

Aneurysmal Bone Cyst: With Uncommon Location and Presentation – A Case Series with Review of Literature

Abstract

Aneurysmal bone cyst (ABC) is an expansile osteolytic bone lesion of unknown aetiology. These lesions are usually present in the long bones, particularly the humerus, femur, tibia, and fibula and are composed of blood-filled cavities with the female predominance. ABC is differentiated into two variants such as primary and secondary. Primary ABC arises *de novo*, whereas secondary arises in association with some pathology. Here, we are presenting three unusual cases of ABC; two cases were reported here because of their rare locations such as talus and ulna, whereas the last case was of giant cell tumor with secondary ABC.

Keywords: Aneurysmal bone cyst, bone tumors, histopathology

Introduction

Aneurysmal bone cyst (ABC) is benign and expansile bone tumors occurring in the first two decades of life, which comprises about 1% of the all primary bone tumors. It was recognized first as a distinct pathological entity in 1942.^[1] The incidence is 0.14/100,000 of the population per year, with a slight female predominance. Almost 80% of the lesions occur in skeletally immature patients who are younger than 20 years. It typically affects the metaphysis of long bones, pelvis, and posterior vertebral elements, but ribs, hand, foot, scapula, clavicle, and mandible are rarely involved. They account for 1% of all biopsied primary bone tumors^[2] and appear as rapidly growing destructive lesions that expand the cortices.^[3] The radiographic appearance of ABC is purely lytic, involves the metaphysis of a long bone, and is eccentric. The radiological findings are characteristic which present as lytic and multiloculated lesion with blowout or expansile character and area of overlying cortical disruption. ABC can exist either as a primary bone lesion (70%) or as a secondary lesion; it is called secondary when a preexisting osseous lesion is identified (30%).^[4] Giant cell tumor (GCT) is a benign bone tumor with malignant potential and accounts for 5% of all primary bone neoplasms.

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Cystic components in GCT which indicates secondary aneurysmal cyst are reported in 14% of cases,^[5] and it is a most frequent tumor associated with secondary ABC.

Case Reports

Case 1

A 34-year-old female presented with painful swelling at the left wrist, which gradually increased from a peanut size to lemon size for 1½ years and was associated with restricted movement at the corresponding joint. Clinical examination revealed tender bony swelling and deformity at the left wrist with no neurological deficit. Clinically provisional diagnosis was GCT of the left ulna. Radiological examination (X-ray) showed an expansile multicystic eccentric lesion of the distal end of the ulna with Soap bubble appearance and no periosteal reaction [Figure 1]. The patient was operated, and formalin-fixed specimen was received in the histopathology section of pathology department. The specimen was club-like bone measuring 10 cm × 6 cm × 6 cm; its cut surface showed multiloculated soft bony friable filled with hemorrhage and myxoid material. Tissue was processed and stained with hematoxylin and eosin. Sections examined show various size of cyst separated by septa. The cysts contain blood. The septa are composed of loosely arranged spindle cells, capillary proliferation, and benign giant cells. The space is lined by cuboidal-appearing cells.

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On the basis of location, clinical-radiological investigation and histopathology present case was diagnosed as ABC [Figure 2].

Case 2

A 28-year-old female presented with pain in the right ankle for 1 year without any evidence of swelling. There was a history of trauma 5 years back. Clinical examination revealed tenderness at the right ankle with no neurological deficit. Her vital parameters were within the normal range. Radiological examination (X-ray) of the right ankle showed a well-circumscribed lesion in the right talus [Figure 3]. Magnetic resonance imaging (MRI) showed ABC right talus with breach posterior wall. Chest X-ray was normal. The histopathology section of pathology department received multiple irregular soft-tissue pieces measuring 2 cm × 2 cm and multiple bone pieces measuring 0.7 cm × 0.5 cm × 0.3 cm. Histopathology examination showed stroma intermingled with osteoclastic giant cell, blood-filled space, and bone trabeculae. All findings suggestive of were aneurysmal bone cyst [Figure 4].

Case 3

A twenty six years patient presented with pain over lower one third of leg, tenderness for 6 months in Out patient Department of UPUMS, Saifai, Etawah (Uttar Pradesh). Clinical examination and Xray findings were suggestive of osteolytic lesion over distal tibia [Figure 5]. Patient was operated and curettage was replaced with bone graft from iliac crest. The curettage was having bony and soft tissue pieces grey/ brown in colour and aggregating 4cmx3cmx1 cm. Histopathology examination revealed numerous osteoclast giant cell, cystic spaces filled with blood, [Figure 6] osteoid elements and fibrous tissue. In between singly arranged atypical cells with appreciable mitotic figures was also [Figure 7]. All above finding were of secondary aneurysmal bone cyst with giant cell tumor.

Discussion

The WHO defines ABC as a benign expansile tumor-like osteolytic lesion of bone consisting of blood-filled spaces separated by connective tissue septa containing trabeculae



Figure 1: Anteroposterior and lateral view radiographic image of ulna showing soap bubble appearance

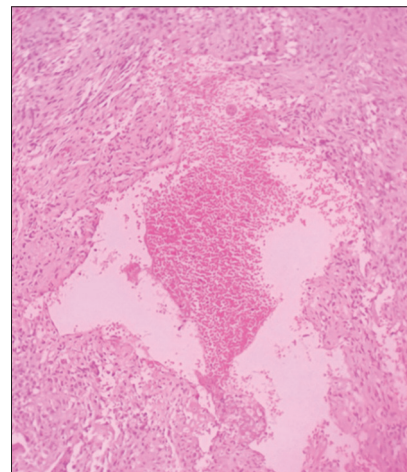


Figure 2: Section showing cystic spaces surrounded by osteoclast like giant cells (H and E, x10)



Figure 3: Radiological examination X-ray of the right ankle showed well-circumscribed lesion in right talus

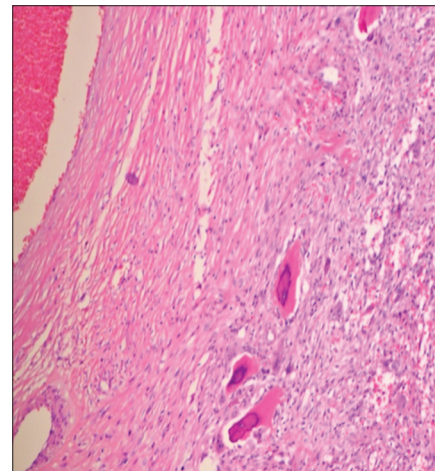


Figure 4: Histopathology examination showed stroma intermingled with osteoclastic giant cell, Blood filled space and Bone elements



Figure 5: Radiographic image of tibia showing osteolytic lesion

or osteoid tissue and occasionally osteoclastic giant cells. It was first described as a distinct entity in 1942 by Jaffe and Lichtenstein^[1] and named by these researchers as aneurysmal bone cyst in 1950. Nowadays, its neoplastic nature is proved, which is characterized by rearrangements of the USP6 gene and USP6 and CDH 11 oncogenes had been identified only in primary ABC whereas absent in secondary ABC.^[6]

This disease is common in the second decade of age and lesions usually present in the long bones.^[3,5] The cause for its origin is strange; there are several examples of ABC apparently arising following fracture. In the present case series, all the cases were found in young age females, and in one case, there was a history of trauma.

Researchers also believe that the ABCs are the result of a vascular malformation within the bone, though the ultimate cause of the malformation is still under debate,^[7] and it affects long bones, pelvis, and posterior vertebra. In the present case series, the two cases were reported because of rare location such as ulna and talus.

On examination, the appearance of the ABC is a subperiosteal, metaphyseal eccentric, or concentric lesion, elevating and inflating the periosteum and progressively eroding the cortex. There are also many septa and ridges in the osteolytic area (honeycomb appearance).^[2] Computed tomography and MRI imaging studies are useful in evaluating soft tissue and intramedullary extension of the lesion,^[3] but they are not always conclusive, and ABC sometimes mimic as eosinophilic granuloma, giant cell tumor, and unicameral bone cyst.^[8] During bone scintigraphy, the increased uptake reflects the true pathologic extent of the lesion. In most cases, the abnormally increased uptake is located around the periphery of the cyst, with less activity in the center (ring-like pattern).^[9]

Histopathology is mandatory for the diagnosis of ABC.^[10] On gross examination, these lesions are frequently curetted and red-brown, granular, fragmented material, whereas the

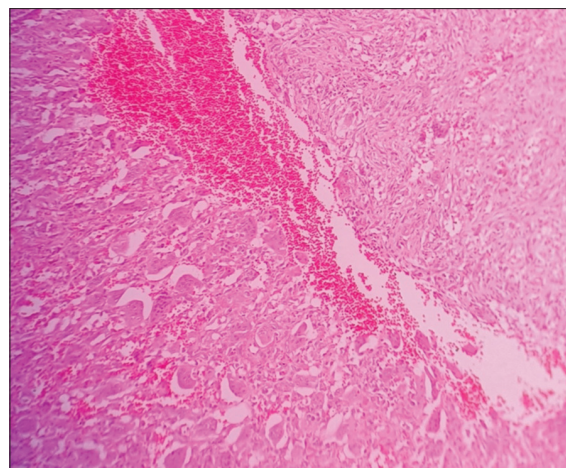


Figure 6: Section showing cystic spaces filled with blood surrounded by osteoclast like giant cells (H and E, ×10)

cut surface shows hemorrhagic, multicystic spaces filled with blood, separated with septa. The same gross findings were seen in our cases.

Its differential includes GCT, low-grade osteogenic sarcoma, and telangiectatic osteosarcoma, differential from these conditions are required to confirm the diagnosis. GCT occurs in long bones and in skeletally mature persons, whereas ABC occurs in skeletally immature persons with metaphyseal as location. Low-grade osteogenic sarcomas are less cellular and have great mitotic activity than ABC. In regards to telangiectatic osteosarcoma, septa in telangiectatic osteosarcoma have pleomorphic atypical cells, whereas as there is no atypia in ABC septa. In the current study, above features were viewed to confirm diagnosis and exclude differential.

There are two theories regarding the genesis of secondary ABC, first suggests that vascular anomaly in the primary bone lesion or a reactive bone growth leads to hemodynamic changes, while other believe that neoplastic origin with chromosomal abnormalities. Despite such theories, etiology, and pathogenesis of this benign, vascular destructive lesion remains unclear.^[11]

Nearly 30%–50% of ABC occurs secondary to primary tumors such as GCT, chondroblastoma, chondromyxoid fibroma, fibrous dysplasia, and osteogenic sarcoma.^[12] GCT is the most prevalent precursor lesion of ABC developing secondarily. GCT is a benign bone tumor which occurs predominantly in the meta epiphyseal regions of the long bone, typically in the distal femur and proximal tibia. Cystic components in it indicate secondary aneurysmal bone cyst. When ABC occurs with GCT bone, MRI finding is able to identify characteristic fluid level as well as solid foci, suggestive for secondary ABC. However, fluid levels also appreciated in GCT, chondroblastoma, simple bone cyst, and telangiectatic osteogenic sarcoma.^[13]

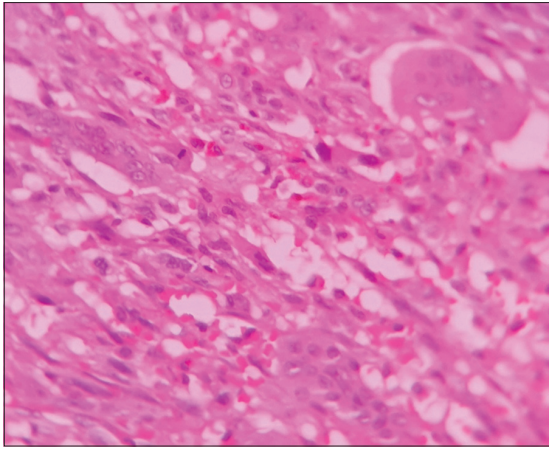


Figure 7: Section showing giant cell, spaces filled with blood and stromal cells having focal mitosis (H and E, ×40)

Therefore, the confirmative diagnosis is based only on histopathology examination. However, the presence of ABC has no implications as far as the management or the outcome of treatment of GCT of bone is considered, but recognizing these patterns is important in differential diagnosis and correct diagnosis.

Conclusion

The diagnosis of ABC is mandatory and specially when its location is other than usual sites and associated with some other pathology. Histopathology is an excellent and confirmative diagnostic tool. Hence, it is important to highlight the associations between imaging and histopathology findings. During treatment decision-making, there are many options with curettage being the most common. Recurrence is a common complication after their treatment, and follow-up is always necessary with attention regarding any change in malignancy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal.

The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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