Case Report

Aneurysmal Bone Cyst: An Uncommon and Aggressive Presentation

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ABSTRACT

The solid variant of aneurysmal bone cyst (ABC) is a rare subtype of aneurysmal bone cyst with a preponderance of solid compared to cystic elements. We present a case of solid ABC affecting the proximal tibia-an unusual site, and presenting with rapid progression mimicking malignant neoplasm.

A 12 year old girl presented with pain and swelling, just below left knee for 9 months. X-Ray and CT scan demonstrated a lytic expansile lesion over metaphyseal region of tibia and diagnosed radiologically as ABC. Histology of curettage established it as solid variant of ABC. After 5 months, patient presented with a fungating mass over skin below knee with foul smelling discharge. Histology of incisional biopsy from outside suggested osteosarcoma, though no osteoid was found. Above knee amputation was done and histology of the whole specimen conclusively proved it to be a case of solid variant of ABC.

Solid ABC has been of great interest because it may be mistaken for malignant tumor, mainly giant cell tumor, osteosarcoma or synovial sarcoma, because of cellularity and variable mitotic activity.

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Introduction

An aneurysmal bone cyst (ABC) is a benign, often rapidly expanding, locally destructive cystic lesion of the bone. Although ABC affects all age groups, the peak incidence is in the first and second decades of life. It accounts for about 1% to 2% of biopsied primary bone tumors. It may be of conventional or solid type. [1,2] The solid variant is a rare sub-type, and it is more difficult to recognize. The term "giant cell reparative granuloma" has been used as a synonym in the pathology literature to describe this variant. Locations include the small bones of the hand and feet, vertebrae, sacrum, and less commonly – long bones. [3] Although it is considered to be reactive in nature, there is evidence that some ABCs are true neoplasms. We present a case of solid ABC affecting the proximal tibia, and presenting with rapid progression mimicking malignant neoplasm.

Case Report

A 12 year old girl from Bihar presented to us with complaints of pain and swelling just below left knee for last 9 months. On X-Ray, a lytic expansile septate lesion was found over metaphysis of tibia. Joint was spared. Provisional diagnosis of ABC was made from X-Ray (Fig. 1A). Her hematological and biochemical parameters were within normal limit. On examination, single ipsilateral lymph node (1x0.5cm) was found in inguinal region. Curettage was attempted. On OT, whole bone fragments came out with haemorrhage and necrotic material. Histopathological examination (HPE) was done and sections(Figure 1B and 1C) showed fragments of bone and a mass predominantly localized in soft tissue composed of cystic and solid areas. Mass was composed of oval to spindle cells diffusely arranged and osteoclastic type giant cells were scattered among the stroma. Mitotic activity was brisk, but atypical mitosis was absent. Very small foci of necrosis were present. A provisional diagnosis of solid variant of Aneurysmal Bone Cyst was made. Patient was put on follow up and discharged. Lymph node found in inguinal region showed reactive hyperplasia.

Four months later, patient came with severe bleeding over wound site and was complaining of weakness. Hb% was 3.6 gm/dl. Color Doppler study revealed arterial sprouting at wound site. Embolisation was done followed by pressure packing over wound. Again, after one month, patient came with a fungating mass over wound (Fig 2A) with foul smelling discharge. Culture sensitivity of the discharge revealed Pseudomonas and appropriate antibiotic treatment was given. Patient was complaining of continuous boring pain in the whole limb. X-Ray (Fig. 2B) showed destruction of upper tibia with cortical breach at joint space. MRI (Fig. 2C) revealed soft tissue extension, skin involvement over wound and almost whole upper end of tibia was lytic. No calcification was noted in MRI. Incision biopsy from the fungating mass was done and sent to two different labs. One reputed private laboratory gave diagnosis of osteosarcoma, although no osteoid formation could be detected. We reviewed the sections and stuck to the diagnosis of solid variant of ABC (Fig 2D).

Decision of amputation with whole mass excision was taken in Medical Board partly due to discrepancy of diagnosis and also because reconstruction was not possible as joint cavity was left with no or little tibial shaft. Above knee amputation was done after obtaining informed consent. Whole mass was sent for Histopathological Examination. During the whole period although the mass was rapidly progressing, but patient's general condition was not deteriorating. Chest x-ray and USG abdomen did not reveal any lesion in lung or liver.

Gross examination of amputated specimen showed a globular mass measuring 19x11x5cm, with grayish white outer surface with blackish and necrotic cystic areas and ulceration of skin. On serial sections multiple well-circumscribed blood filled cavities were seen separated by fibrous bands (Fig. 2E). No calcification was noted in the mass.

Microscopic examination showed spindle cells in haphazard arrangement interspersed with blood filled cavities. The cells showed minimal pleomorphism with focal reactive atypia, necrosis and few scattered mitotic

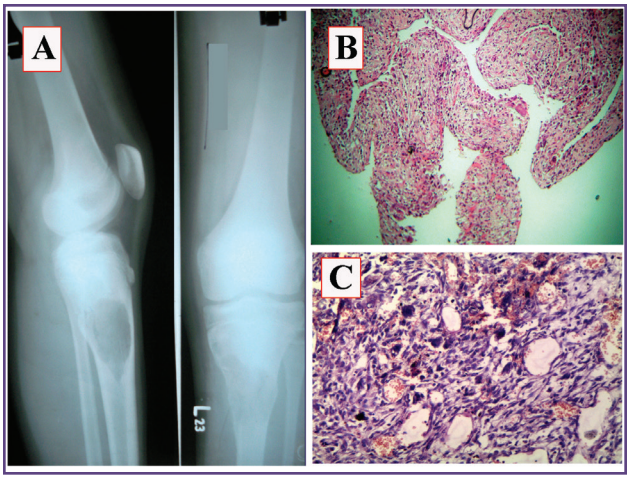

Fig. 1A : Initial x-ray showing punched out osteolytic lesion in metaphysis, joint cavity intact. 1B: Initial curettage showing cystic solid mass predominantly consist of plump spindle cells (x200, H&E) 1C: Initial curettage showing scattered osteoclastic type giant cells and cystic spaces without endothelial lining (x400, H&E).

figures (Fig. 3D). However, atypical mitosis could not be detected in any sections. Hypocellular areas with fibromyxoid degeneration (Fig. 3C), inflammatory cells and histiocytes were scattered at places. Stromal spindle cells were arranged in storiform pattern in few places (Fig. 3A). Collection of osteoclast like giant cells was present haphazardly. The blood filled cavities lacked any endothelial lining and were lined by fibroblasts and giant cells instead (Fig. 3B). Osteoid formation could not be detected in any of the sections despite thorough search. Overall histopathological features were suggestive of solid aneurysmal bone cyst/ giant cell reparative granuloma.

Patient was then put on follow up with prosthesis. Currently patient is doing well.

Discussion

Solid ABC is a rare ABC variant with the same or similar clinical, radiologic features and prognosis as a conventional lesion of an ABC, but with the preponderance of solid to cystic elements. In 1983, Sanerkin et al. [4] first described a variant of ABC in which the predominant histological features were solid, and they used the term "solid variant of aneurysmal bone cyst".

Solid aneurysmal bone cysts are most commonly located in the small bones of the hand and feet, vertebrae, sacrum, and less commonly involves long bones of extremities. [3] In the latter locations, they tend to have a metaphyseal location.

In our case, considering the age group, site of tumor and aggressive presentation, the following possibilities were kept in mind as differential diagnosis: Giant Cell Tumor (GCT), Osteosarcoma, spindle cell sarcomas like Malignant Fibrous Histiocytoma (MFH), Synovial Sarcoma, Fibrosarcoma. Histological examination remained the main tool in differentiating solid ABC from other type of tumors.

Although Giant cell tumor is rarely seen in patients below 20 years of age, it is more common in women and the classic location is the epiphysis of a long bone, from which it may spread into the metaphyseal area, break through the cortex, invade intermuscular septa, or even cross a joint space. The sites most commonly affected are the lower end of the femur, the upper end of the tibia. Histologically Giant cell tumors have a uniform distribution of the giant cells and diffuse arrangement of round to oval stromal cells. In our case histologically the giant cells were usually scattered or gathered in small clusters around the cystic spaces which lacked endothelial lining. The stromal cells were spindle-shaped without histologic evidence of sarcomatous transformation. These favored more towards a non-GCT neoplasm like ABC. [5]

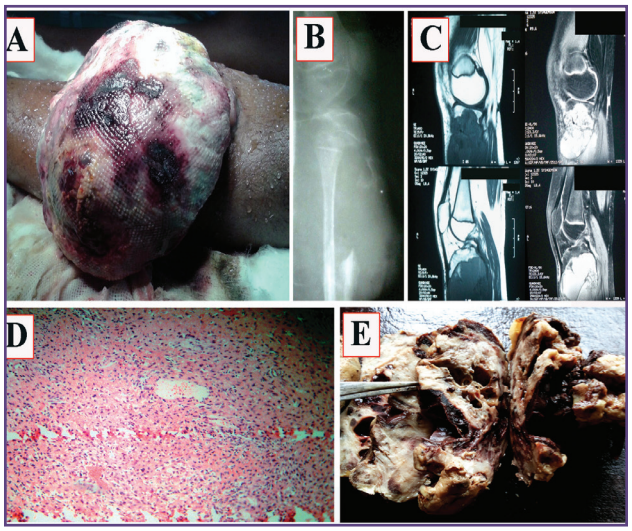

Fig. 2A: Fungating mass over leg involving skin and soft tissue 2B: X-ray showing destruction of upper tibia with cortical breach near joint space. 2C: MRI of left upper tibia showing contrast enhancing mass extending from bone into joint and soft tissue 2D : solid sheet like plump spindle cells with focal atypia (x400, H&E) 2E : gross picture of amputated mass showing blood filled cystic and solid septate areas.

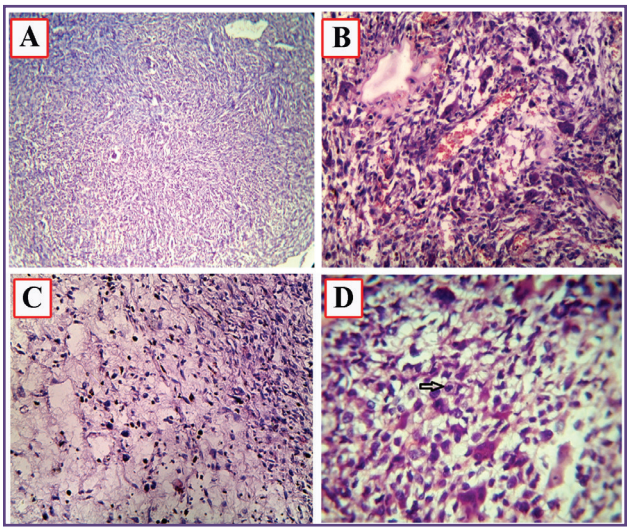

Fig 3A : Storiform pattern of spindle cell in amputated specimen (x200, H&E) 3B :Scattered osteoclastic type giant cells, vascular spaces, and cysts without endothelial lining (x200,H&E) 3C : Myxoid hyaline areas in tumour mass (x400, H&E) 3D :Mitotic figure in tumour mass (x400, H&E)

Osteosarcoma usually occurs in patients between 10 and 25 years of age. Most osteosarcomas are located in the metaphyseal area of the long bones, particularly the lower end of the femur, the upper end of the tibia, and the upper end of the humerus. Both age distribution and tumor location were similar but in our case the pattern of reactive bone production was different, from the malignant hyaline lacy osteoid that one expects to see in low-grade and high-grade osteosarcomas [6]. In ABCs, the reactive bony trabeculae are lined by plump, benign-appearing osteoblasts, which are not a feature of osteosarcomas. The stromal cells also showed less atypia and no atypical mitosis which further point towards a benign lesion like ABC.

Spindle cell sarcomas like MFH, Synovial Sarcoma and Fibrosarcoma were considered as differential diagnosis because of the very aggressive presentation of the tumor and they also show storiform pattern and cellular atypia [7, 8]. But the high degree of nuclear pleomorphism and atypical mitosis which are typical in these sarcomas were lacking in our case. Clinico-radiological correlation, absence of atypical mitotic figure and typical immunohistochemical features may help to distinguish.

Although ABCs are known to be non-neoplastic and probably reactive lesions, more recent studies have shown that at least some of the ABCs of all types are of a neoplastic nature, owing to clonal chromosomal aberrations like clonal rearrangements of chromosomal bands 16q22 and 17p13, indicating a neoplastic basis in at least some ABCs[9].

Differentiation of ABC from other malignant bony neoplasm is of utmost importance, because chemo-radiation is not indicated in ABC; also prognosis is drastically different. Aneurysmal bone cysts are usually treated by thorough curettage. Occasionally they recur, and further curettage with either cryosurgery or phenol instillation can be used[10].

Conclusion

Solid variant of ABC is a challenging entity both to clinicians, radiologists and histopathologists, because of its diverse morphological picture in different areas and resemblance to other mesenchymal neoplasm. Immunohistochemistry is not very helpful in these cases. The unique feature of this case is aggressive presentation of a benign bony neoplasm.

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Competing Interests

None declared

Reference

1. Bertoni F, Bacchini P, Capanna R, Ruggieri P, Biagini R, Ferruzzi A, *et al*. Solid variant of aneurysmal bone cyst. Cancer 1993;71:729–34.
2. Unni KK, Inwards CY. Tumors of the osteoarticular system. In: Fletcher CDM, editor. Diagnostic Histopathology of Tumors, 3rd edn. Vol. 2. New York: Churchill Livingstone; 2007;p.1634–36.
3. Ilaslan H, Sundaram M, Unni KK. Solid variant of aneurysmal bone cysts in long tubular bones: giant cell reparative granuloma. Am J Roentgenol 2003;180:1681-7.
4. Sanerkin NG, Mott MG, Roylance J. An unusual intraosseous lesion with fibroblastic, osteoclastic, osteoblastic, aneurysmal and fibromyxoid elements. "Solid" variant of aneurysmal bone cyst. Cancer 1983;51:2278-86.
5. Chakarun CJ, Forrester DM,Gottsegen CJ,Patel DB, White EA,Matcuk GR. Giant Cell Tumor of Bone: Review, Mimics, and New Developments in Treatment. RadioGraphics 2013;33:197–211.
6. Klein MJ, Siegal GP. Osteosarcoma: Anatomic and Histologic Variants. Am J Clin Pathol 2006;125:555-81.
7. Fisher C. Synovial Sarcoma. Ann Diagn Pathol 1998;2(6):401-21.
8. Eyden BP, Manson C, Banerjee SS, Roberts ISD, Harris M. Sclerosing epithelioid fibrosarcoma: a study of five cases emphasizing diagnostic criteria. Histopathology 1998;33:354–60.
9. Sciot R, Dorfman H, Brys P, Dal CP, De Wever I, Fletcher CD, *et al*. Cytogenetic-morphologic correlations in aneurysmal bone cyst, giant cell tumor of bone and combined lesions: a report from the CHAMP study group. Mod Pathol 2000;13:1206-10.
10. Schreuder HWB, Veth RPH, Pruszczynski M, Lemmens JAM, Koops HS, Molenaar WM. Aneurysmal bone cysts treated by curettage, cryotherapy and bone grafting. J Bone Joint Surg [Br] 1997;79-B:20-5.