Case Report

Polyostotic fibrous dysplasia with secondary aneurysmal bone cyst in tibia

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ABSTRACT

Fibrous dysplasia is a benign tumor-like lesion of bone believed to be developmental in origin. Polyostotic fibrous dysplasia (FD) is a rare condition. Our case was further complicated by the presence of secondary aneurysmal bone cyst (ABC). This is the second reported case of polyostotic FD with secondary ABC.

Keywords: *Aneurysmal bone cyst, polyostotic fibrous dysplasia, fibrous dysplasia*

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Introduction

FD is a benign tumor-like lesion of bone presenting in children and young adults. FD constitutes 2.5-10% of the total bone tumors. Polyostotic FD forms 30% of total FD. FD is characterized by the replacement of medullary bone by fibro-osseous tissue. ABC is a benign condition characterized by cavernous vascular spaces lined by fibrous tissue, osteoclastic giant cells, and woven bone. In our case, ABC developed secondary to FD.

Case Report

A 22-year-old female came to the Orthopaedic Outpatient Department with complaint of pain in left lower limb and swelling just below the left knee since 1 year. The pain was a deep dull ache that responded to painkillers initially, but since the last month, it had become more severe and had restricted her movements. There was no history of trauma. There was no history of similar pain in any other part of body. On examination, the patient had normal temperature, pulse, and blood pressure. She walked with a limp. Both lower limbs were of equal length and girth. Movements of the left hip and ankle joint were normal. Knee movements were painful therefore restricted. A bony swelling measuring 6 cm × 8 cm was palpable in the left upper leg on the anterior aspect, which was tender. Her neurological examination was within normal limits. She was sent for plain X-ray examination of the left lower limb [Figure 1]. The left tibia showed an expansile lesion in proximal one third of diaphysis, partly extending in metaphysis without cortical reaction. There was thinning and break of cortex in the anteromedial aspect. Subsequent computerized tomography (CT) showed expansile lesion with ground glass appearance and internal septae [Figure 2a]. Magnetic resonance imaging (MRI) images in T1 coronal, T2 sagittal, and T1 contrast planes showed well demarcated, mildly expansile solid cystic lesion in proximal one-third of the left tibial diaphysis, partly extending to the metaphysis on the anteromedial aspect [Figure 2b and c]. Thinning of adjoining anterior, medial, and lateral cortex was noted, with cortical breaks at multiple places with minimal adjacent anterior extraosseous soft tissue extension. Posterior cortex was intact. The solid component showed T1 hypointense and T2 mildly hyperintense signal intensity with moderate post-contrast enhancement. The hypointense septae also showed post-contrast enhancement with no enhancement of cystic spaces. No periosteal reaction was noted. No adjoining marrow infiltration or edema was observed. A three-phase radionucleid bone scan [Figure 3] was suggestive of mixed osteoblastic and osteolytic lesion in the upper end of left tibia, with focal osteoblastic lesions in the skull and pelvis. Based on these findings, a diagnosis of polyostotic fibrous dysplasia (FD) was made. The routine hematological and biochemical investigations including serum calcium,

phosphorus, and alkaline phosphatase yielded normal results. Café au lait spots were searched and were found to be absent. There were no clinical signs of endocrinopathies. She attained puberty at 13 years and had a regular, normal menstrual cycle. Patient was taken for open biopsy. Tissue received in surgical pathology department consisted of several small bits of bony, gritty tissue, totally measuring 2 cc. Histopathologically, the lesion was made up of fibrous tissue consisting of spindle-shaped fibroblasts and irregularly shaped bony trabeculae interspersed in between [Figure 4a]. These bony trabeculae showed osteoblastic rimming in few places. In addition, vascular spaces lined by osteoclastic giant cells and fibrous tissue were noted [Figure 4b]. A diagnosis of FD with secondary aneurysmal bone cyst (ABC) was made. The patient underwent curetting of the lesion with bone grafting. Postoperative recovery was uneventful. She was administered bisphosphonates and advised regular follow up.

Discussion

FD is a benign tumor-like disorder of unknown etiology, in which fibro-osseus tissue replaces the medullary bone. It constitutes 2.5-10% of bone tumors. It is common in children and young adults and is found equally in both sexes. It can present as monostotic type, polyostotic type, or as part of the McCune–Albright syndrome. The polyostotic type is less common (30%) and the McCune–Albright syndrome is very rare.[1] FD most often presents in long bones of upper and lower extremity, ribs, pelvis, and craniofacial bones. In polyostotic FD, bones such as facial

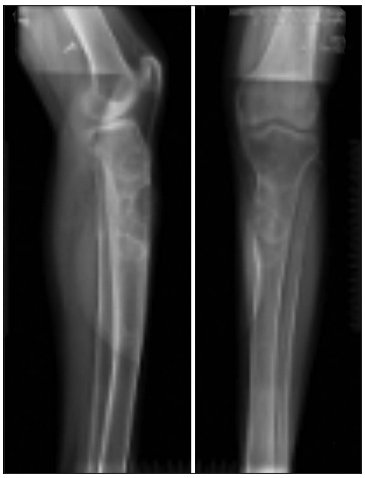

Figure 1: X-ray of left leg: AP and lateral view showing an expansile, lytic lesion

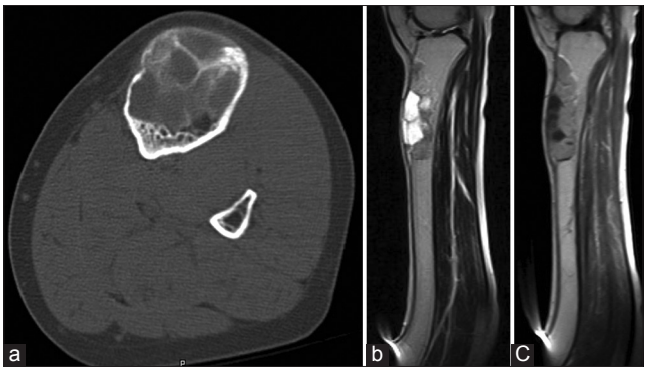

Figure 2: (a) CT axial left tibia showing ground glass appearance of the expansile lesion. (b) MRI Image in T2 sagittal plane, and (c) MRI image in T1 sagittal plane with contrast showed well-demarcated mildly expansile solid cystic lesion in proximal one-third of left tibial diaphysis

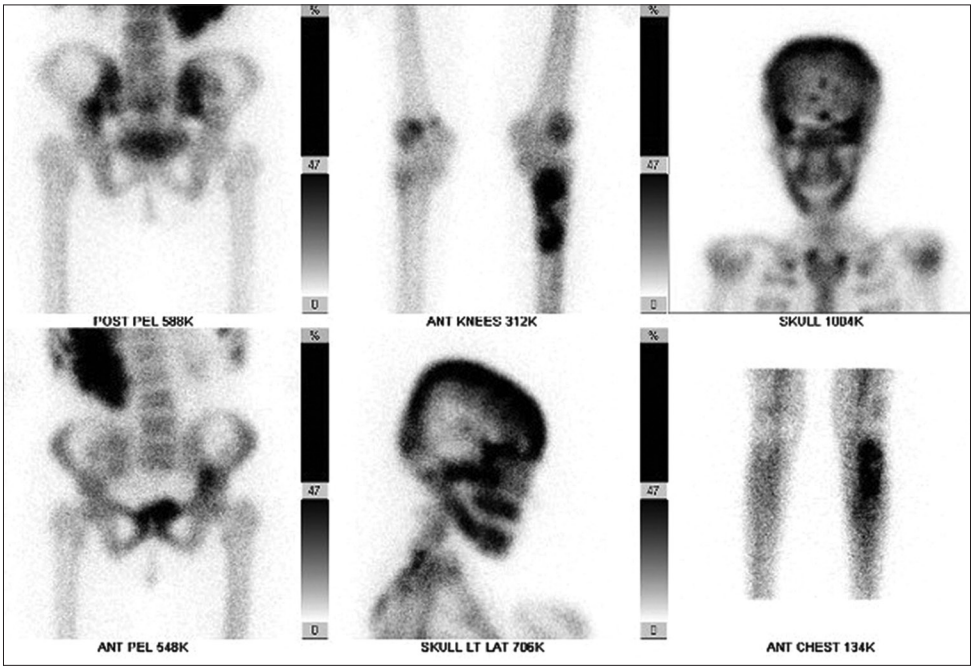

Figure 3: A three-phase radionucleid bone scan showing mixed osteoblastic and osteolytic lesion in upper end of left tibia with focal osteoblastic lesions in skull and pelvis

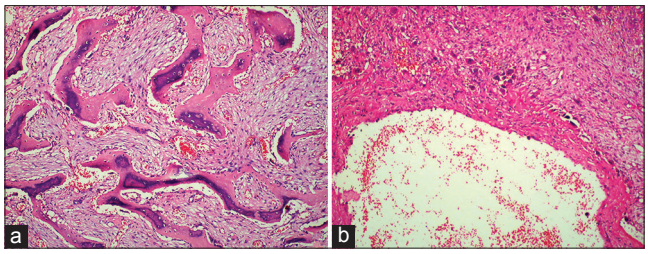

Figure 4: (a) Fibrous tissue and irregularly shaped bony trabeculae interspersed in between, (b) vascular spaces lined by osteoclastic giant cells and fibrous tissue

bones, skull base, long bones, and occasionally ribs are typically involved. Recent research showed an association between FD and mutation in the Gs gene, which is situated on chromosome 20q13.2-13.3.[2] Radiologically, on plain X-ray and CT, an expansile metaphyseal lesion showing ground glass appearance is typical of FD. MRI of FD reveals typically low signal intensity on T1- and T2-weighted images with contrast enhancement.[3]

An ABC is a benign cystic lesion of bone. It may occur *de novo* or secondary to other lesions of bone (20-30%). It is composed of blood-filled cystic spaces lined by fibroblasts, osteoclastic multinucleate giant cells, and reactive woven bone. The primary diseases known to be associated with ABC are osteoclastoma, osteosarcoma, osteoblastoma, and hemangioma.[4] On plain X-ray and CT, an expansile bony cyst is seen with varying degree of cortical thinning. MRI shows high-signal intensity on T1-weighted images.[3] In some cases, a fluid-fluid level is clearly visible. The exact pathogenesis of secondary ABC is not known. It is believed to be a secondary reactive lesion due to the primary tumor-induced anomalous vascular process.[5]

FD with secondary ABC is rare. It has been reported in facial bones, skull base, and calvarium.[1,6] It has been reported in the spine.[7] Montalti *et al.* reported a case involving the proximal femur.[8] Choi *et al.* reported a case affecting the proximal tibia in a toddler.[9] Thus, our case is the second case affecting the proximal tibia, but our patient is a young adult. Most of the reported cases have occurred with monostotic FD. We found one case where ABC occurred with plyostotic FD.[10] Thus, our case is the second case reported in a polyostoic FD. The presentation of both FD and ABC depends on the site of involvement, rate of growth, and proximity to important structures such as brain, eye, and nerves. Symptoms may include a painless or painful mass, as in our case. Pressure symptoms may also be a presenting feature. Pathological fracture may occur. The diagnostic modalities include imaging studies, especially CT and histopathological examination. Treatment of FD includes bisphosphonates with regular follow up, and surgery in some symptomatic cases. ABC on its own may be treated by selective arterial embolization or by surgery. Treatment of secondary ABC is that which is appropriate for the underlying lesion.

Conclusion

Our case highlights the importance of histopathology in diagnosis of FD with ABC. Without this investigation, the diagnosis would not be possible.

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