

Aneurysmal Bone Cyst in an Adult Patient Located in the Femoral Head and Neck; Positive and Differential Diagnosis

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Abstract

Aneurysmal bone cyst (ABC) is a benign but locally aggressive primary bone tumor, predominant in children and young adults. It can affect any bone in the body with possible soft tissue involvement, but the metaphysis of the long bones is the preferred location. The systemic approach to make the diagnosis of this lesion consists of simple X-rays, however cross-sectional imaging, especially MRI, allows more details and the exclusion of differential diagnoses, mainly telangiectatic osteosarcoma. We report the case of a 40-year-old man who presented with a painful hip whose radiograph showed a well-defined lytic lesion of the head and neck of the right femur revealed to be an ABC on MRI. We approach through this case the main diagnostic criteria of an ABC and the mimicking situations including the "ABC-like".

Keywords: Aneurysmal Bone Cyst; Extensive Pancephalic; Lesion; MRI; Differential Diagnosis

Introduction

Aneurysmal bone cyst is a benign primary bone tumor in which the word "aneurysmal" refers to the marked expansion and "cyst" to the fluid-filled cavities. It is relatively a rare lesion accounting approximately 1% of all bone tumors [1,2]. It occurs in 95% in the first three decades [3] with a predilection to long bones in 50% and pelvis in 12% of cases. The long femur is a common location but usually at the level of the metaphysis and not in the head. The old terminology of primary and secondary ABC was changed in 2020 according to the WHO classification into "ABC-likes changes" defining changes characteristic of ABC occurring within preexistent primary bone tumors and representing 30% of cases. These changes are usually seen with chondroblastoma, giant cell tumor, fibrous dysplasia, non-ossifying fibroma, osteoblastoma and osteosarcoma [3-5].

Case Presentation

A 45-year-old man with no medical history, presented to the trauma consultation for a painful right hip since months with no other associated sign. A plain radiograph was performed and revealed a well-defined radiolucent lesion with sharp margins and a discrete sclerotic area (Figure 1) containing fine septa delimiting cavities. An aneurysmal bone cyst was raised at first, however the age of the patient and the location in the head and neck of the femur left room for some doubt. A complementary MRI was decided and revealed an occupying mass of sharp margins containing fluid filled cysts some of which were hyperintense on T1WI, with fluid levels without associated marrow edema and respecting adjacent soft tissues (Figure 2). At injection of gadolinium, there was no remarkable enhancement and the diagnosis of ABC was then confirmed.

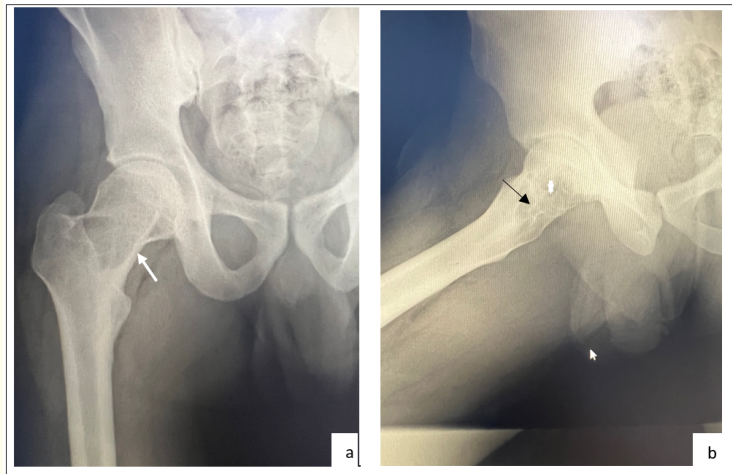


Figure 1: X-Ray of the hip according to anteroposterior (a) and oblique (b) incidences showing a well-defined lytic lesion with a sharp transition zone (white arrow). Thin septa (black arrow) are identified within this lesion delimiting cavities (asterisk).

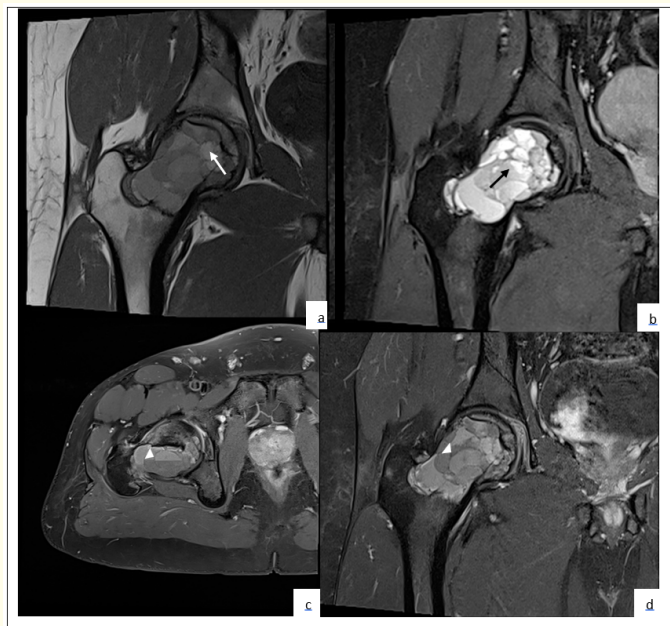


Figure 2: MRI of the hip according to coronal T1WI (a), coronal DPFS (b), axial and coronal T1 with gadolinium injection (c, d) showing a well-limited lesion with inner cavities some of which are slightly hyperintense (white arrow). These cysts are fluid-filled (back arrow) with fluid-fluid levels and internal thin septa that are not enhancing (arrowhead).

Discussion

Positive diagnosis

Aneurysmal bone cyst is a benign cell-rich osteoclastic tumor composed of blood-filled cystic spaces separated by thin septa and surrounded by a thin shell of reactive bone. Symptoms are usually pain with an insidious onset, swelling or palpable mass and may present as acute in the event of a complication such as a pathological fracture or compression of neurological structures, particularly in the spine.

The pathophysiology of ABC is still poorly understood with several theories put forward, some of which evoke the hypothesis of increased pressure leading to bone destruction with an aberrant repair process, others that of trauma. However, the identification of chromosomal translocations in the USP6 gene, although nonspecific and present in approximately 70% of cases, confirms the clonal neoplastic process [6-8]. Although the ABC can affect any bone, the metaphysis of the long bones dominated by the femur and the tibia are the most frequent sites followed by the spine. The presentation in our patient is different by its location at the level of the head and neck of the femur.

There are several proposed classifications for ABC depending on the natural history, the activity and the morphological appearance. The one that could be applied radiologically lists 5 subtypes: Type I is a centrally located lesion with no outline or slightly expanded outline often met in short bones of the hand and feet. Type II are expansile tumors with cortical thinning interesting the whole bone segment. Type III being the most common subtype are eccentric within the metaphysis involving only one cortex. Subtype IV is subperiosteal and growing out of the bone and V is periosteally based with peripheral expansion and extension to the underlying cortex [9].

The first imaging modality to approach the diagnosis of ABC, as the case of all bone tumors is X-Ray where it takes the form of a lytic expansile geographic metaphyseal lesion with possible visible internal septations. A periosteal reaction has also been described, smooth and unilamellar often associated with a healing fracture, with some reported cases of an aggressive one simulating a Codman triangle [10-12]. MRI and CT are useful for diagnosis confirmation fracture prediction and further pre-procedural planning with slightly higher specificity of MRI than X-Ray. It allows an exact delineation of the lesion, extent to soft tissue and visualization of internal composition. Fluid-fluid levels highly suggestive of ABC but not pathognomonic are better identified on MRI than CT. Perilesional edema is sometimes seen and signs a rapid growth. At injection of gadolinium, there is usually a certain degree of enhancement, which has been reported in decreasing order as being peripheral, septal then central diffuse. CT is useful for 3D-reconstructions in some complex anatomical regions such as spine, facial bones, pelvis and hindfoot. It also recognizes better, areas of mineralization, cortical destruction and periosteal reaction when present [13,14].

Differential diagnosis

Differential diagnoses of an aneurysmal bone cyst are variable, as it may occur in any bone, but mostly dominated by unicameral bone cyst, giant cell tumor and telangiectatic osteosarcoma, this latter being the most feared because of its malignant character. Some subtle imaging features are important to recognize and are of paramount importance in setting the correct diagnosis. Telangiectatic osteosarcoma is also an osteolytic lesion but with a broader zone of transition, no sclerotic area, with cortical destruction, soft tissue extension and a higher incidence of pathological fractures seen the rapid growth. Fluid-fluid levels tend to be more peripheral than in ABC, with a mineralized matrix and thick internal septations of nodular enhancement. Lack of USP6 gene rearrangement in telangiectatic osteosarcoma may help confirm the diagnosis. Giant cell tumor appears as a radiolucent eccentric well-defined lesion with no sclerotic margins involving usually the epiphysis in skeletally mature people. On MRI enhancing solid tissue components may be seen associated to soft tissue extension with surrounding marrow edema. Some fluid-fluid levels may also be present representing more "ABC-like" changes. Unicameral bone cyst is usually an asymptomatic bone tumor diagnosed in the second decade of life, in the humerus first followed by

femur. It is a more a geographic lytic lesion with a sclerotic rim centrally located that when associated with a pathological fracture, shows the pathognomonic sign of “the fallen bone fragment” [15-17].

“ABC-like” changes may be seen with other tumors, but a systematic approach taking into account the clinical, epidemiological and radiological features allow the accurate diagnosis. For example, fibrous dysplasia and non-ossifying fibroma occur in diaphysis. Chondroblastoma is characterized by an extensive surrounding inflammatory reaction, but what is more important remains the features of the primary bone lesion.

Several therapeutic options are performed for ABC, going from a conservative approach in some selected cases where spontaneous healing occurs, to aggressive one. These options include, total excision, curettage, sclerotherapy using different agent and selective arterial embolization with variable results. The choice of each attitude depends on the symptomatology, the location of the lesion, its extent and its inner character [18,19].

Conclusion

ABC is, according to the current WHO classification (2020), a benign but aggressive lesion with specific radiological characteristics occurring mainly in the metaphysis of the long bones in children and young adults. However, this is not a rule, and older age or different location are possible, thus requiring careful analysis and synthesis of the various components, especially since some of the differential diagnoses may be malignant.

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Conflict of Interest

Authors declare having no conflict of interest.

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