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CASE REPORT

Diaphyseal aneurysmal bone cyst: A case report

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Abstract

Aneurysmal bone cyst (ABC) is benign lesion of long bone metaphysis, but it can also become locally aggressive lesion. ABC affects mostly medullary region of long bone metaphysis that predominantly affects children and young adults. Meta-diaphysis of the long bones and posterior elements of the vertebrae are the most common sites of involvement. ABCs can involve every skeletal site, but the most common locations are the metaphysis of the long bones, followed by the spine. However, diaphyseal lesions may occur. Patients with ABCs typically present with insidious onset of pain, swelling or a palpable mass. The imaging appearance of ABC is variable; however, a lytic and expansile lesion with fluid-fluid levels is the most common presentation. Although incidence of intracortical variety of ABCs is rare but surgeons might evaluate its severity in view of aggressive benign or malignant lesions of diaphysis. We present the case of a 11-year-old female with left shaft ulna ABC.

Keywords: cyst, aneurysmal, bone, metaphysis, children

Introduction

Aneurysmal bone cyst (ABC) is a benign but locally aggressive lesion that predominantly affects children and young adults.[1] ABC are benign lesions of long bone metaphysis affecting mostly medullary region. These may be originated rarely either in the cortex or in the superficial regions of diaphysis.[2]

The most common sites of involvement are the meta-diaphysis of the long bones and posterior elements of the vertebrae.[3] ABCs can involve every skeletal site, but the most common locations are the metaphysis of the long bones, followed by the spine. However, diaphyseal lesions may occur. Patients with ABCs typically present with insidious onset of pain, swelling or a palpable mass. Some variability in clinical presentation exists because ABCs, even in the same location, can have different growth rates with doubling times ranging from months to years. This variability in growth rate also contributes to the differences in imaging appearance.[1]

As with any bone tumor, the initial evaluation of ABCs should be done with radiographs followed by magnetic resonance imaging or less frequently computed tomography for further characterization. The imaging appearance of ABC is variable; however, a lytic and expansile lesion with fluid-fluid levels is the most common presentation.[1] Although incidence of intracortical variety of ABCs is rare but surgeons might evaluate its severity in view of aggressive benign or malignant lesions of diaphysis.[2] We present the case of a 11-year-old female with left shaft ulna ABC and describe the symptoms, signs, and radiographic appearance of the ABC. She underwent an MRI that suggested ABC.

Case Report

A 12-year-old female came to our institution with complaints of a lump on left forearm since 1 year ago. The lump is getting progressively enlarged. Complaints accompanied by pain that increases with acti-

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vity. There was a history of trauma 2 years ago, the patient fell from a bicycle with the right arm supporting the body.

On physical exam at left forearm revealed there was a painful and solid lump, measuring 6x4×2 cm, regular borders, firm borders, wrist joint range of motion within normal limits. Conventional Xray (figure 1) show a minimally septated expansile lytic lesion of the left os ulna diaphysis, suspect primary bone tumor which is aneurysmal bone cyst. MRI of left antebrachii (figure 2) revealed a solid mass with a cystic component inside, in the distal 1/3 area of the left ulna diaphysis that gives isointense signal intensity changes with hypointense components inside on T1WI, as well as hypointense with hyperintense components inside on T2WI and T2FS, and gives a partially restricted area on DWI-ADC with an ADC value of 1.0-1.3 x 10-3 mm2/s suggestive of aneurysmal bone cyst. The patient underwent biopsy (figure 3), confirming the diagnosis of aneurysmal bone cyst of the left forearm.

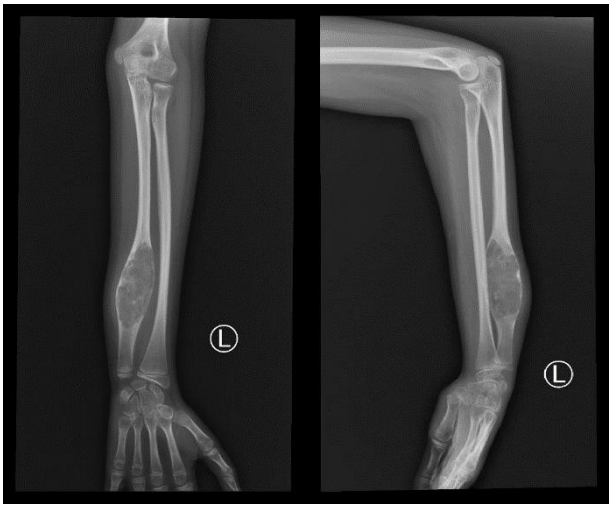

Figure 1. Conventional Xray
(There was a minimally septated expansile lytic lesion of the left os ulna diaphysis, suspect primary bone tumor which is aneurysmal bone cyst.)

Discussion

Aneurysmal bone cyst (ABC) are expansile, hemorrhagic, non-neoplastic lesions that can be locally destructive and that can arise either de novo or secondary to another benign or malignant lesion.[4] Almost 80% of these lesions present in the first two decades of life and have prediction to affect the metaphyseal region of the long bones.[2]

ABC is a descriptive term in which the word “aneurysmal” refers to the marked expansion and the word “cyst” refers to “fluid-filled cavities.”[1] ABC is a rare neoplasm with an annual prevalence of 0.32 per 100,000 young population, 0.14 per 100,000 general population, and comprising about 2.5% of all bone tumors.[3]

Historically, it was thought that an ABC develops as the result of an underlying vascular event; increased venous blood flow; or a reaction to prior trauma. However, in light of the recent molecular findings of the recurrent rearrangement involving the USP6 gene (chr.17p13.2 locus). ABC is now considered a neoplasm rather than a reactive lesion.[3] The most frequent sites are the humerus, the femur, the tibia, and the fibula. Other sites that may be affected are the skull and the posterior elements of the spine.[5]

Considering the nature of the other associated bony lesions, three stages are recognized in the formation of the ABC, First is the initial phase, with predominant osteolysis and non- characteristic appearance. Second is the growth phase, with the rapid growth of the tumor, marked bone destruction, and expansion of the bone. The tumor is not circumscribed, and bony septa are indistinct. Progressively, the first signs of a bony shell appear around the tumor and the last stage is the stabilization phase, with a well-defined unilocular or multilocular radiolucency with histological features of blood-filled sinusoidal spaces along with fibrocellular connective tissue stroma.[6]

The diagnosis is mostly based on radiological findings and imaging, showing bone lesions (cortex, medullary or mixed) and surrounding soft tissues.[2] Radiographic findings often aid in narrowing down the differential diagnoses. As a general rule, any epiphyseal or diaphyseal-based lesion raises the possibility of ABC-like changes more than the primary ABC.[3] Diaphyseal lesions present radiologically as an osteolytic lesion, similar to metaphyseal one but occur eccentrically with thinning of surrounding cortex, thus appear as expansile fusiform lesions with multiple septae.[2]

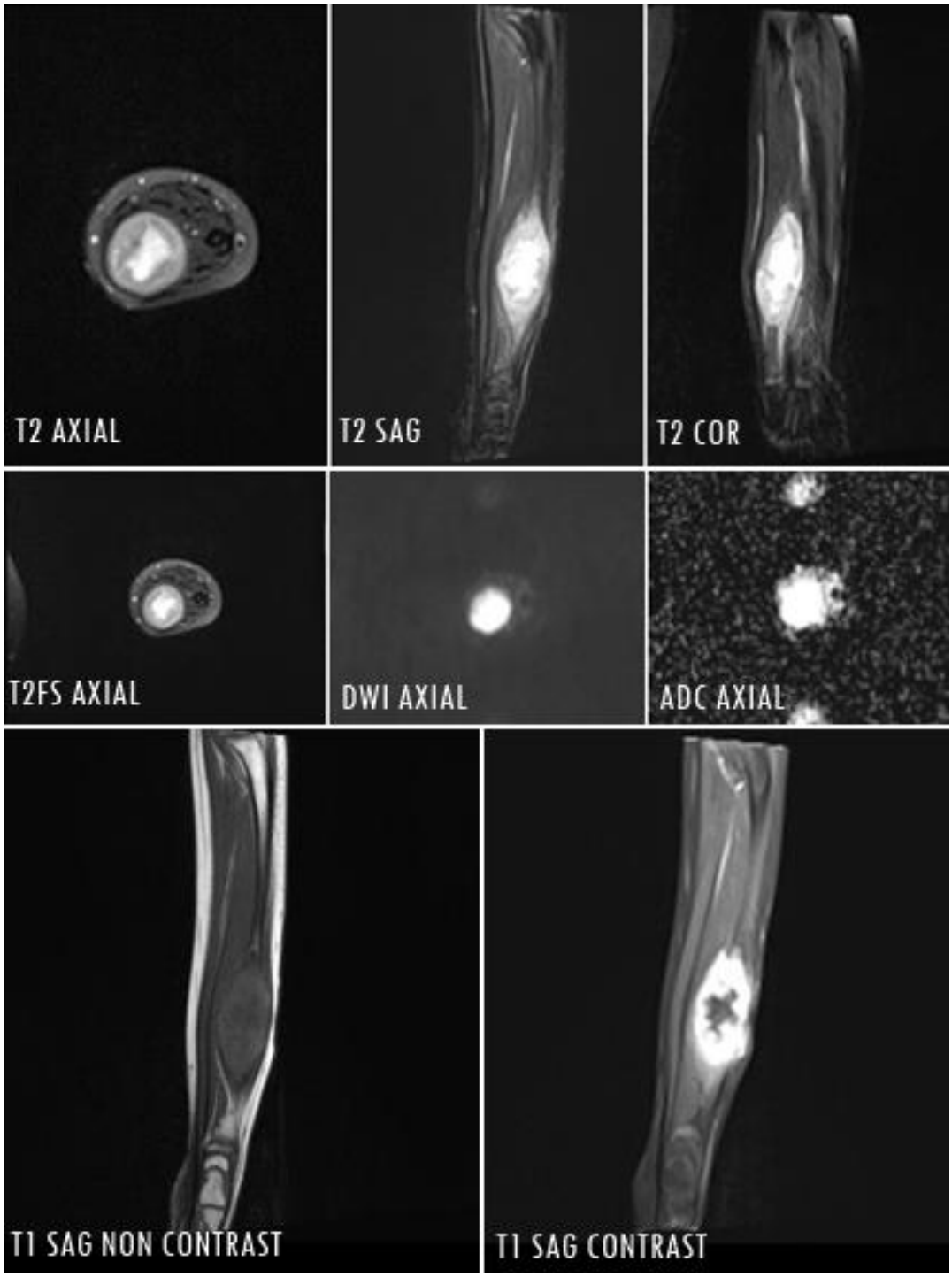

Figure 2. Left antebrachii MRI
(There was a well-demarcated, regular-edged mass with a fluid component inside, measuring approximately 2.63 x 2.67 x 5.78 cm, no periosteal reaction in the area of the distal 1/3 of the diaphysis of the left ulna bone which gave a change in signal intensity of isointense with a hypointese component inside on T1WI, and hypointense with a hyperintense component inside on T2WI and T2FS, and gave a partially restricted area on DWI-ADC with an ADC value of 1.0-1.3 x 10-3 mm2/s. Post contrast scanning provides enhancement at the edges. The scanned neurovascular and muscular structures still provide normal shape and signal intensity. There is no infiltration or destruction of the surrounding bones.)

The radiographic features of ABC are quite distinct and aid in diagnosing the disease. Conventional radiographs show an eccentric radiolucent lesion with expansile remodeling of bone. A thin surrounding rim of the periosteum and sub periosteal bone is usually present. The cyst wall trabeculae impart the multilocular appearance. The best imaging modality to identify the fluid-fluid levels is magnetic res- onance

imaging. The cysts usually demonstrate variable signal intensity with a rim of low T1 and T2 signal. T1 post-contrast sequence may show some enhancement of septations.[3]

MRI, besides lacking ionizing radiation, has the advan- tage of a better contrast resolution, thus allowing a pre- cise delineation of the extent of the lesion (e.g., physeal/ epiphyseal involvement, soft-tissue extension), perilesional edema and the internal composition (e.g., fluid- fluid levels, enhancing nodularity, septations, soft-tissue mass). Fluid- fluid levels of different signal intensity, which are better identified on MRI (especially on fluid sensitive sequences compared to CT scan, are highly suggestive but not pathognomonic of ABCs.[1]

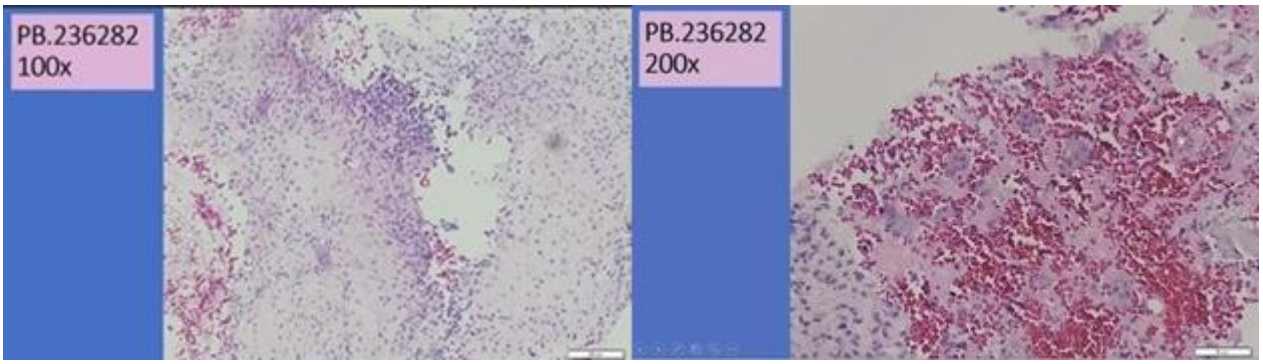

Figure 3. The left fore arm appeared to have a cavity wall lined with connective tissue stroma consisting of hyperplastic, compacted fibroblast cells, with nuclei within normal limits. Some of them appear to be hemorrhages. Osteoclast like giant cells were also seen. Blue chondroid like material was seen. No woven bone structure was seen. No malignant signs were seen.

The diaphyseal lesions start in the cortex of diaphysis and gradually involve adjacent cortex with microfractures leading to the deformities and even the pathological fractures. The chances of early pathological fractures may be due to destructive nature of lesion and their location in the diaphyseal area, which is relatively away from the joint providing a long lever arm causing notable mechanical forces, leading to the breakage of bone. These lesions originated in cortex and progressed gradually, resulting as a pathological fracture, hence usually required internal fixation along with curettage and bone grafting.[2]

Conclusion

ABC is a benign but locally aggressive bone lesion. ABC more prone to pathological fractures having long lever arm at diaphyseal location and stress riser at the site of cortical destruction. Therefore it is important to detecting and distinguishing it from conventional type. Our case is an example of a diaphyseal aneurysmal bone cyst that was diagnosed using MR.

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