Review Article

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Chondrosarcoma-A Multi-Faceted Entity

Sujata Patnaik[1]*, Shantiveer Uppin[2] and Y Jyostnarani[3]

[1]*Additional Professor, Department of Radiology and Imageology, Nizam's Institute of Medical Sciences, Hyderabad, India*

[2]*Professor of Pathology, Department of Pathology, Nizam's Institute of Medical Sciences, Hyderabad, India*

[3]*Professor of Radiology, Department of Radiology and Imageology, Nizam's Institute of Medical Sciences, Hyderabad, India*

***Corresponding author**

Sujata Patnaik, Additional Professor, Department of Radiology and Imageology, Nizam's Institute of Medical Sciences, Hyderabad, India

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Introduction

Chondrosarcoma (CS) accounts for 20-27% of all primary malignant tumors of bone mostly seen in older age group [1]. It may be primary or secondary occurring in 2nd to 5th decade. The long bones especially the metaphysis are the most common sites. Axial skeleton, skull, jaw, small bones of hand and feet are other sites that can be involved. Though rare, extraskeletal CS may occur anywhere in the body. The conventional CS are primary or secondary. Primary chondrosarcoma may be intramedullary or Juxtacortical. Secondary CS arises from Osteochondroma or Enchondroma. Non-conventional CS are clear cell, myxoid, mesenchymal, extraskeletal, and dedifferentiated. Usual distribution of these tumors is long bones (45%) or pelvis (25%). The spine (7%), scapula (5%), cervical spine (6-7%), craniofacial bones (2%), hand and foot are rarely affected [2].

Our Observation

37 cases of HPE proven chondrosarcomas were analysed. Maximum were in age group of 40-60 yrs. M: F ratio was 25:12. Long bone involvement was seen in 16 cases. Skull (n=3), jaw (n=1), vertebrae (n=1), sacrum (n=3), ribs (n=3), pelvis (n= 6) were other sites. There was 1 case of synchronous CS. Our study also included extraskeletal soft tissue CS (n=2) and periosteal chondrosarcoma (n=1). There were 3 cases having metastases in lung. Three had dedifferentiated component.

Discussion

Pathologically these lesions are multi-lobulated due to hyaline cartilaginous nodules with central high water content and peripheral endochondral calcification. High water content appears as high signal on T2W-images and calcification appears ring and arc shaped on radiograph as well as on CT scan. According to pathology they can be graded into 4 types. Histologically CS are classified as clear cell type, well differentiated and dedifferentiated high grade which may be having osteoid component/fibroblastic/malignant fibrous histiocytoma.

Conventional Primary Chondroblastomas

These are hyaline cartilage tumors. Hence growth pattern is lobular deep, endosteal scalloping, focal area of deep penetration and associated soft tissue component. Majority present with osteolytic lesion metadiaphysis with cortical destruction in 61% cases. Size is more than 4cms occupying more than 30% -50% of bone length (Figure 1A, Figure 1B and 2). Cortical thickening endosteal scalloping are other findings. Pathological fracture may be seen (Figure 3). Periosteal reaction may be unilamellar or malignant type appearing spiculated, multi-layered in 6% cases. Matrix mineralisation seen in 77% in radiographs and 82% on CT scan (Figure 4) and 66% show chondroid mineralisation on MR appearing as high signal lobules, hypointense foci in 20% onT2W observed representing medullary calcification (Figure 4B). Soft tissue component is appreciated on radiograph 25%, CT- 81% and MR -80% (Figure 5). On contrast the mass shows enhancement.

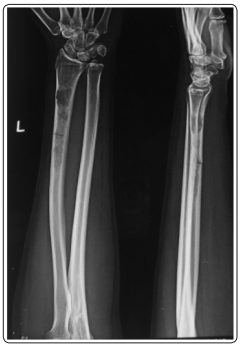

Figure 1A: 42, M: Chondrosarcoma-the length of involvement is more than 30% of bone length

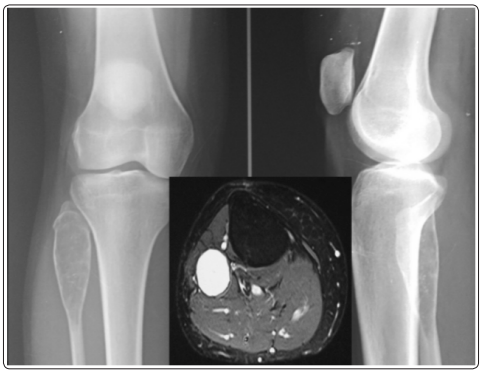

Figure 1B: 25, M: Cortical scalloping, thinning of cortex more than 2/3rds thickness in low grade Chondrosarcoma

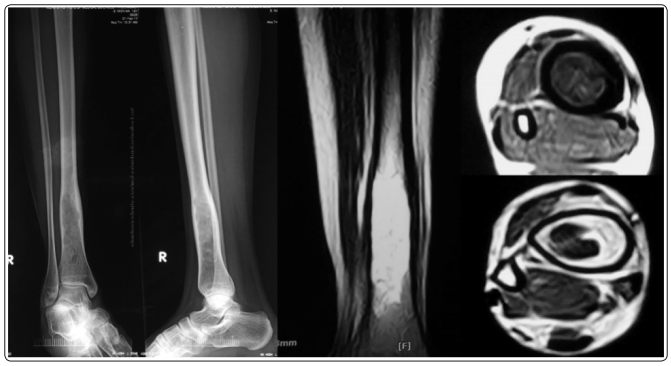

Figure 2: 19, F: Chondrosarcoma of low-grade. Length of lesion > 30% of bone length, endosteal Scalloping, cortical thickening and matrix calcification can be seen

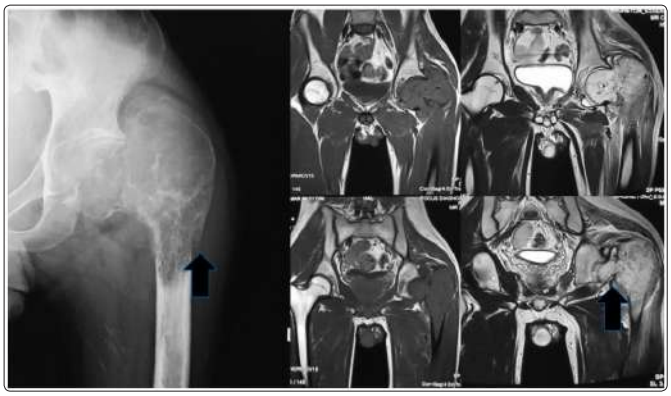

Figure 3: CS with pathological fracture

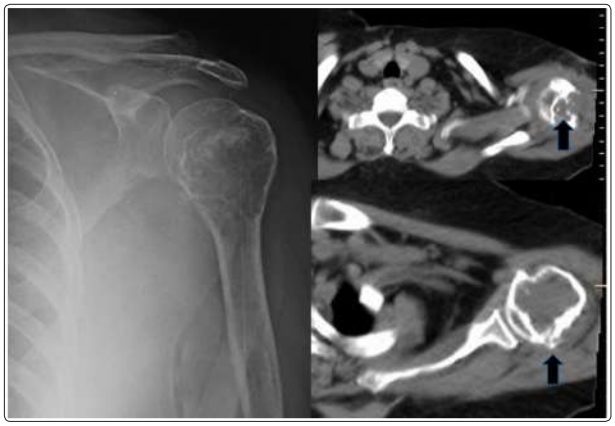

Figure 4A: 60, F: Cortical disruption and matrix calcification on CT scan

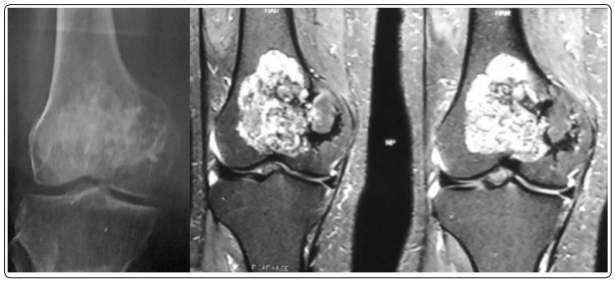

Figure 4B: 55, F: Chondrosarcoma-Matrix calcification appearing as hypo-intense foci in STIR W MRI; high-signal signify Chondroid matrix

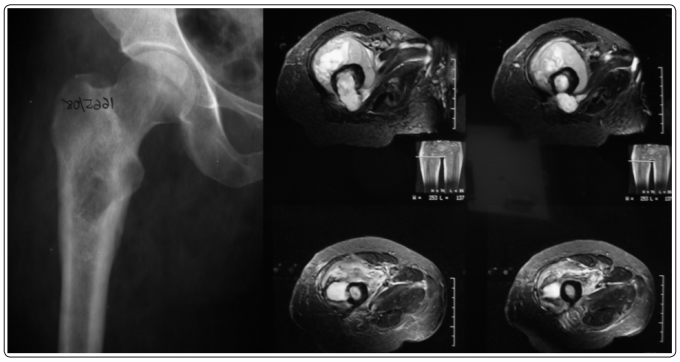

Figure 5: 31, F: Large soft-tissue component is well appreciated in MRI and not on radiograph

Secondary Chondrosarcomas

1% of solitary osteochondroma and 3-5% of multiple exostoses undergo malignant transformation. Increasing pain, swelling, increasing size after skeletal maturation raises malignant change. Thick cartilage cap more than 2cms, development of soft tissue mass with or without calcification or dispersed calcifications, bone destruction indicates malignant transformation of Osteochondroma (Figure 6 and Figure 7). Similarly, when the size of enchondroma is more than 5-6cms, cortical breach or permeative/moth eaten bone destruction are noted there is suspicion of malignant change. Soft tissue component is also another feature of malignant transformation (Figure 1 and Figure 2). Small tubular bones of hand and feet are uncommon sites for CS whereas lesions in pelvis, spine, and ribs are common sites. Malignant transformation in primary synovial chondromatosis though rare, has been reported. Sarcomatous change is considered if there is rapid growth /recurrence after synovectomy. Radiologic distinction is not possible.

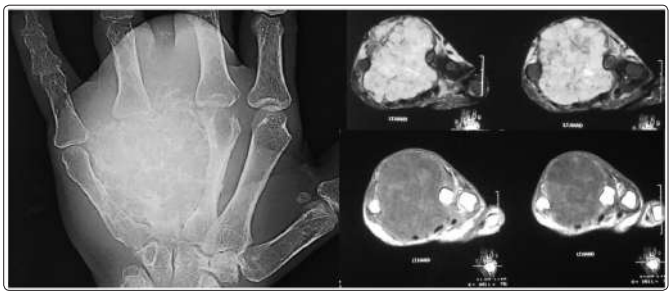

Figure 6: 40, M: Secondary Chondrosarcoma- Destruction of 4th metacarpal-Lobulated; T2 W High signal lobule of cartilaginous nature

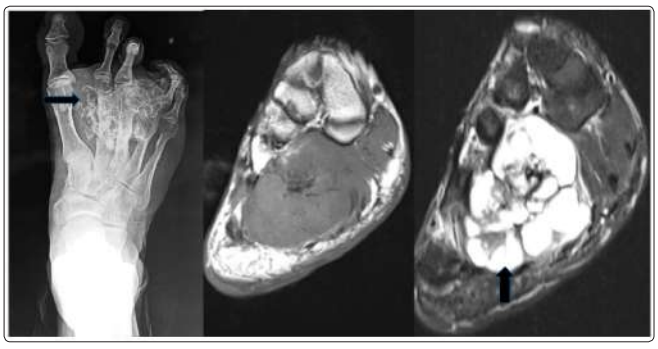

Figure 7: 36, M: Exostosis turned into Chondrosarcoma: Destruction of cartilagenous outline; large cartilagenous cap

Juxtacortical Chondrosarcomas

Periosteal Chondrosarcomas

It arises from outer surface with radiological features similar to conventional CS. It may be low/moderate/severe grade occurring 3rd to 4th decade. This tumor accounts for 2% of all CS and 0.2% of all bone tumors [3]. These arise from surface of bone, lifting the periosteum over them with fibrous pseudo capsule. Underlying cortex is thickened/may be eroded; however extension to medullary cavity is uncommon. Codman triangle and matrix calcification may be observed. Lesion is covered by pseudocapsule which is continuous with periosteum. Peripheral and septal enhancement is noted in CT and MRI (Figure 8). Differentials are periosteal osteosarcoma and juxtacortical chondroma. Periosteal OS occurs in younger age 20-30 years and are chondroblastic. Mid-diaphysis is involved. Periosteal chondroma has similar appearance as periosteal chondrosarcoma but these lesions are smaller and less than 3cms. Soft tissue invasion is noted in periosteal CS.

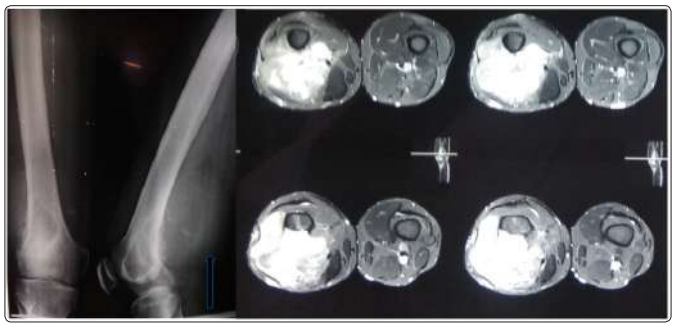

Figure 8: 39, M: Periosteal chondrosarcoma: Note large soft tissue with calcification, cortex eroded; No medullary involvement

Chondrosarcoma at various site have some peculiar features, CS of pelvis has predilection for triradiate cartilage- acetabulum. Usually the size is large at presentation. Radiographically lesions are underestimated and appear small. Any chondromatous lesion in rib is CS as it is highly potential site. They usually occur at costochondral junction. Similar occurrence is possible in sternum (Figure 9, 10). In contrast, hand or foot lesions are regarded as benign as these are common sites for enchondroma .If sarcomatous change occurs these are difficult to differentiate. In spine posterior elements are common site. 40% occur in posterior elements, 15% in vertebral body and 40% in both (Figure 11). These lesions can cross the intervertebral disc and to ribs (Figure 11) [4].

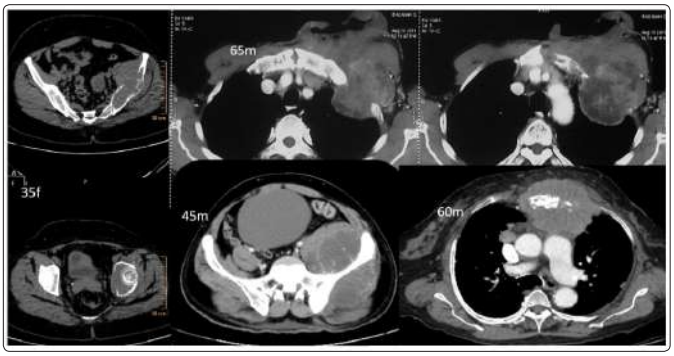

Figure 9: Cases of chondrosarcoma with lesions in pelvis, rib, sternum; large soft tissue component showing high water content, calcifications and heterogeneous enhancement

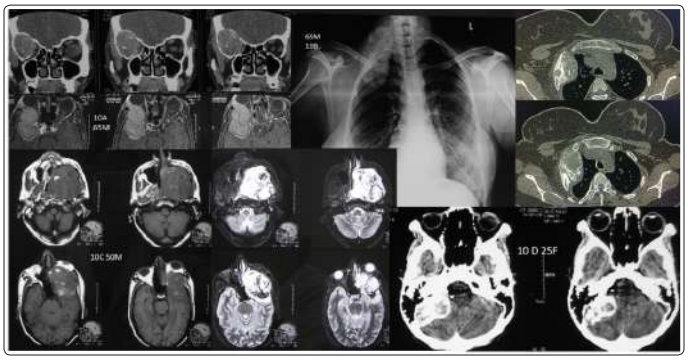

Figure 10: Chondrosarcoma of sphenoid, rib, maxilla and petrous bones

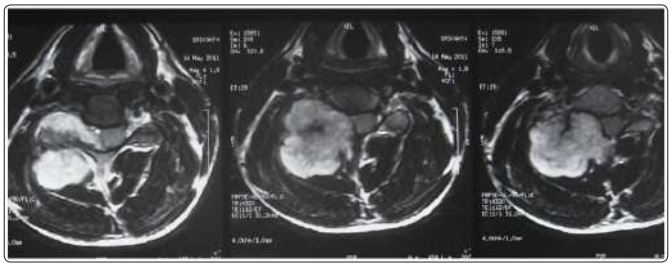

Figure 11: 20, M: Chondrosarcoma of Spine with typical involvement of posterior element

Craniofacial CS accounts for 2% of all CS. Maxilla is a common site. These have to be differentiated from chondroma which occurs a decade later than CS and grows rapidly [5].

Clear Cell Type Chondrosarcomas

Clear cell type is rare accounting approximately 1-2% of all chondrosarcoma and 0.2% of all bone tumors. These tumors are low grade malignancy, slow growing occurring 3rd to 4th decade with male predominance [6]. They have better prognosis and radiologically predominant lytic lesion occurring in Femur, Humerus in proximal end epiphysis. Sometime there is large area of haemorrhage/cystic changes. Mild expansion of bone and peripheral sclerosis may be observed. CT demonstrates soft tissue component and calcification. It is to be differentiated from Chondroblastoma (Figure12 and Figure 13).

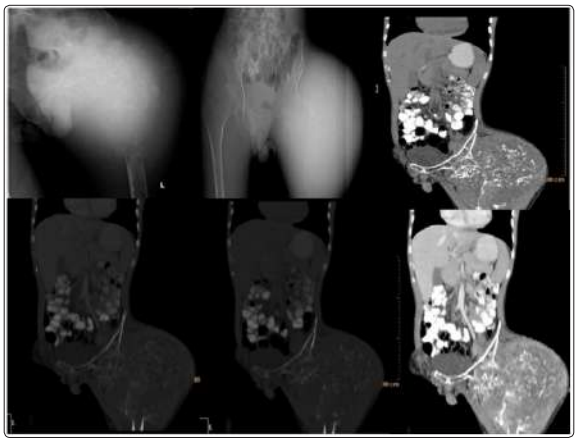

Figure 12: 21, M: Epiphyseal involvement with a calcific foci in a case of Clear cell Chondrosarcoma

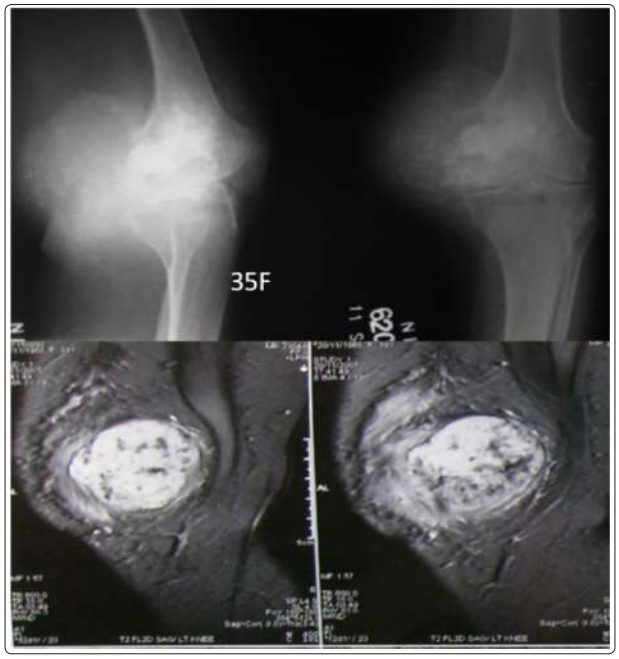

Figure 13: Clear-cell Chondrosarcoma mimicking Chondroblastoma

Myxoid Chondrosarcomas

It accounts for 2% of CS occurring at 50yrs of age. They can occur in bone and soft tissue. Half of the cases are seen in femur. Osseous mass with soft tissue component. Matrix mineralisation noted on CT. On CT and MR, these lesions enhances mildly. These are of intermediate grade malignancy. In soft tissue they occur in extremity, deeper tissue is usually involved. Only 25% occur in subcutaneous tissue. Course is aggressive; metastases and local recurrence are common (Figure 14) [7].

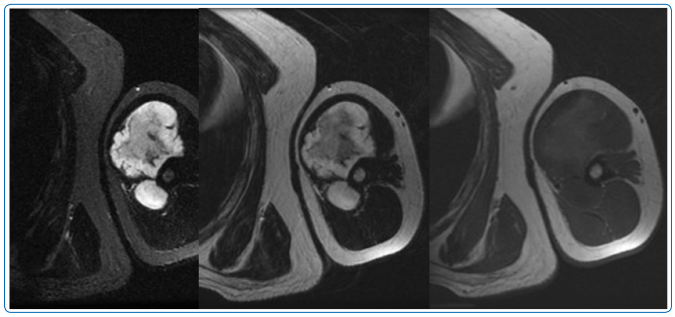

Figure 14: Extra skeletal Chondrosarcoma: Myxoid type in deep soft tissue of upper extremity

Mesenchymal Chondrosarcomas

2% CS are Mesenchymal CS being very aggressive high grade with tendency to metastasise [8]. Common sites are axial skeleton, ribs, jaw, long bone mostly diaphysis. Aggressive bone destruction either permeative/moth eaten appearance noted in radiographs. Lesions are associated with ill-defined multi-layered periosteal reactions. Matrix has rings or arc like calcifications. CT scan reveals all these features more clearly. Chondroid mineralisation is best depicted. On MR the mass has low to intermediate signal both on T1W, T2W images with homogenous/heterogenous enhancement. Main differentials are small round cell tumors like EWS.

Low-Grade Chondrosarcomas

These tumors are lytic sclerotic with size more than 4cms usually more than 10cms. Lesions grows in lobular pattern with endosteal scalloping with or without cortical penetration. Ring/arc like matrix mineralisation seen. DD from enchondroma is by destruction of trabeculae, more than 2/3rd cortical scalloping, and soft tissue extension (Figure1 and Figure 2).

Dedifferentiated Chondrosarcomas

10% of conventional CS undergo dedifferentiation in the 5th to 7th decade [9]. Pathological fracture and large soft tissue component may be seen. Imaging features depend on proportion of conventional CS and non-cartilageneous focus increasing size. There will be rapid increase in size, progression of aggressive lysis, decrease calcification and soft tissue expansion. Hence presence of large non-mineralized soft tissue mass predicts dedifferentiation in cartilaginous tumor helping the diagnosis and choosing the site for biopsy. CT and MR show distinct area of low and high grade components. Septal peripheral enhancement in low grade component and diffuse enhancement in high grade component is also another feature of differentiation. Sometimes there may be nodular enhancement instead of peripheral enhancement. Dedifferentiated components suggest 'high grade' either osteoblastic/fibroblastic/malignant fibrous histiocytoma (Figure 15). Other rare components may be rhabdomyosarcoma, angiosarcoma, and leiomyosarcoma. The cartilaginous and non- cartilaginous components are seen adjacent to one another and term them as collision of two tumors [2].

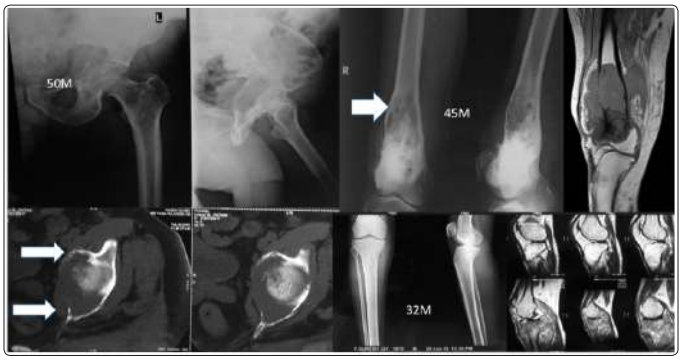

Figure 15: Three different cases of conventional chondrosarcoma turned to De-differentiated chondrosarcoma; Arrow points to cartilaginous and non-cartilaginous dedifferentiated component

Extraskeletal Chondrosarcoma

Extraskeletal chondrosarcoma are highly aggressive occurring in extremity predominantly. Other sites are thigh, meninges, orbit and brain [10]. They are either myxoid/mesenchymal in type (Figure 14) [2]. Non- specific findings on CT include heterogenous tumor in soft tissue with HU value of 20-40 slightly lower than muscle showing minimal enhancement. CT may demonstrate ill-defined cloud like matrix with calcified whorls and arc (Figure 16).

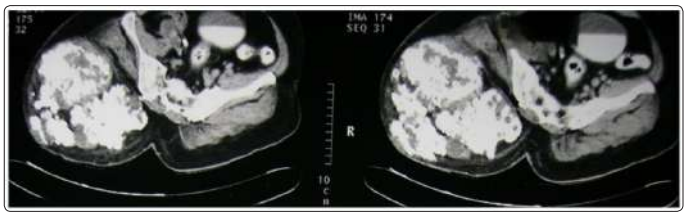

Figure 16: Extra-skeletal gluteal chondrosarcoma

Mutifocal Chondrosarcomas

Multicentric CS are rare may be monomelic/ multi centric (Figure 17). It may be mesenchymal. Only a few cases are reported in literature. Multiple hereditary exostoses may have increased chance of sarcomatous changes in 5-25%.

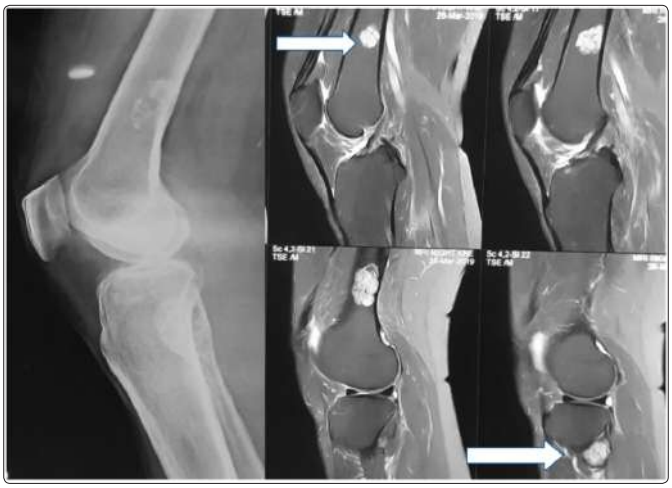

Figure 17: 60, F: Multifocal chondrosarcoma (in femur and fibula on right): Low grade

Conclusions

Most of chondrosarcomas are low grade conventional type. Mesenchymal, myxoid and dedifferentiated chondrosarcoma variants are regarded as high grade. Clear cell and extraosseous chondrosarcoma though rare, may be observed at times. Hence one should be aware of their imaging features. Aggressive bone destruction like moth eaten appearance, permeative pattern, non-mineralised matrix, homogenous /nodular enhancement suggest high grade tumor. Tumor biphasicity is determined by identifying cartilaginous and non cartilageneous component.

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