CHONDROBLASTOMA WITH SECONDARY ANEURYSMAL BONE CYST

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ABSTRACT
A devastating rare tumor chondroblastoma (calcified giant cell tumor, chondromatous osteoclastoma) is a cartilage forming tumor which occurs commonly in individuals below the age of 20 years. Males are affected more frequently with a ratio of 2:1. Most common site of tumor is epiphyseal end of long bones before the epiphyseal cartilage disappears; in distal end of femur, proximal end of humerus, proximal end of tibia. Sometimes it arises in metaphysis or arises in epiphysis and extend into metaphysis. On imaging the tumor is seen as a well delineated lytic lesion with areas of rarefaction. The reason behind presenting this case is that Chondroblastoma with Secondary Aneurysmal bone cyst is rare. Case Presentation: 21 year male presented with complains of pain in the left shoulder since 2 years. He was asymptomatic 2 years back, developed pain in left shoulder during lifting of weight which increases gradually. On examination there is a tenderness over the joint line without any inflammatory signs. There is limitation of movement of joint when abducting beyond 90 and in flexion beyond 30. X ray shows an expansile mass in the upper end of humerus. CT scan shows a lytic lesion in the epiphysis of left humerus. Excision and curettage with femoral head bone grafting was performed. The tumor was sent for histopathology. Histopathological studies were performed and Chondroblastoma with Secondary Aneurysmal Bone Cyst was diagnosed. Chondroblastoma with aneurysmal bone cyst is very rare. Present study suggests that this tumor should be included in the differential diagnosis of any lytic lesion of bone.

KEYWORDS: Chondroblastoma, Aneurysmal bone cyst, lytic lesion.

INTRODUCTION
Chondroblastoma is a rare cartilage forming tumor that accounts for less than 1% of primary bone tumors. The tumor arises from chondroblasts of epiphyseal plate. Metaphysical or diaphyseal Chondroblastomas are rare which occurs in less than 2% of cases. In non-epiphyseal cases, the tumor is intramedullary.² Chondroblastoma occurs commonly in individuals below 20 years of age. But cases are reported in older individuals also. Males are affected more frequently with male to female ratio of 2:1. Though this tumor is benign it has some unusual features like mitotic activity, necrosis, and cellular atypia.³ Along with unusual features it has unusual locations also like skull (Clivus), long bone diaphysis (tibia and humerus), facial bones and small tubular bones of hands and feet (navicular bone).⁴⁵⁶

Chondroblastoma of humerus is rare. The most frequent sites for Chondroblastoma are proximal part of the humerus.⁵ Most of the cases present with pain and limitation of movements of joints. But clinical features depend upon the size and location of tumor. Severe headache, left facial dysesthesias and diplopia is seen in Chondroblastoma involving the clivus.⁶ The tumor is limited to the bone but sometimes it becomes aggressive and invades the surrounding soft tissues.²³ Chondroblastoma is a benign tumor but some studies report malignant transformation of this tumor along with metastasis.⁷⁸¹⁶ The cell of origin of Chondroblastoma is chondrocytes. The tumor arises from transition during endochondral formation. This transition occurs in response to alteration in local environment.¹³ On X ray Chondroblastoma is a radiolucent lesion with smooth or lobulated margin without periosteal reaction. Computed tomography which is helpful in diagnosis shows a lytic lesion with sclerotic rim and calcification.⁹¹³ Microscopy of this tumor shows round to polygonal cells, chicken wire appearance of chondrocytes, osteoclast like giant cells, cystic spaces filled with blood.¹⁵¹⁶

CASE REPORT
Clinical History
21 year male presented to the outpatient clinic of orthopedic department of Osmania general hospital...
(Tertiary Care Centre) with complains of pain in his left shoulder since 2 weeks. Upon enquiry the patient revealed that he was asymptomatic 2 years back, developed pain in his left shoulder which subsides temporarily with Analgesics. For the past 2 months along with pain he observed limitation of joint movement.

**Physical Examination**

There were no scars, no sinuses, no enlarged veins and skin over the swelling was normal. Vital signs were stable. There is tenderness over the joint line with no inflammatory signs. Limitation of movement of jointis present when abducting beyond 90 and in flexion beyond 30.

**Investigations**

CBC, ESR, CRP were within normal limits.

X-ray: showed a lytic lesion in the upper part of humerus Fig 1A.

CT scan: showed a lytic expansile lesion in the epiphysis, eccentric location with sclerotic margin and internal calcification Fig 1 B&C.

**Differential Diagnosis**

1. Aneurysmal bone cyst
2. Chondroblastoma
3. Giant cell tumor
5. Chondromyxoid Fibroma.

**Histopathology**

**Gross**

Received multiple grey white firm masses. Cut sections revealed grey white areas with calcification, necrosis, multiple cystic spaces filled with blood.

**Microscopy**

Hematoxylin and eosin stained sections shows hypercellular lesion with cells arranged in sheets, areas of hemorrhage, necrosis and cysts filled with blood Fig 2A. Individual cells are round to polygonal with distinct cell borders, moderate amount of pink cytoplasm, round to oval nuclei with longitudinal grooves. Scant mitotic activity is seen. Some cells show clear cytoplasm. Scattered in between the tumor cells are osteoclast like giant cells and spindle cells Fig 2B. Deposits of cartilaginous matrix surrounding the individual cells with calcification giving the typical appearance of chickenwire appearance. This feature is pathognomonic of Chondroblastoma Fig 2C. Also seen are cystic and cavernous spaces filled with blood and surrounded by fibrous tissue and intermixed with osteoclast like giant cells Fig 2D & E.
Figure 2: Chondroblastoma with Secondary Aneursmal Bone Cyst.

A- Chondroblastoma with areas of calcification, hemorrhage and necrosis. Cysts filled with blood are also seen.  B- Sheets of round to polygonal cells intermixed with giant cells. C- Calcification surrounding the individual cells giving appearance of chicken wire. D & E –Cystic and cavernous spaces filled with blood and surrounded by spindle cells and intermixed with osteoclast like giant cells. Both cartilaginous and osteoid matrix is seen. (H & E stain - A x100, B x400, C x400, D x100, E x40)

Diagnosis
Chondroblastoma with Secondary Aneursmal Bone Cyst.

DISCUSSION
Chondroblastoma is a benign cartilage forming tumor first described by Codman in 1931. It accounts for 1% of all bone tumors. The tumor arises from epiphyses of long bones. But it can arise from metaphysis as well as diaphysis. Chondroblastoma of upper end of humerus is rare. In a study done by Xu H et al, the most commonly involved sites were proximal part of tibia followed by proximal part of femur and the distal part of femur. Some of these tumors are seen at unusual sites also like temporal bone, squamous portion being most common.

Majority of the tumors are benign but there are cases which show malignant transformation and metastasis to lung. Chondroblastoma is slow growing tumor. Sometimes the tumor grows rapidly and destroys the cortex and soft tissues. But in our case there is no soft tissue destruction as well as there is no metastasis.

Mean age of presentation is below 20 years. Patients usually present with pain and limitation of movement of joints as is seen in our case. Clinical features depend upon the site of involvement and size of tumor. In a study done by Jonathan Liu et al, the main clinical feature in their case was severe headache, left facial dysesthesias, and diplopia. This is because the site of tumor was clivus which is also very rare site. Pain and limp is present if the Chondroblastoma is situated in the
upper part of femur.[7] The cell of origin of Chondroblastoma is chondrocytes. Harner et al states that Chondroblastoma arises from transition during cartilaginous and endochondral formation.[12] Varvares et al states that Chondroblastoma arise from normal chondrocytes that transform in response to alteration in local environment.[13]

Imaging is very important in the diagnosis. On X-ray the tumor appears as a well-defined radiolucent lesion, with either smooth or lobulated margins or a thin sclerotic rim, arising eccentrically in the epiphysis of long bones.

CT Scan shows a lytic expansile lesion in the epiphysis, eccentric location with smooth sclerotic margin and internal calcification without any periosteal reaction. [17]

The Key histopathological features of Chondroblastoma are round to polygonal cells, osteoclastic type giant cells and chicken wire appearance of chondrocytes. [14] These features along with cysts and cavernous spaces filled with blood and surrounded by spindle cells and intermixed with osteoclast like giant cells points the diagnosis towards Chondroblastoma with secondary aneurysmal bone cyst.[15][16] Study conducted by Bloem et al states that in 15% of the Chondroblastoma secondary aneurysmal bone cyst is observed. Chondroblastoma is positive for S100. A new IHC marker which is also helpful in the diagnosis of this tumor is DOG1.[30] Behjati S et al found Mutation in H3F3B in Chondroblastoma cases which is one of the two genes for histone H3.3.[31]

Differentiation between Chondroblastoma and other related benign tumors is important in terms of management and prognosis. Aneurysmal bone cyst (ABC) consists of cystic spaces filled with blood and surrounding fibrous tissue without round to polygonal cells and chicken wire appearance. There is mild reactive bone formation seen in ABC. Chondroblastoma doesn’t have hemorrhagic cystic spaces. Cells with chicken wire appearance and longitudinal grooves in nuclei is absent in giant cell tumor. Chondromyxoid fibroma is composed of hypocellular lobules of hyaline cartilage composed of chondroblasts with pink cytoplasm and myxoidstroma and fibrous septa containing spindle cells. Giant cells are few. Longitudinal grooves and chicken wire appearance of cells in not seen. Giant cell tumor and Chondroblastoma with Secondary ABC has many similarities but the margins of tumor is clearly visible in Chondroblastoma.[22]

CONCLUSION
Histopathological confirmation of diagnosis of Chondroblastoma with Secondary ABC is very important for proper management. Since it’s a benign tumor, excision and curettage with femoral head bone grafting is the surgical procedure performed in our case and there was no recurrence of tumor.

REFERENCES
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