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Case Report

A large mixed pattern aneurysmal bone cysts: An extremely rare case with unusual radiological findings and management sequela

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ABSTRACT

A mixed pattern of aneurysmal bone cysts (ABCs) is an extremely rare anatomic subtype. Typical ABCs usually arise in the metaphysis of long bones, predominantly in childhood-age patients. Here, we report a case of a 37-year-old female presenting with the left leg pain and mass for 2 years. Conventional radiographs and computed tomography scans revealed a well-demarcated expansile lytic lesion at the diaphysis of the left tibia. Magnetic resonance imaging showed fluid-fluid levels with associated bone edema. These findings were suggestive of ABC. An open incisional biopsy was done and was confirmatory. After 8 months, the patient showed radiographic complete ossification of the lesion with persistent symptoms. Excision and intralesional curettage of the lesion was performed and supported with a tibia locking plate. On 18 months of follow-up, the patient was asymptomatic, with no recurrence observed on conventional radiographs.

Keywords: Aneurysmal bone cyst, Bone marrow edema, Magnetic resonance, Mixed pattern, Tibia

INTRODUCTION

Aneurysmal bone cysts (ABCs) are rare, benign, yet locally destructive, expansile bone tumors with an incidence rate of 0.14–0.32 cases per 100,000 individuals.[1] They constitute around 1% of all primary bone tumors.[2] They usually occur in the metaphysis of long bones, especially around the knee, although they can be found almost anywhere in the appendicular or axial skeleton.[1,2] Almost 80% of these lesions occur in patients who are younger than 20 years old.[1,2] Clinically, ABCs commonly present with pain, swelling, or an expansile mass.[1] Capanna *et al.* classified ABCs into five morphologic subgroups based on radiographic appearances. Types I–III represent typical ABCs with a medullary origin, whereas types IV and V are both considered "surface" ABCs.[1-4] Surface ABCs are less common, originate on the surface of the bone, and can extend into the surrounding soft tissue.[1-3] Maiya *et al.* described a mixed variant of ABCs where precise origin could not easily be determined, as they may present with features suggestive of one pattern and then develop features of another pattern and tend to be more aggressive than surface ABCs with an incident rate of 2.7% of all ABCs.[3] Here, we report a case of a giant, mixed pattern ABCs in the diaphysis of a left tibia patient in her late fourth decade. Our case illustrates the

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importance of listing ABCs in the differential diagnoses of expansile mixed cystic diaphyseal lesions in adult-age patients. Furthermore, this case is important in sharing clinic-radiological features, treatment outcomes, and follow-up regarding this rare case.

CASE REPORT

A 37-year-old woman presented with gradually increasing pain during weight-bearing and swelling in her left leg over the past 2 years. She had no substantial personal or family medical history or any prior history of trauma. Locally, no skin changes were visible, but a palpably large mass was observed in the anteromedial portion of the patient's left leg. She had neither restriction in her range of motion nor neurovascular compromise. Her initial workup ruled out an infectious process and other lesions of her skeleton. A conventional radiograph and computed tomography (CT) scan of the left tibia revealed a well-demarcated expansile lytic diaphyseal mass [Figure 1]. Magnetic resonance imaging (MRI) was ordered and was suggestive of ABC [Figure 2]. A pre-operative core biopsy was done, which revealed histological findings suggestive of ABC, but this was not conclusive. The patient was booked for a surgical open biopsy. The open biopsy came back confirmatory for ABC. The patient was discharged with outpatient follow-up. The patient, however, did not show up at the clinic for her scheduled follow-up appointment.

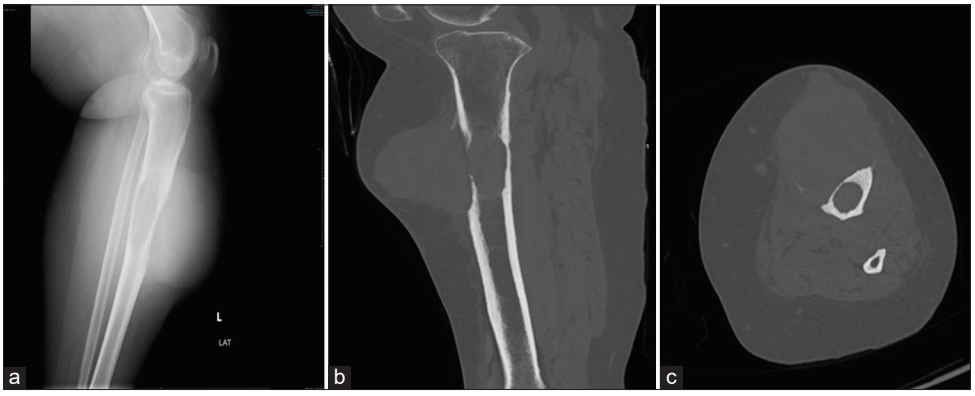

Figure 1: Pre-operative imagining studies of the left tibia. (a) Lateral radiograph shows a well-demarcated expansile eccentric lytic lesion ballooning appearance with cortical bone destruction. (b and c) Sagittal and axial computed tomography cuts demonstrate heterogeneous protruding soft-tissue mass with intramedullary extension marked intra- and extraosseous components measuring 7.5 × 6.8 × 10 cm in maximum axial and craniocaudal dimensions, respectively. Notably limited formation of the periosteal shell.

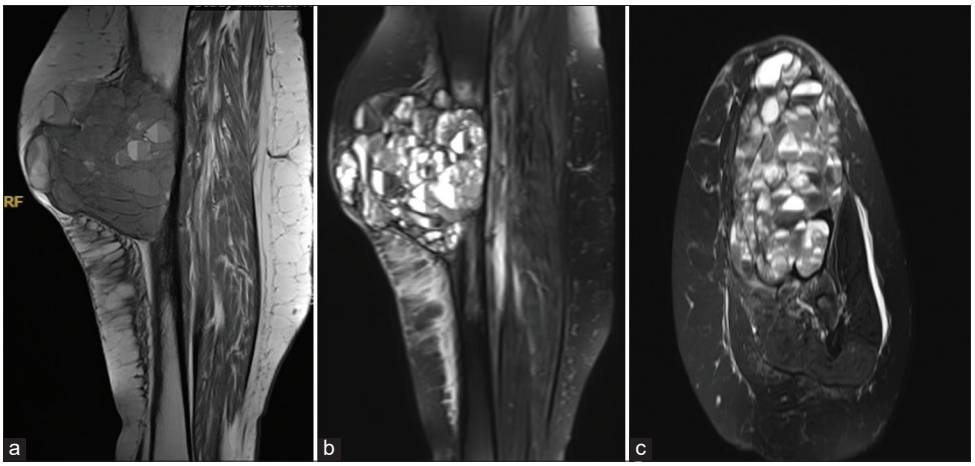

Figure 2: Pre-operative magnetic resonance imaging. (a) T1-weighted sagittal cut, (b) T2-weighted sagittal cut, and (c) T2-weighted axial cut show proximal-mid tibial diaphyseal osteolytic lesion. The lesion characteristically shows multiple cysts with fluid-fluid levels denoting different ages of blood products. Although significant intraosseous component intramedullary, the bulk of the lesion is subperiosteal. There is surrounding muscle, perilesional, and intramedullary edema.

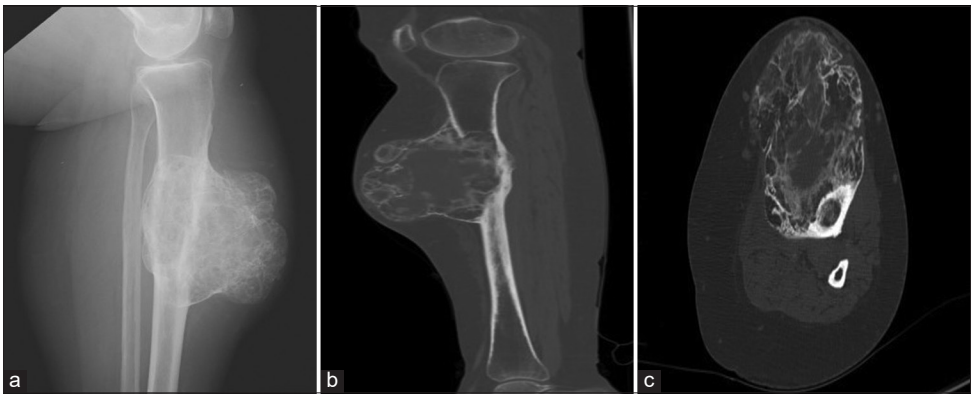

Figure 3: Post-incisional open biopsy images. (a) Lateral radiograph. (b and c) Sagittal and axial computed tomography scan cuts showing further interval mineralization and ossification of both extra and intraosseous components. It measures approximately 8.5 × 9.9 × 11.3 cm in maximum axial and craniocaudal dimensions, respectively.

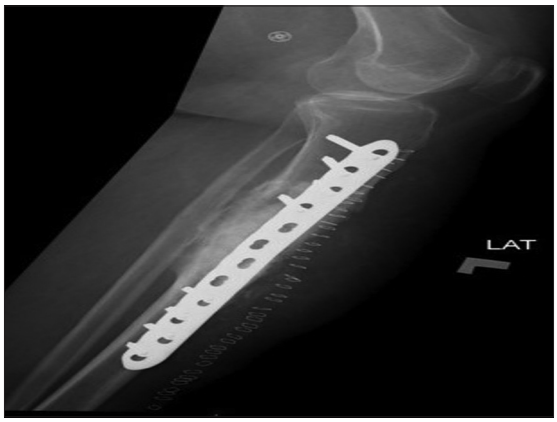

Figure 4: A 38-year-old female with aneurysmal bone cysts (ABCs) of the left tibia. Lateral view radiograph shows postexcision and intralesional curettage of the left tibia ABCs which are supported by tibia locking plate.

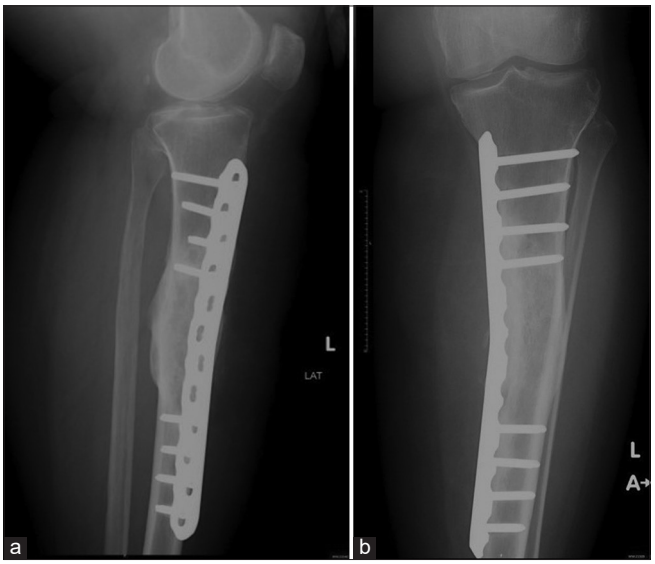

Figure 5: (a and b) 18 months follow-up conventional radiographs show bone remodeling changes.

Nevertheless, she returned 8 months after her left leg swelling and pain had worsened, and she could no longer bear weight. The radiograph of the left tibia revealed complete ossification of the lesion [Figure 3a]. Surgery was planned after the CT scan [Figure 3b and c] during the same week. The patient underwent an excision of the lesion and intralesional curettage, which was supported with a locking tibial plate osteosynthesis [Figure 4]. The histological findings of the excised mass revealed cystic lesions lined by fibrous walls with mild inflammation, amorphous contents, and scanty multinucleated giant cells. During the 18-month follow-up, the patient was asymptomatic, with no recurrence observed on conventional radiographs [Figure 5a and b].

DISCUSSION

ABCs were described as distinct entities by Jaffe and Liechtenstein for the 1st time in 1942. They are described as expansile lesions with cavities and thin blood-filled walls.[1,3] There were many theories regarding the etiology of primary ABC. These include traumatic, vascular, and, to a lesser extent, genetic pathophysiology. They are primary benign bone neoplasms that were once thought to be reactive but are now known to be neoplastic based on variations in the USP6-gene.[1,2] Surface ABCs are a rare ABC type extending beyond the bone outline. They usually occur within the cortex or, conversely, below the periosteum, limited by the periosteum externally and inwardly constrained by the endosteum.[3,4] Mixed pattern ABCs, on the other hand, are rare and have features of both medullary and surface ABCs. They can be challenging to diagnose, as they can mimic other bone tumors, such as osteosarcoma, especially the telangiectatic type.[1,3,4] ABCs can cause diagnostic uncertainty because

they can exhibit a destructive radiographic appearance, usually observed as lytic, expansive, and multi-loculated lesions with disturbed cortices.[2] Radiographically, periosteal reactive formation, also known as the periosteal shell, in ABCs can be complete, partial, or absent with respect to the activity level of the lesion. Aggressive ABCs in which the periosteal shell is absent may mimic malignant bone tumors radiographically.[5] Infrequently, CT might show a finding of fluid-fluid levels sign but is usually used preoperatively to better outline the bony boundaries of the lesion.[1,5] Although the MRI considered the modality of choice for detecting fluid-fluid levels, this finding is also a radiographic feature of the telangiectatic osteosarcoma (2nd most common), giant cell tumor, chondroblastoma, osteoblastoma, and fibrous dysplasia.[3,5,6] Thus, at the moment, a histological evaluation of incisional biopsies is the benchmark for diagnosing ABCs.[1-4] Interestingly, the MRI finding of bone marrow edema along the diaphyseal shaft in our current case is most unusual and rarely reported without a pathological fracture.[6]

The reported surface/mixed variant ABCs treatment is curettage, excision, or resection. The prognosis is generally good, with no evidence of recurrence.[3-5]

There is no uniform consensus on conservative treatment for these lesions.[7] It has been reported that ABCs occasionally heal spontaneously after a pathological fracture or biopsy alone. Intrinsic healing potential after minimally invasive curettage with the healing of 81% of the lesions was also reported.[8] Our case, on the other hand, showed a large diameter and aggressive lesion went to complete ossification after open biopsy but did not relieve patient symptoms. Due to the destructive nature of ABCs (i.e., cortical destruction) and especially when it is located in the diaphyseal area (long lever arm at the site of cortical destruction), causing stress riser necessitating an adequate internal fixation using intramedullary nail or plate osteosynthesis to prevent fractures.[6]

To the best of our knowledge, this case has not been reported with such clinical presentation (i.e., age group), radiological features (i.e., diameters of the lesion and bone marrow edema), or intrinsic healing after surgical open biopsy.

CONCLUSION

Mixed pattern ABCs have rarely been reported in the literature. It may share similar clinical presentation, radiographic features, and age distributions to that of osteosarcoma, especially the telangiectatic type. Treatment modalities of large lesions should include adequate rigid fixation internal fixation using intramedullary nails or plate osteosynthesis, especially if located in the diaphysis of long bone.

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ETHICAL APPROVAL

Approved by the Research Ethics Committee at King Fahed Medical City, Riyadh, Saudi Arabia. IRB number H-01-R-012, dated March 31, 2022.

AUTHORS' CONTRIBUTIONS

AA wrote the initial and final drafts of the article and provided logistic support. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

USE OF ARTIFICIAL INTELLIGENCE (AI)-ASSISTED TECHNOLOGY FOR MANUSCRIPT PREPARATION

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

DECLARATION OF PATIENT CONSENT

The authors' certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

CONFLICTS OF INTEREST

There are no conflicting relationships or activities.

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