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REVIEW ARTICLE

Benign Cartilaginous Lesions

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Introduction

Majority of the cartilaginous lesions occur in bone. However, many have been described in soft tissues including larynx, synovium etc. Benign cartilaginous tumors constitute about 50% of all bone tumors. These are described in Table 1

Table 1: Benign cartilaginous tumors

- Enchondroma – single / multiple
- Ecchondroma
- Osteochondroma
- Post radiation and post traumatic osteochondroma
- Multiple osteochondromata
- Paraosteal chondroma
- Periosteal chondroma
- Epiphyseal chondroma
- Chondroblastoma
- Chondromyxoid fibroma
- Soft tissue chondroma
- Miscellaneous – focal, fibrochondral dysplasia,synovial chondromatosis,
- Ollier's dyschondroplasia

Radiological investigations include the following Table 2.

Table 2:

- Conventional
- Ultrasonography
- MDCT

- Pet CT
- Radionuclide scanning
- MRI

All these investigations may not be necessary in all patients. The general principles applied in the diagnosis of bone tumors are given in Table 3.

Table 3:

- Age and sex
- Single or multiple Lesions
- Type of bone involved
- Site of the lesion within the bone
- Site of origin of the lesion
- Nature and extent of bone change
- Appearance of the transitional zone
- Nature of the tumour matrix
- Type of periosteal reaction
- Soft tissue involvement

Diagnosis of chondroid tumours mainly rests on conventional radiology and is made by the anatomical location, transitional zone and mineralization of matrix. The calcified matrix may be minimal or heavy. When there is no mineralization of cortex, it is difficult to make a firm diagnosis. In these instances CT or MRI may be helpful. Endosteal scalloping, thick periosteal reaction and cortical hook when present may add to the correct diagnosis.

Polyostotic

- Multiple enchondromata
- Metachondromatosis (Hereditary)
- Ollier's dyschondrodysplasia
- Maffuci's syndrome
- Multiple osteochondromata

Enchondroma

The incidence is 3-17% of biopsied primary bone tumors. This is the second most common cartilage-containing tumor. It is due to continued growth of residual benign rests of cartilage displaced from the growth plate. It is generally asymptomatic except when there is trauma that result in a pathological fracture (Figure 1).

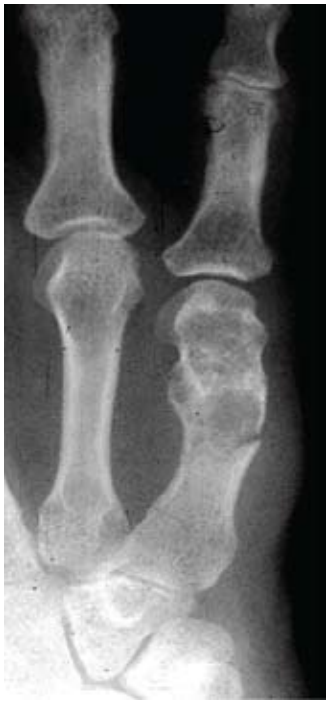

Figure 1: Enchondroma of fifth metacarpal with pathological fracture

It commonly presents in subjects between 10 – 30 yrs of age. When multiple these are located in tubular bones of hands (Figure 2).

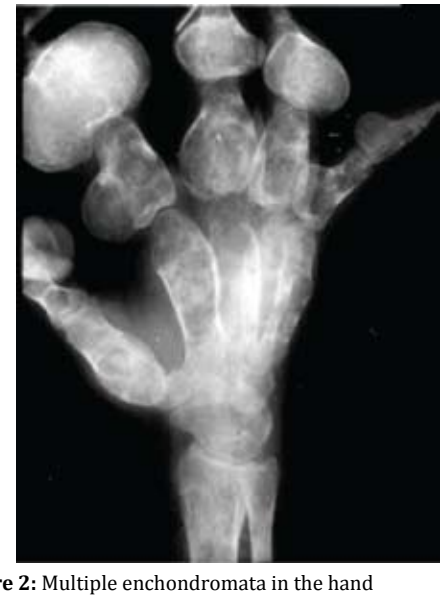

Figure 2: Multiple enchondromata in the hand

Multiple enchondromata in the hands is also called Ollier's syndrome. Other sites affected include long bones including femora, humeri, clavicles and ribs (Figure 3abc).

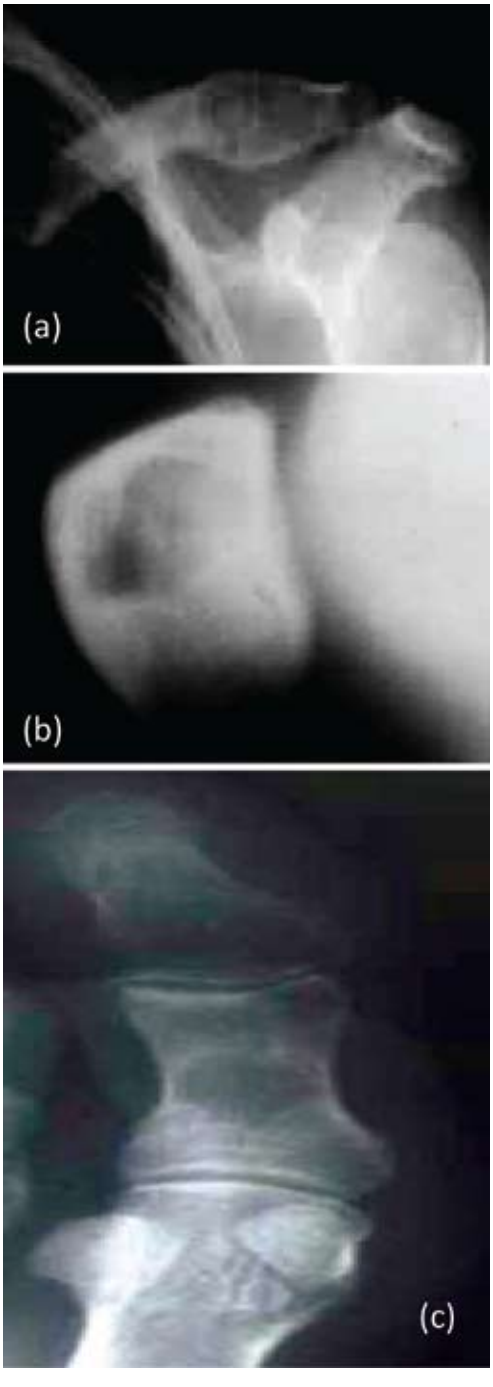

Figure 3: Enchondroma in (a) Clavicle (b) Patella (c) Big toe

Enchondroma is located in the central medullar canal of the metaphysis. The incidence is 3-17% of biopsied primary bone tumors and it is the second most common cartilage containing tumor. The average size is smaller than 3cm. When it exceeds 5cm low grade malignancy should be considered. It is due to continued growth of residual benign rests of cartilage displaced from the growth plate. The calcifications of the matrix may assume the shape of punctuate, arc like, circular, nodular, or a mixture of these (Figure 4ab). CT shows early mineralization much better than conventional radiography.

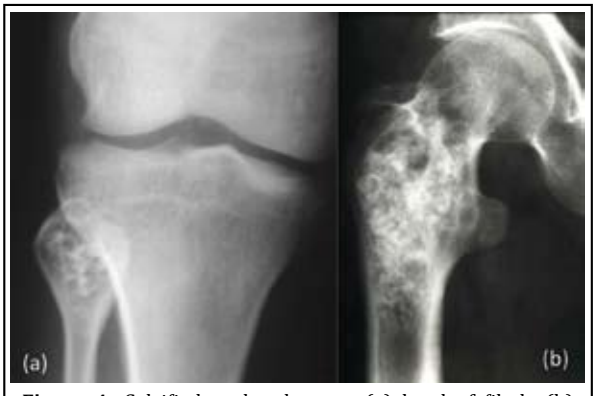

Figure 4: Calcified enchondromata (a) head of fibula (b) intertrochanteric area of femur

Another finding in cartilaginous tumors is the presence of a hook at the margins (Figure 5).

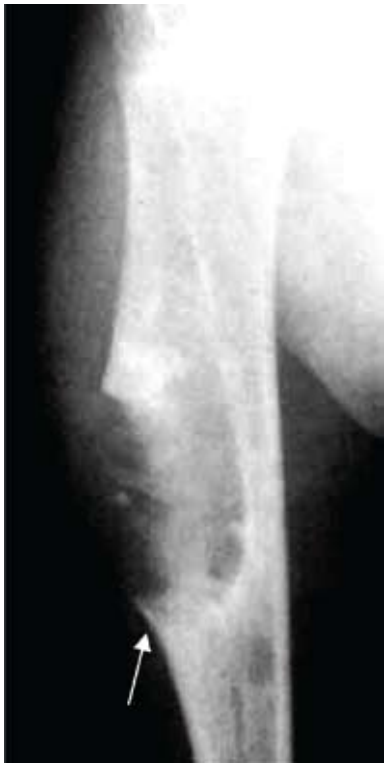

Figure 5: Intracortical chondroma showing calcifications and the presence of hook inferiorly

Periosteal chondroma is also called juxta cortical chondroma and is diagnosed by the anatomical location and the presence of calcific matrix (Figure 6ab).

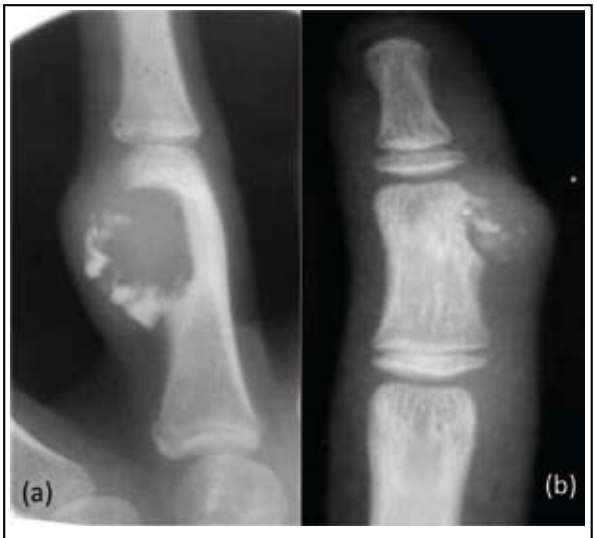

Figure 6ab: Periosteal chondroma in the phalanges with calcifications extending into the soft tissues

The complications of enchondroma include pathological fracture and malignant degeneration particularly when located in the long-bones whose incidence is between 15 and 20%. In the differential diagnosis, epidermoid inclusion cyst (phalangeal tuft), with history of trauma, unicameral bone cyst (rare in hands, more radiolucent), giant cell tumor of tendon sheath (commonly erodes bone, soft-tissue mass outside bone) fibrous dysplasia (mostly polyostotic) and bone infarct (Figure 7ab) need to be continued.

Figure 7ab: AP and lateral views of lower end of femur showing bone infarct. Calcification simulates enchondroma

Maffucci syndrome constitutes multiple enchondromata and soft tissue hemangiomas. It is actually a non-hereditary mesodermal dysplasia. In this connection, an increased prevalence of malignancies in the bone, ovary, duodenum and central nervous system. The diagnosis of Maffucci syndrome is made by the presence of enchondromata and presence of phleboliths in the soft tissue hemangiomas (Figure 8).

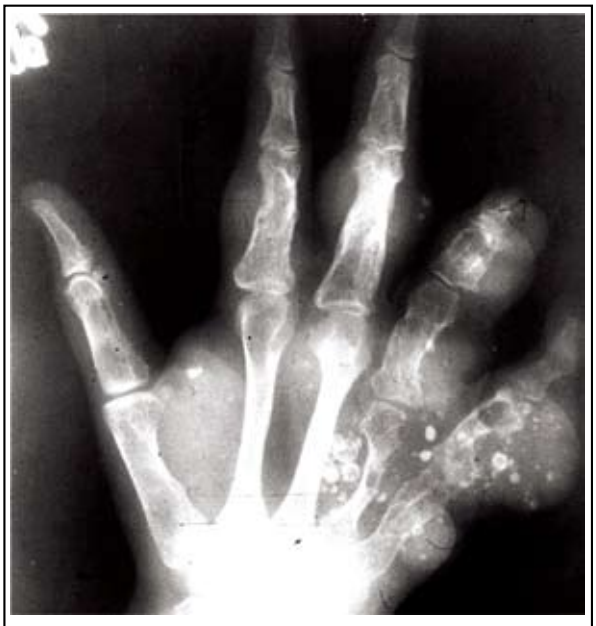

Figure 8: Maffuci syndrome. Note the enchondromata and calcified phleboliths in hemangiomata

Subcutaneous soft tissue chondromas are encountered without the involvement of underlying bones. They show amorphous, lumpy and homogenous calcifications (Figure 9ab). In occasional, instance calcified soft tissue hematoma and localized Myositis ossificans may be considered in the differential diagnosis.

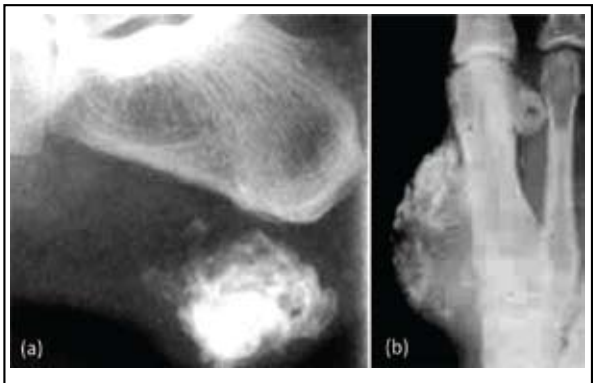

Figure 9: (a) heel (b) hand – soft tissue enchondromata

They may be seen in intra and extra articular locations (Figure 10ab). They may also occur in structures like nasal septum, larynx etc.

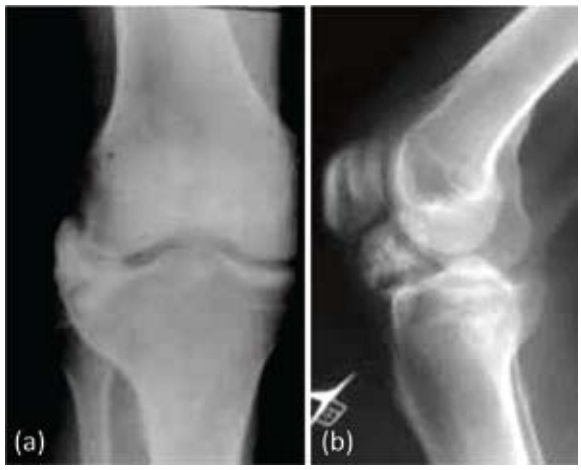

Figure 10ab: Intra articular chondroma of the knee

Ollier's dyschondroplasia is a developmental disorder. Chondromatous lesions are encountered in several bones and usually affect children in the age group of 1 to 12 yrs. These lesions are generally distributed on one side of the body. These may lead to shortening of the limb (s). Radiological findings include cortically oriented lesions with calcifications. The hook sign indicates that it is a cartilaginous lesion. This lesion prevents normal growth of the bone and may lead to shortening of the bone (Figure 11ab).

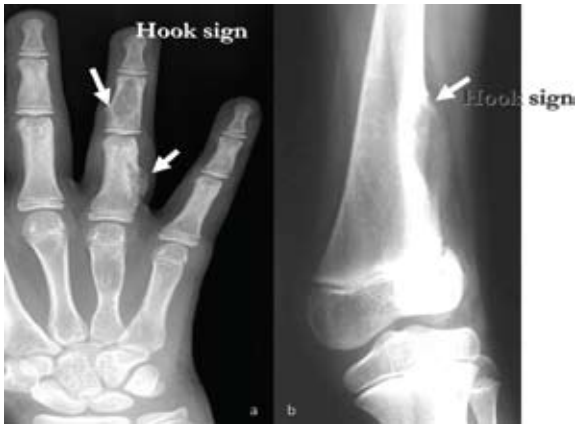

Figure 11: Ollier's dyschondroplasia (a) hand (b) femur. Note the hook sign as well as the enchondroma. Shortening of femur is noted

Chondroblastoma is a benign cartilage tumor and constitutes 1-3% of all bone tumors. Male and Female ratio is 2:1 and is encountered in 2nd & 3rd decades. Long & short tubular bones are frequently affected. Radiologically a lytic area is noted in the epiphysis or metaphysis, or the lesion may involve both epiphysis and metaphysis. Chondroid calcification is present in 1/3rd of cases (Figure 12ab).

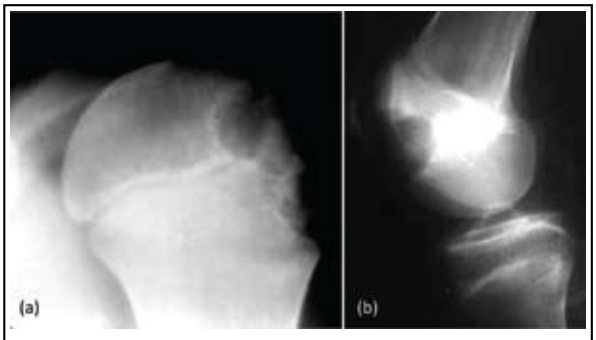

Figure 12: Chondroblastoma in (a) humeral head (b) lower end of femur

When it is primarily located in the metaphysis periosteal reaction may be noted (Figure 13).

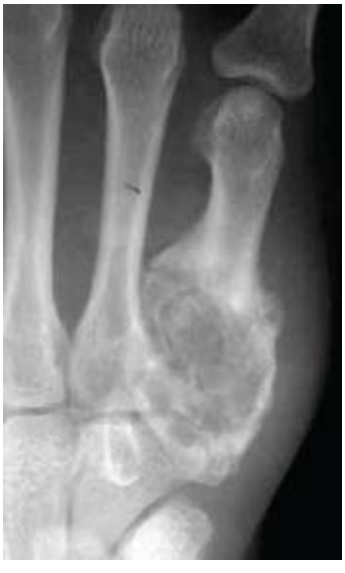

Figure 13: Chondroblastoma with periosteal reaction at the base of the fifth metacarpal. Note the pathological fracture.

In about 1/3rd of the cases aneurysmal bone cyst may be associated in which instance, MRI shows fluid – fluid levels (Figure 14ab).

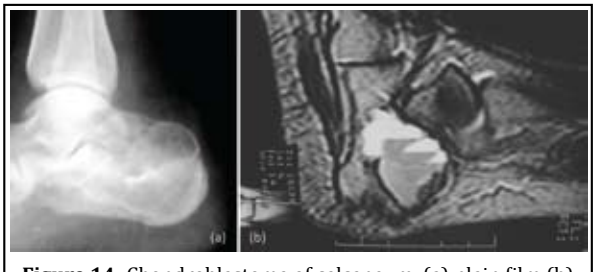

Figure 14: Chondroblastoma of calcaneum. (a) plain film (b) MRI showing fluid – fluid levels indicating secondary ABC

Chondromyxoid fibroma is a rare benign cartilaginous tumor although it is also included in fibrogenic

tumors. It is encountered in 2nd and 3rd decades. It occurs in the metaphysis of long tubular bones as well as short bones of the feet. Radiologically, it is a lytic expanding lesion with lobulated appearance, tending to be eccentric. Trabeculations may be seen with endosteal sclerosis (Figure 15ab).

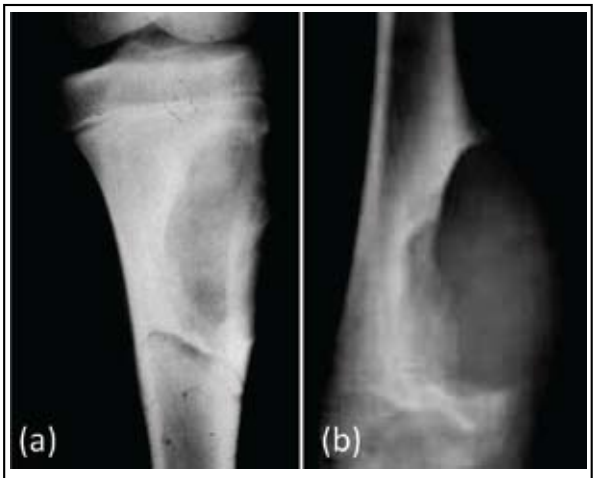

Figure 15: Chondromyxoid fibroma of (a) proximal tibia (b) distal tibia

This is one of the benign aggressive lesions simulating radiologically an aneurysmal bone cyst. Calcification is rarely noted, although punctate calcifications are noted histologically. Advanced imaging does not really help in the diagnosis.

Solitary osteochondroma (exostosis) is generally encountered in the first four decades. When a single lesion is noted it is appropriate that the whole body is investigated for detecting multiple lesions. Radiologically, it occurs in the metaphysis and

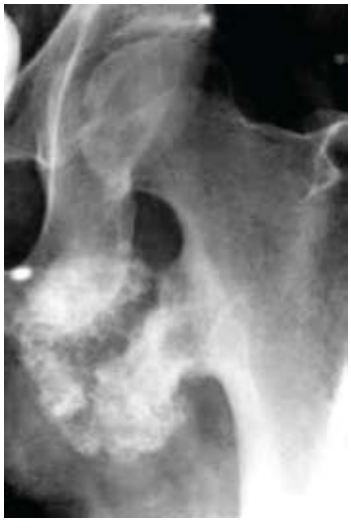

Figure 16a: Exostosis of lesser trochanter of femur

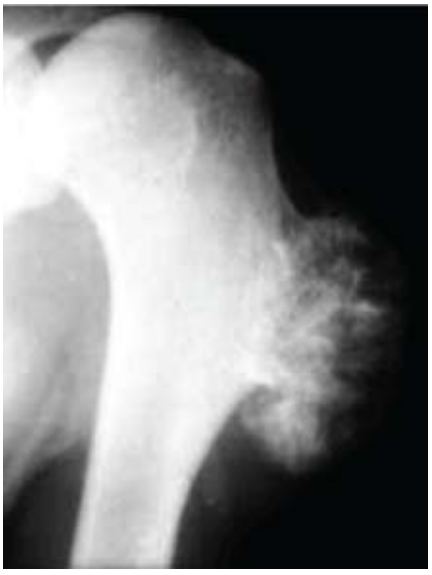

Figure16b: Enostosis of humerus. Note cauliflower like osteochondroma

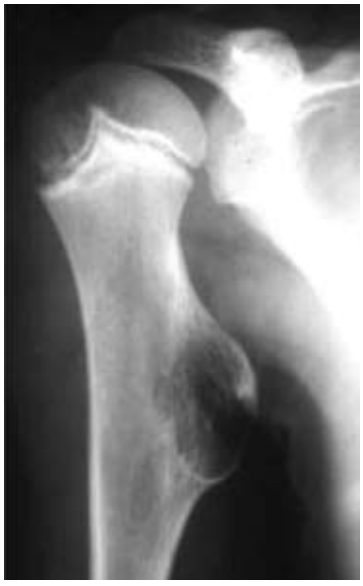

Figure 16c: Sessile Exostosis of humerus seen end on

diametaphysis and projects out of the underlying bone. It may be sessile or pedunculated. The femora and tibia are the common sitesof its occurrence. The medularic cavity is in continuity of the parental bone. In long bones, these lesions grow away from metaphysis. Diaphyseal eclasis growth takes places in the cartilaginous cap. Growth generally diminishes after skeletal maturity (Figure 16abc). The average thickness of the cartilage is about 2cm.

Multiple osteochondromata (exostoses) are hereditary and autosomal dominant. These are

encountered in younger age groups. They are bilateral and often symmetrical. Radiologically, the findings are that of solitary exostosis. However, since they are developmental, shortening and deformity of limbs may be noted (Figure 17abcd).

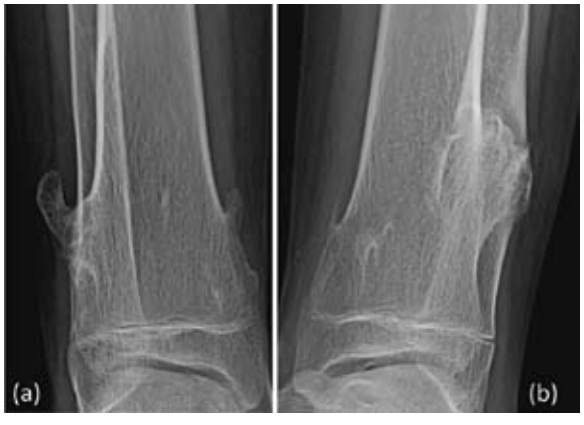

Figure 17ab: Bilateral osteochondromata. Note hook like projections

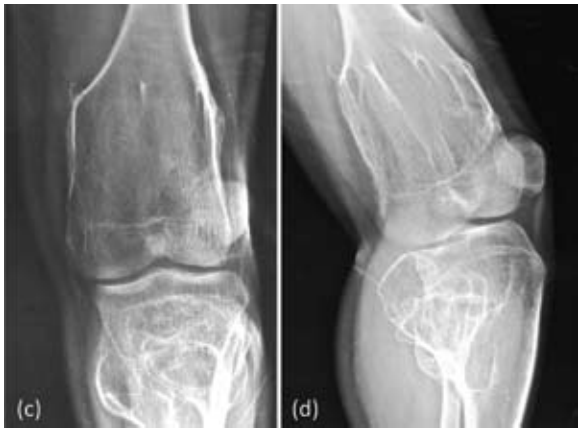

Figure 17cd: Multiple symmetrical osteochondromata of femur and tibia

Complications include fracture, bursal formation, compression of nerves and vessels and malignant transformation (Figure 18ab).

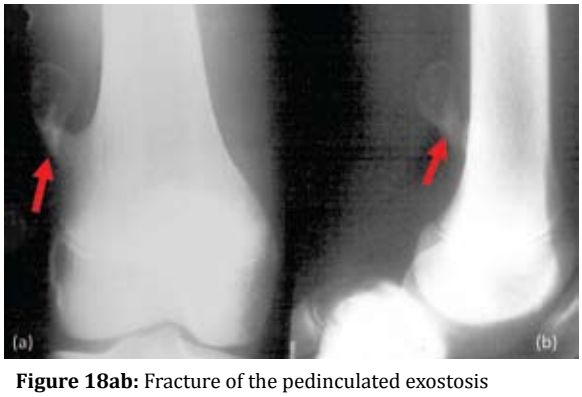

Figure 18ab: Fracture of the pedinculated exostosis

In solitary exostosis/ Enchondroma, the incidence of malignancy is ~1% (Figure 19) in multiple exostoses the incidence of malignant transformation may be up to 10 – 15%. In Maffucci syndrome the incidence of malignancy is 20 – 30% which includes non-osseous tumors.

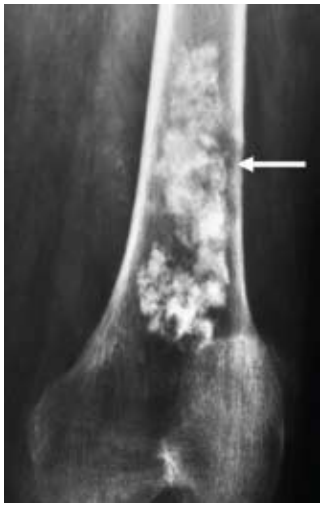

Figure 19: Benign enchondroma undergoing malignant transformation in the lateral aspect eroding the cortex.

Post traumatic exostosis has been described and when its shape is like a tent it is named as Turret exostosis. Another exostosis is due to osteoperiostitis or Nora's lesion as a sequel to trauma (Figure 20ab). Post radiation osteochondromata have also been described.

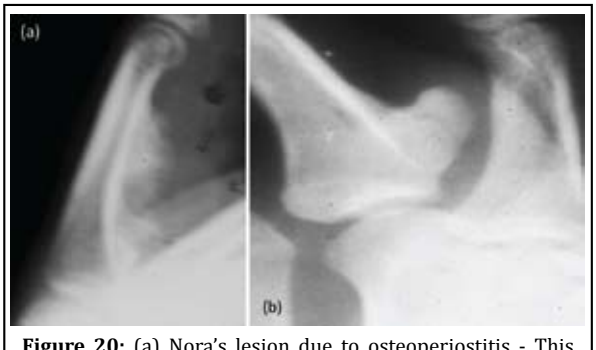

Figure 20: (a) Nora's lesion due to osteoperiostitis - This is also known as Bizarre Paraosteal Osteochondromatous Proliferation (BPOP) (b) Turret exostosis due to previous trauma.

Trevor's Disease (Dysplasia epiphysialis hemimelica) is a disorder with eccentric growth of ossification centre associated with calcified mass. It may be considered as epiphyseal osteochondroma. This may be hereditary and deformity of joint may develop. Generally, monoarticular but more than

one joint may be affected. In this case talus and distal femoral epiphyses are most commonly involved. Radiologically, focal calcifications occur at the epiphysis or apophysis. Affected centres may become hypertrophied resulting in severe deformities (Figure 21abc).

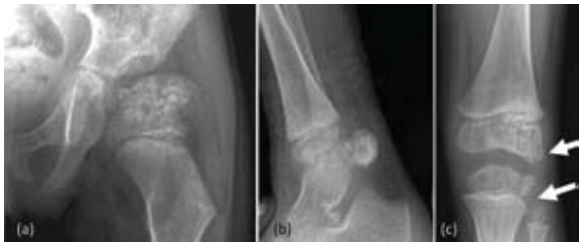

Figure 21: Trevor's disease involving (a) Femoral head (b) Talus and (c) Knee

Synovial chondromatosis may be primary or secondary. Primary synovial chondromatosis may be intra articular or extraarticular (Figure 22).

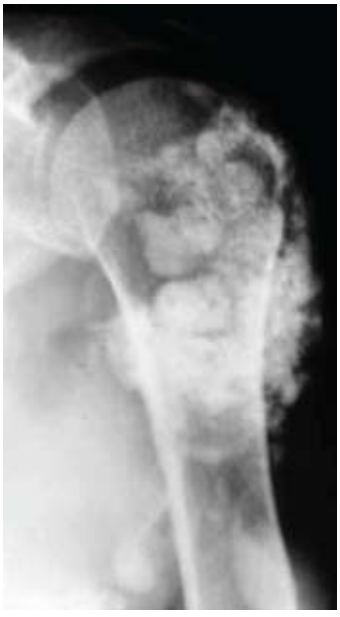

Figure 22: Primary synovial chondromatosis of the shoulder

Secondary synovial chondromatosis is due to advanced degenerative arthritis. The degenerated and fragmented cartilage and synovium may be separated and form a nidus for secondary synovial osteochondromatosis. Primary synovial chondromatosis is due to metaplasia of the synovium which gets fragmented and nourished by synovial fluid and eventually calcifies and ossifies and form chondromatosis (Figure 23ab).

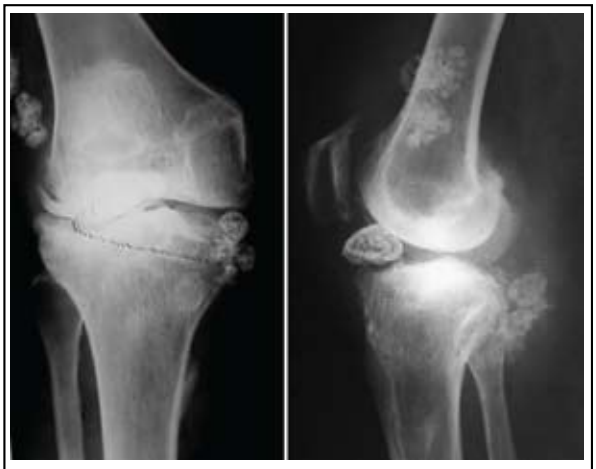

Figure 23ab: Osteoarthritis with secondary synovial osteochondromatosis (joint mice)

Conclusion

Benign cartilaginous tumors are common and constitute ~50% of all bone tumors. Conventional radiology is adequate enough to diagnose these tumors. Mineralization of matrix in the form of arcs, circles and flocculations is characteristic. Previously, we were the first to describe (7, 8) a "hook sign" in cartilaginous tumors. The classical radiological findings on conventional radiograph are illustrated.

Aknowledgements

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