GIANT CELL-RICH LESIONS OF BONE AND JOINTS: A ONE YEAR PROSPECTIVE STUDY

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ABSTRACT

BACKGROUND

Giant cell-rich lesions constitute a group of biologically and morphologically diverse bone and joint tumours. The common feature is presence of numerous multinucleated osteoclast-like giant cells. However, they differ from each other by in terms of clinical and radiographic features and in many cases by their distinct morphological features.

METHODS

All the bone and joint specimens with giant cell-rich lesions received in the period of one year were studied along with clinical and radiological data available. Gross and microscopic findings were noted.

RESULTS

In a period of one year, 10 cases of giant cell-rich lesions of bone and joints have been studied, which were and correlated with clinical and radiological findings. Five were lesions from bone and two were from joints, which are chondroblastoma, chondromyxoid fibroma, osteoclastoma, aneurysmal bone cyst, pigmented villonodular synovitis, giant cell lesion of tendon sheath, and tendinous xanthoma.

CONCLUSION

In the present study, variety of giant cell lesions of bone and joints are studied. Of which, the mean age in young patients being 20 years and in elderly patients being 50 years. The common site being lower end of femur.

KEYWORDS

Giant cell-rich lesions, Bones, Joints.

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INTRODUCTION: Giant cell-rich lesions comprise a variety of entities seen in bones and joints. The diagnosis of giant cell lesions of bone is often a challenge for an experienced pathologist. The histological picture should be interpreted with x-ray and per operative findings. Differential diagnosis and proper diagnostic criteria should be kept in mind-to-differentiate between the various neoplastic and non-neoplastic giant cell-rich lesions affecting the bones and joints.

AIMS AND OBJECTIVES: To study the various giant cell-rich lesions affecting the bones and joints over a period of one year and to identify the age, sex, and anatomical site-wise distribution of giant cell-rich lesions of bones and joints and to correlate with radiological and histopathological findings.

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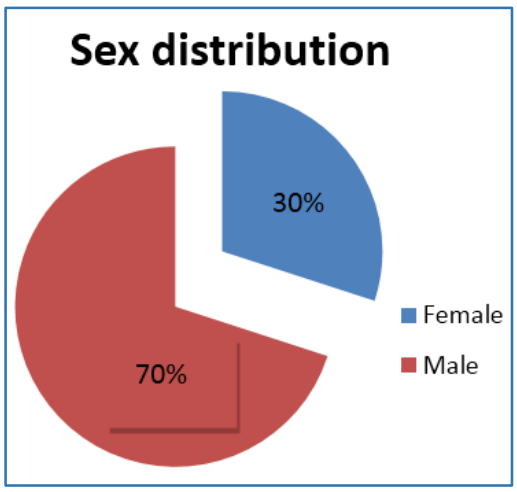
MATERIALS AND METHODS: The present study is conducted at the Department of Pathology, Maharajah's Institute of Medical Sciences from July 2014 to July 2015.

All patients with significant lesions in x-ray were selected. For histopathological examination, the tissues were formalin-fixed and paraffin-embedded and decalcification was done in bony tissues. Gross appearances of the specimens were properly studied along with the adequate clinical and radiological findings of the cases. Extensive grossing was done and sections were stained with routine H and E staining procedure. Special stains were used whenever required to aid in the diagnosis.

RESULTS: Ten cases of giant cell-rich lesions of bone and joints have been reported over a period of one year. Five were lesions from bone and two were from joints, which are chondroblastoma, chondromyxoid fibroma, osteoclastoma, aneurysmal bone cyst, pigmented villonodular synovitis, giant cell lesion of tendon sheath, and tendinous xanthoma.

Parameter	Age (years)
Mean Age	20
Youngest Age	5
Oldest Age	50

Table 1: Age Distribution



Site Distribution	Number of Cases	Histopathological Diagnosis
Lower end of femur	3	Chondroblastoma, Osteoclastoma, Aneurysmal Bone Cyst
Upper end tibia	1	Chondromyxoid Fibroma
Shaft of humerus	2	Aneurysmal Bone Cyst
Knee joint	2	Pigmentary Villonodular Synovitis
Distal phalanx	2	Tendinous Xanthoma, Giant Cell Lesion of Tendon Sheath

Table 2: Site Distribution

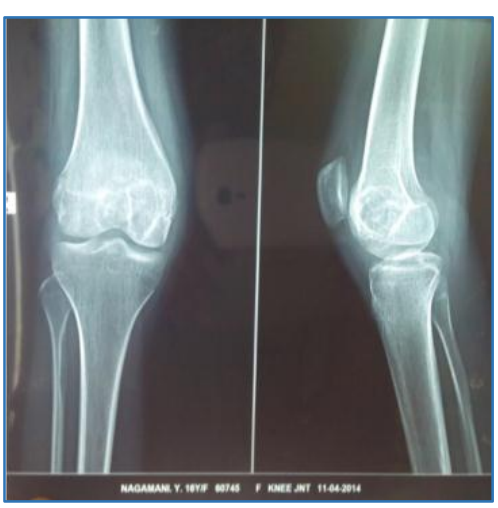


Fig. 1a: Chondroblastoma - X-Ray Picture Showing Lytic Lesion on Lower End of Femur

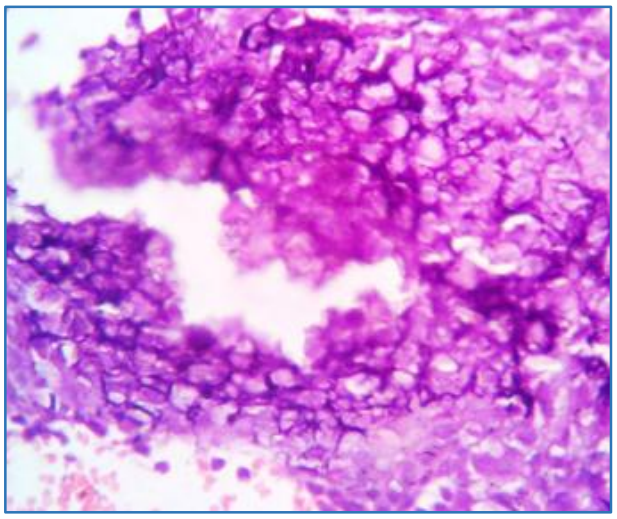


Fig. 1b: Chondroblastoma- Microscopic Picture Showing Typical Lace like Chicken Wire Calcifications. (H and E) 400x

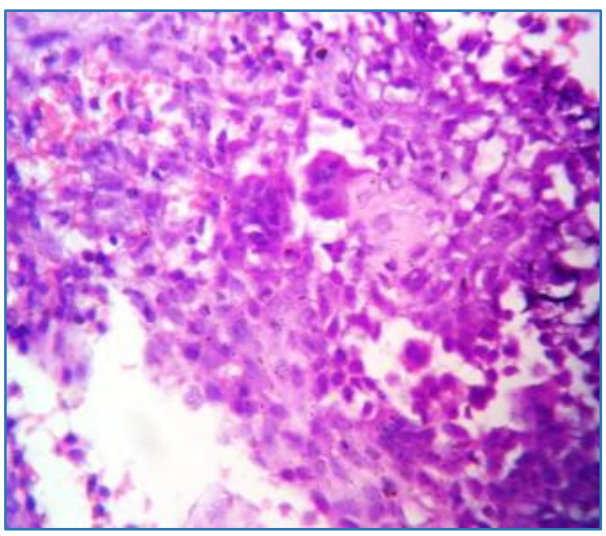


Fig. 1c: Chondroblastoma - Microscopic Picture Monotonous Population of Small Round Cells with Osteoclastic Giant Cells. (H and E 400x)

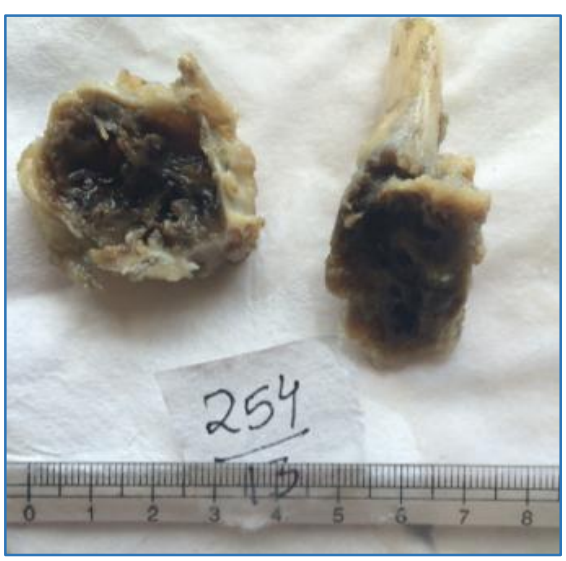


Fig. 2a: Osteoclastoma - Gross Photograph Showing Greyish Brown Haemorrhagic Areas with Thin Shell of Bone at the Periphery

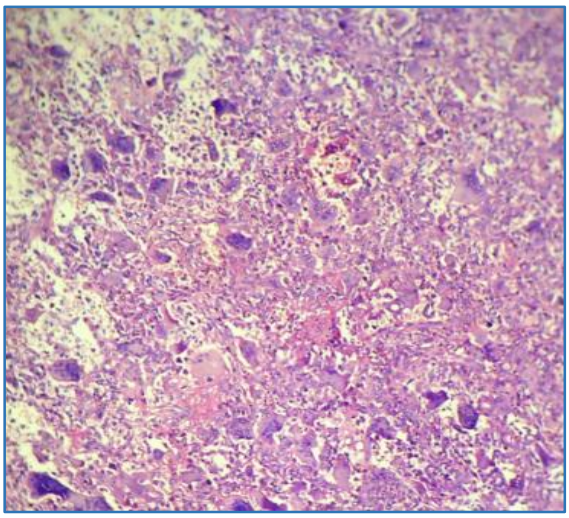


Fig. 2b: Osteoclastoma-Round to Oval Mononuclear Cells with Giant Cells. (H and E) 100x

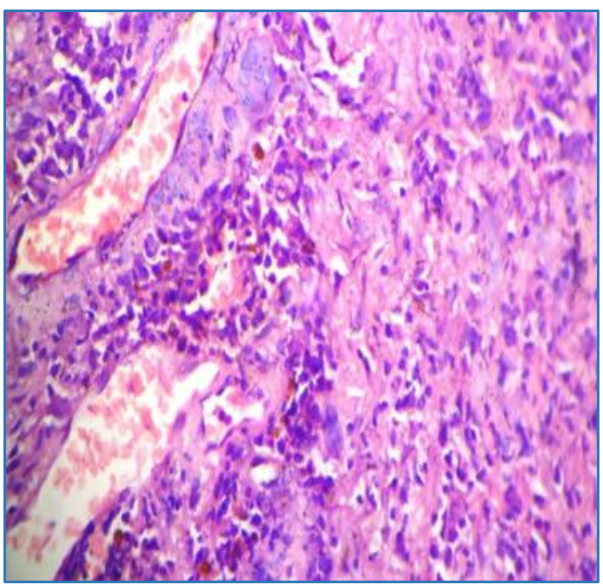


Fig. 3a: Aneurysmal Bone Cyst-Vascular Spaces with No Endothelial Lining and Giant Cells. (H and E) 400x

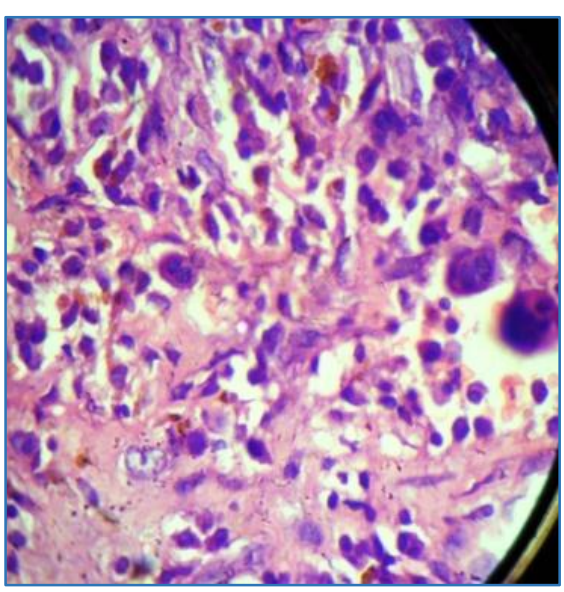


Fig. 3b: Aneurysmal Bone Cyst-Hemosiderin Pigment and Giant Cells. (H and E) 400x

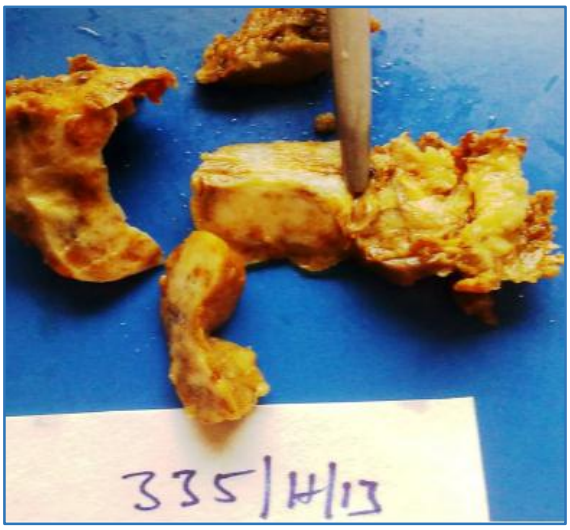


Fig. 4a: Pigmented Villonodular Synovitis-Orange Yellow Lesion with Villous-Like Projections of Synovium

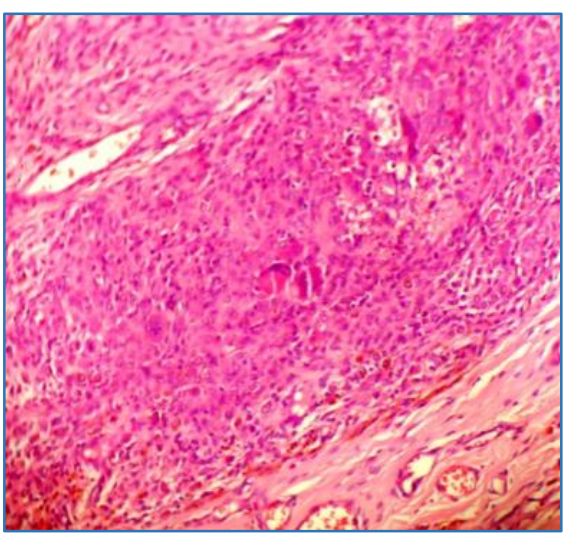


Fig. 4b: Pigmented Villonodular Synovitis - Small Round Cells, Giant Cells, and Hemosiderin Pigment. (H and E) 400x

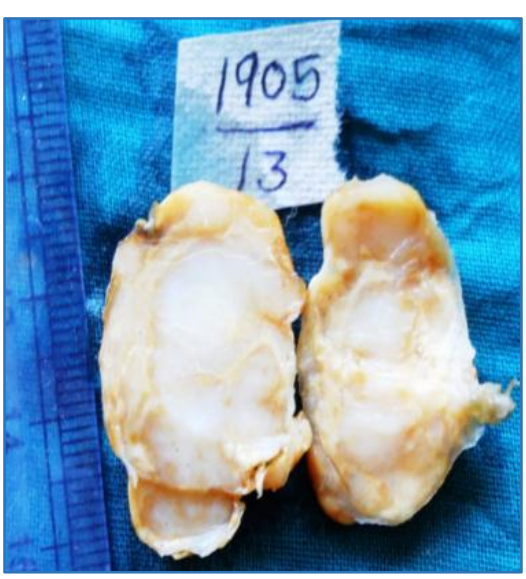


Fig. 5a: Giant Cell Tumour of Tendon Sheath-Firm and Lobulated Mass and Cut Surface Shows Yellowish White Areas

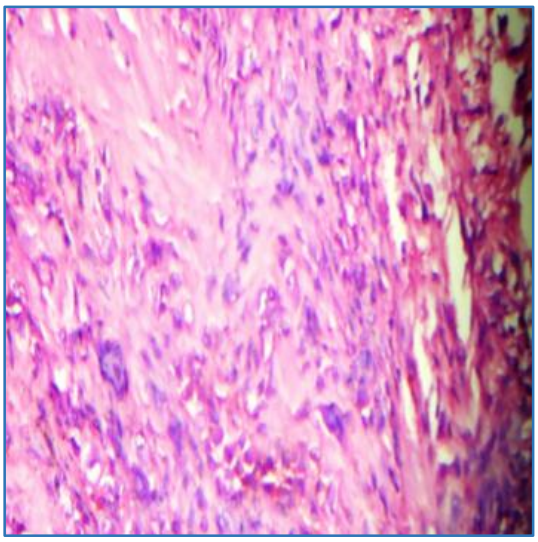


Fig. 5b: Giant Cell Tumour of Tendon Sheath: Areas of Hyalinization and Giant Cells. (H and E) 400x

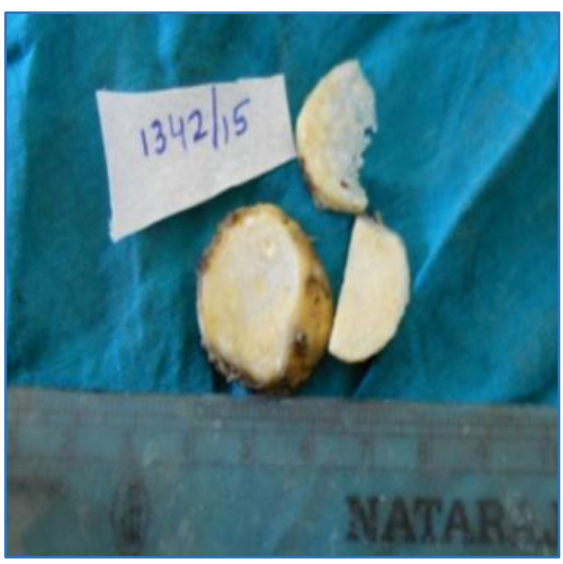


Fig. 6a: Tendinous Xanthoma: Gross: Well-Circumscribed Mass on Cut Section: Greyish White and Greyish Yellow Areas are Seen

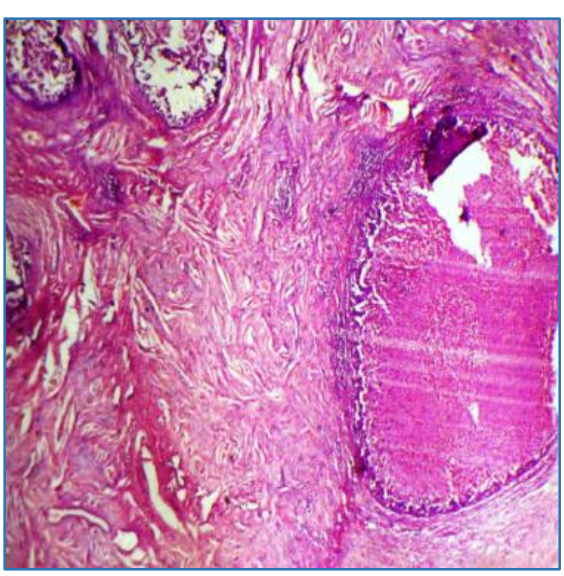


Fig. 6b: Tendinous Xanthoma: Note Plenty of Cholesterol Clefts and Some Cystic Spaces. (H and E) 40x

DISCUSSION: In general, the lesions with osteoclastic giant cells are:

- Chondromyxoid fibroma, Chondroblastoma, Chondrosarcoma.
- Osteoblastoma, Osteoid osteoma, Osteoclastoma, Telangiectatic osteosarcoma.
- Metaphyseal fibrous defect, Brown tumour of hyperparathyroidism, Giant cell reparative granuloma, Aneurysmal bone cyst, Simple bone cyst, Pigmented villonodular synovitis, Giant cell tumour of tendon sheath.

Chondroblastoma: It is a benign cartilaginous producing neoplasm usually arising from the epiphysis of skeletally immature patients particularly in the distal end of femur, proximal end of humerus, and proximal end of tibia.[1] It is mostly seen at the age of 10-20 years predominantly in males and can be quite painful. The present case was seen in a female of sixteen years who presented with pain and swelling of lower end of femur. The diagnostic microscopic features include pericellular calcifications.[2] (Chicken-wire calcification Fig 1b), which are network of thin lines (Chicken wire). Extremely cellular with round to polyhedral cells with round to indented and lobulated nuclei and scattered collections of giant cells. Radiologically, lytic lesions were seen in the lower end of femur (Fig 1a). The differential diagnoses are giant cell tumour, aneurysmal bone cyst, and chondromyxoid fibroma. Immunohistochemically, it is positive for S100, vimentin, muscle specific actin, low molecular weight keratin, neuron specific enolase. Recurrence is rare. Metastasis is to the lungs through lymphatics. Aggressive form is seen in pelvis.[3]

Chondromyxoid Fibroma: It is a rare benign lesion arising mostly in the metaphysis of long bones.[1] most common age group is between 20-30 years. Radiologically, it is sharply defined and of large size. Grossly, it is solid and yellowish white to tan, replaces bone, and thins the cortex. In the present case, it is seen in 21 years female who presented with painful swelling over upper end of tibia. Microscopically, it is characterised by hypocellular lobules with collagenous to myxoid matrix with stellate cells separated by intersecting bands of highly cellular tissue composed of fibroblast-like spindle cells and osteoclasts.[2] Differential diagnosis include fibromyxoma, chondroblastoma. It is positive for S100. Usually, it has a good prognosis, recurrence, and metastasis are rare.[3]

Osteoclastoma: It is a benign locally aggressive neoplasm, which is composed of sheets of neoplastic ovoid mononuclear cells interspersed with uniformly distributed large osteoclastic giant cells.[1] It is usually seen in patients over 20 years of age. The classic location is the epiphysis of a long bone from which it may spread into the metaphyseal area and break through the cortex. Radiologically, the typical appearance is that of an entirely lytic expansile lesion in the epiphysis without peripheral bone sclerosis or periosteal reaction.[2]

In the present case, it is seen in a 50 year male who presented with painful swelling over lower end of femur. Radiologically, there was a lytic lesion and grossly it showed greyish brown haemorrhagic areas with a thin rim of reactive bone at periphery. Microscopic examination revealed round to oval mononuclear cells with plenty of giant cells, which have more than 20-30 nuclei (Fig 2a and 2b). It is the prominence of these multinucleated giant cells that gives the tumour its name. Many evidences suggest that these multinucleate giant cells are not the neoplastic elements, but rather the result of fusion of circulating monocytes that have been recruited into the lesion. By contrast, the mononuclear stromal cell is to be interpreted as neoplastic. It is the only proliferating element in the lesion and the one exhibiting atypia in the rare cytologically malignant examples of this tumour.

These mononuclear cells are clearly mesenchymal in nature and share many features with normal mesenchymal stromal cells. Differential diagnosis includes all benign lesions with giant cells. Although, almost all neoplasms of bone may contain giant cells, giant cell tumours have a prominent proliferation of osteoclastic giant cells. One of the main microscopic differences between true giant cell tumour and other entities or variants containing giant cells is the spatial relationship between the giant and stromal cells. The former tend to be distributed regularly and uniformly in giant cell tumour whereas in the lesions that simulate it foci containing numerous clumped giant cells alternate with large areas completely lacking this component. The natural history of giant cell tumour is that of a low-grade malignancy.

Aneurysmal Bone Cyst: It is a benign cystic lesion of bone composed of blood filled spaces separated by connective tissue septa containing fibroblasts, osteoclast type giant cells, and reactive woven bone.[1] it is usually seen in patients between 10 and 20 years of age and slightly more common in females. It occurs mainly in the vertebrae and flat bones, but can also arise in the shaft of long bones. Multiple involvement is frequent in the vertebral lesions. Radiologically, aneurysmal bone cyst shows eccentric expansion of the bone with erosion and destruction of the cortex and a small peripheral areas of periosteal new bone formation.

Grossly, it forms a spongy haemorrhagic mass covered by a thin shell of reactive bone, which may extend into the soft tissue. Microscopically, large spaces filled with blood are seen. In the present study, two cases of aneurysmal bone cyst were noted. One case was a young female with swelling over left lower end of femur and another one was a 5 years male child with swelling over humerus. Grossly, lesion is spongy and has cystic spaces filled with haemorrhage. Histopathological picture revealed vascular spaces with no endothelial cell lining, giant cells, and hemosiderin pigment (Fig. 3a and 3b). Differential diagnosis are solitary bone cyst, giant cell tumour, haemangioma, telangiectatic osteosarcoma, and giant cell reparative granuloma. Secondary aneurysmal bone cyst is seen rarely.[4]

Pigmented Villonodular Synovitis: It is closely related to tenosynovial giant-cell tumour. Diffuse type of tenosynovial giant cell tumour is also known as extra-articular pigmented villonodular synovitis. It tends to occur in young adults. The most common site of involvement is knee joint. Occasionally, the lesion may penetrate within the underlying bone. The process may be focal or diffuse. When diffuse, it is made up of brownish-yellow spongy tissue. Its appearance depends on the content of hemosiderin pigment. In the present study, a 36 year male presented with a painful swelling over knee with limitation of movements. Grossly, lesion was orange yellow in appearance with villous-like projections of the synovium (Fig 4a). Microscopic findings include small round cells, giant cells, and hemosiderin pigment (Fig 4b). Differential diagnosis are degenerative joint diseases, rheumatoid synovitis, and foreign body reaction to materials. It has an excellent prognosis and usually the recurrence is very rare.[4,5,6]

Giant Cell Lesion of Tendon Sheath: It is also called tenosynovial giant cell tumour. It is a common lesion that occurs more frequently in women than in men usually in young and middle-aged persons. Most cases are located in-between the wrist and finger tips and in-between the ankle and toe tips. Vertebral column also can be affected. In the present study, a 42 year female presented with a painless solitary lesion with complaint of discomfort in movements of the fingers. Grossly, it was a solitary, firm, lobulated mass, which on cut section showed yellowish white areas (Fig 5a). Microscopic findings include areas of giant cells and hyalinization (Fig 5b). Differential diagnosis are fibroma of tendon sheath and pigmented villonodular synovitis.[4,6] It gives positive staining for desmin and CD68. The multinucleated giant cells have the phenotypic features of osteoclasts. The great cellularity of this tumour, its variable pattern, and the presence of mitotic figures may lead to an erroneous diagnosis of sarcoma. However, these tumours are nearly always benign. They may erode contiguous bone by pressure. If incompletely removed, they may recur locally.

Tendinous Xanthoma: Tendon xanthomas are associated with type II hyperlipidemia.[5,7] 43 years male presented with painless swelling over ring finger. Grossly, it is a well circumscribed mass, which on cut section showed greyish white and greyish yellow areas (Fig 6a). Histopathology revealed plenty of cholesterol clefts, few cystic spaces, giant cells, and areas of hyalinisation (Fig 6b). It is also called as giant cell lesion of tendon sheath-Localised type. Immunohistochemically, it is positive for CD68, muscle specific actin and desmin. It has a good prognosis and the recurrence is 40%.[4,6]

CONCLUSION: Different entities of giant cell-rich lesions of bones and joints were studied in the present study. The mean age of presentation in young patients being 20 years and in elderly patients being 50 years. The common site being lower end of femur.

Since many differential diagnoses come under the umbrella of giant cell lesions, clinical, radiological, and pathological correlation is essential for accurate diagnosis of these lesions. One of the main microscopic differences between true giant cell tumour and these so-called variants or giant cell-rich lesions, resides in the spatial relationship between the giant and stromal cells. The former tend to be distributed regularly and uniformly in giant cell tumour whereas in the lesions that simulate it, foci containing numerous, clumped giant cells alternate with large areas completely lacking this component. The giant cells themselves do not differ significantly in the two groups of disease, morphologically, histochemically, or immunohistochemically.

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