-cm cystic lesion was palpable in the right parietal region, extending above the skull by 6 cm. Skull roentgenograms and cranial CT scans showed a cystic expansion of the skull consisting of fibrous dysplasia, and a soft-tissue mass with an abnormal rim of contrast enhancement superficial to the skull defect (Fig. 5).

Operation. A 6 \( \times \) 8-cm lesion filled with dark viscous fluid was removed. It had completely eroded the inner and outer tables of the skull. There was no dural invasion, and at the borders of the lesion the skull was

Fig. 5. Case 2. Left: Computerized tomography scan showing a cystic area with abnormal enhancement superficial to the lesion. Right: Plain roentgenogram of the skull, anteroposterior view, showing changes of fibrous dysplasia and cystic expansion of the right parietal bone. The appearance was strikingly similar to that in Case 1.
soft, thickened, and gritty. The cystic lesion was excised and the defect was filled with methyl methacylate.

Pathological Examination. Microscopic studies of the excised lesion showed regions characteristic of fibrous dysplasia adjacent to cavernous spaces lined by endothelium and surrounded by multiple benign appearing giant cells characteristic of aneurysmal bone cyst (Fig. 6).

Discussion

The association of an aneurysmal bone cyst with fibrous dysplasia is a rare but previously documented entity;\(^3,5,7,11,12,23\) to our knowledge it has not been reported before in the bones of the calvaria. The aneurysmal bone cyst is a distinct lesion described initially by Jaffe and Lichtenstein;\(^13,14,17\) it may occur in the vertebrae, the long bones, or the flat bones. Initial symptoms include pain, rapid enlargement, and possible skeletal or neurological dysfunction, depending on the lesion's location and the extent of bone destruction. Aneurysmal bone cysts were initially described as primary lesions arising from traumatic or anomalous venous disruption in the diploe with subsequent increase in venous pressure and development of a dilated and engorged vascular bed in the affected bone.\(^17\) More than 300 cases of primary aneurysmal bone cysts have been reported,\(^3,4,9,21-23\) and the primary form of this lesion appears to be the predominant form.

Subsequent reports have confirmed the association of aneurysmal bone cysts with other lesions in bone, including solitary bone cysts, osteoclastoma, osteosarcoma, nonosteogenic fibroma, osteoblastoma, hemagioendothelioma, hemangiofibroma, and fibrous dysplasia.\(^3,5,11,12,16\) Cacdac, et al.,\(^6\) reported the association of an aneurysmal bone cyst with a hemangiofibroma of the parietal bone. These reports support the view that a secondary form of aneurysmal bone cyst may arise from a disruption in the osseous circulation caused by a primary lesion. In our patients no history of head trauma, mild or otherwise, could be elicited.

The largest aneurysmal bone cyst reported was occipital and measured 32 \(\times\) 21 \(\times\) 8 cm.\(^{21}\) Bonakdarpour, et al.,\(^4\) reviewing the radiographic characteristics of primary and secondary aneurysmal bone cysts, reported that when an aneurysmal bone cyst was associated with another primary lesion the latter was diagnosed correctly in the majority of cases. If a malignant lesion was associated, it was always diagnosed correctly despite its association with a cyst. Thus, a primary aneurysmal bone cyst appears to have a characteristic radiographic appearance, while the associated lesion generally has the radiographic appearance of its precursor.

A primary aneurysmal bone cyst appears on plain x-ray films as a round or ovoid lesion with varying degrees of expansion or cortical thinning and septation.\(^5,17\) Computerized tomography confirms that it is an expansile diploic lesion.\(^25\) The microscopic appearance is of a septated vascular lesion with scattered endothelium and multinucleated giant cells lining the sinusoids. Around it is less-cellular fibroblastic tissue, with fibrous hyaline strands and irregular osteoid trabeculae surrounded by osteoblasts and osteoclasts.\(^17,18,26\) When isolated from the cystic structure, these areas may resemble other benign or malignant lesions such as osteoblastoma, osteosarcoma,\(^28\) giant-cell granuloma, or even fibrous dysplasia.\(^26\) The entire lesion must be examined in order to assure the correct diagnosis.

Fibrous dysplasia is a separate entity with a distinctive radiographic and pathological appearance.\(^9,19\) Recent reviews\(^10,20\) confirm that it is generally a disease of adolescents; in the monostotic form, the craniofacial bones are involved in about 10% of cases, while in the polyostotic form, 50% to 100% involvement has been reported. Neurological symptoms of skull fibrous dysplasia are generally secondary to compression of optic or other cranial nerves. Radiographically, the skull lesions are sclerotic, pagetoid, or lytic (cystic). The disease is generally self-limiting in the second and third decades of life, but the potential for malignant transformation apparently is present for as long as 30 years after onset. For that reason, new symptoms in an old focus of fibrous dysplasia must be investigated. Malignant transformation is increased with radiation therapy, which is now contraindicated.

It is well recognized that fibrous dysplasia affects the basal skull and calvaria. That aneurysmal bone cysts occur in the calvarial bones is also documented.\(^1,15\) Keuskamp, et al.,\(^15\) reviewing 43 cases of aneurysmal bone cysts of the skull reported in the world literature since 1942, found that the lesions were most often periorbital, but that they were found also (in decreasing order of frequency) in the occipital, temporal, basal, parietal, and frontal bones. In none of these cases or those added by Ameli, et al.,\(^1\) was there radiographic or histological evidence of associated fibrous dysplasia. In our patients, fibrous dysplasia was noted radiograph-
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ically in the skull months or years before there was evidence of an associated aneurysmal bone cyst. Fibrous dysplasia in the bone surrounding the aneurysmal bone cyst was diagnosed histologically by multiple independent consultants. No sarcomatous changes were found.

The potential for malignant degeneration in fibrous dysplasia is less than 1% in nonirradiated patients.19 Surgical excision is usually required only for neural decompensation or cosmetic reconstruction.

On the other hand, complete excision followed by bone or methyl methacrylate reconstruction is the treatment of choice for an aneurysmal bone cyst of the skull. The potential for recurrence is low, and may be related to the age of the patient, the size of the lesion, the presence of mitoses, or the incompleteness of the resection.12,23,26 The recurrence rate does not appear to be altered when a second primary lesion is present.1,12,16 In our patients, the aneurysmal bone cyst was completely excised but residual fibrous dysplasia remained in the surrounding bone. A methyl methacrylate plate was inserted in both patients in anticipation of the eventual remission of the fibrous dysplasia. Cryosurgery or radiation therapy are alternative options for the aneurysmal bone cyst in which the location precludes complete removal and where recurrence is potentially devastating. Irradiation in doses ranging from 600 to 3000 rads has been suggested.3,12 However, radiation therapy would seem to be contraindicated in the treatment of aneurysmal bone cyst contiguous with associated fibrous dysplasia.

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References


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