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RESEARCH ARTICLE

CHONDROBLASTOMA IN THE DISTAL FEMUR: A CASE REPORT WITH LITERATURE REVIEW.

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Abstract

Chondroblastoma is an aggressive tumor of bone with the capacity for recurrence and metastasis with a relatively high incidence in older children and adolescents during the period of active epiphyseal growth. It is generally regarded as a benign neoplasm, but sometimes it grows aggressively or recurs.14-year-old young boy operated operated twice for chondroblastoma. Currently he presented with right knee joint pain, which started two months ago. Radiography and magnetic resonance imaging studies of the right knee showed concluded to recurrence of chondroblastoma of the distal femur. The histologic study of the biopsy curettage showed a chondroblastic with large polygonal cells. The final diagnostic was chondroblastoma.Chondroblastoma is a rare primary bone tumor of young people that typically arises in the ends of the long bones Radiologic investigations show a small, circumscribed, lytic lesion. The tumor is characterized histologically by the proliferation of chondroblasts along with areas of mature cartilage, giant cells, and occasionally, secondary aneurysmal bone cyst formation. Recently, immunohistochemical stains such as DOG1 and SOX9 have been described in Chondroblastoma.

Introduction:-

Chondroblastoma is an uncommon osseous neoplasm that accounts for less than 1% of all bone tumours. It characteristically arises in the epiphysis or epimetaphyseal region of long bones and has been reported to affect people of all ages with slight male predilection. The histogenesis of chondroblastomas is controversial, although cartilage stem cells or epiphyseal cartilage cells are presumed to be the cells of origin (1). It was first described as a "giant cell tumor with calcifications" by Kolodny in 1927, next as a "calcifying giant cell tumor" by Ewing in 1928 and as an "epiphyseal chondromatous giant cell tumor" by Codman in 1931 (2). Although it is generally regarded as a benign neoplasm, it sometimes grows aggressively or recurs; rarely, it metastasizes to the lungs (3). Clinical presentation typically consists of gradually increasing pain and local tenderness, followed by swelling and limitation of movement of the adjacent joint (4). Although the clinical presentation is highly variable, local ± gradually increasing ± pain, soft tissue swelling and tenderness are the most frequent presenting complaints (5). The typical radiological finding is an eccentric osteolytic lesion, frequently accompanied by a thin sclerotic rim (1). The tumor is characterized histologically by the proliferation of chondroblasts along with areas of mature cartilage, giant cells,

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and occasionally, secondary aneurysmal bone cyst formation (6). Recently, studies have documented novel immunohistochemical and molecular findings that may aid in the diagnosis (6).

To prevent recurrence, complete curettage is important; however, such an approach becomes extremely difficult to perform precisely when the chondroblastoma arises deep in the epiphysis (3).

Case presentation:

A 14-year-old boy operated two times for chondroblastomaof the distal femur. The first intervention was made two years ago and the second was one year ago. Currently, the patient presented right knee joint pain with joint motion and a decrease in weight-bearing ability, which started two months ago.

On physical examination, spontaneous pain, tenderness in the medial side of the right, solid mass and joint stiffness of the knee were noted. There was no color change or redness of the overlying skin. Laboratory test results showed no abnormalities. Radiographs of the right knee showed a radiolucent lesion with marginal sclerosis in the distal epiphysis of the right femur (Figure 1). Mri sagital slice: lesion of inferior femoral epiphysis in low signal T1 presenting enhancement by gadolinium (Figure 2). The location and morphology of this lesion, as well as the age of the patient, were suggestive for recurrence of chondroblastoma. A thorough curettage was performed and the histological examination showed large polygonal cells with prominent central nuclei rimmed embedded in a chondroid matrix (Figure 3). In the more cellular areas the chondroid matrix was less obvious and the tumour cells were intermingled with multinucleated giant cells. The final diagnosis was chondroblastoma.

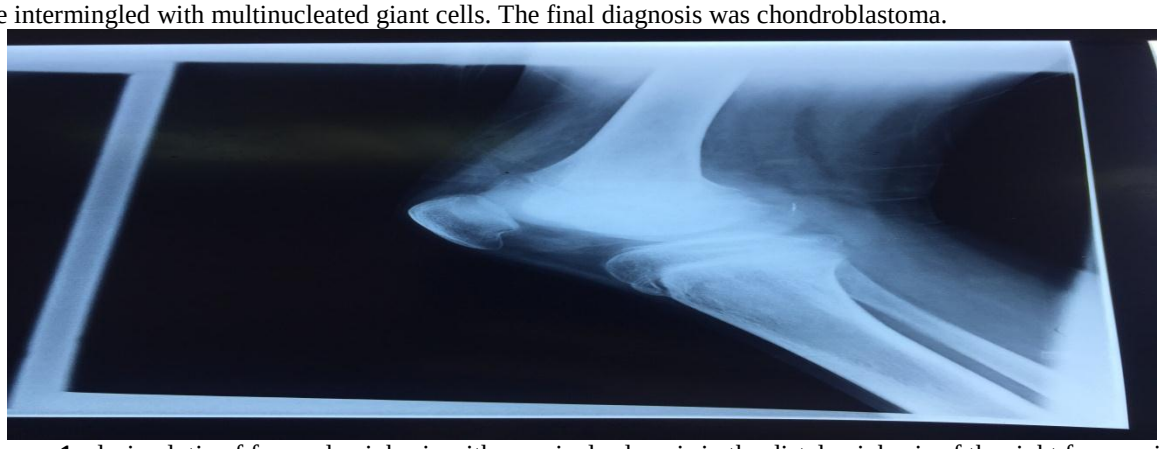

Figure 1:- lesion lytic of femoral epiphysis with marginal sclerosis in the distal epiphysis of the right femur with internal calcification

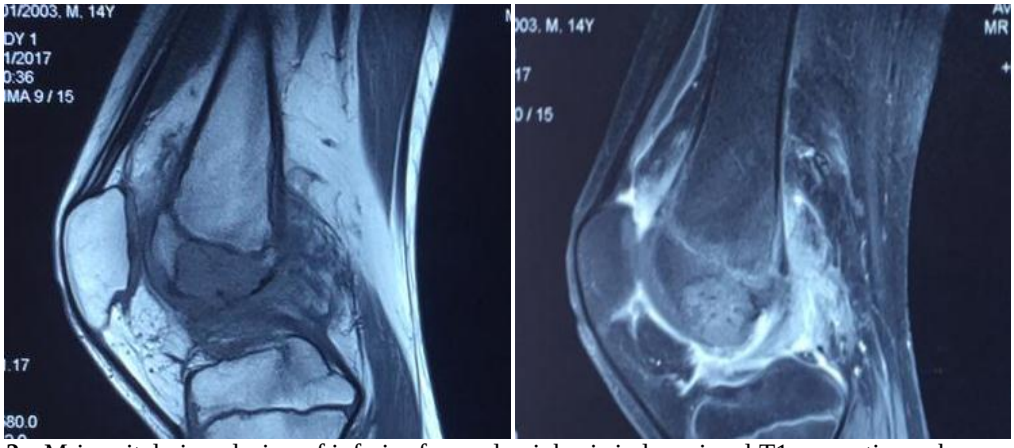

Figure 2:- Mri sagital view: lesion of inferior femoral epiphysis in low signal T1 presenting enhancement by gadolinium

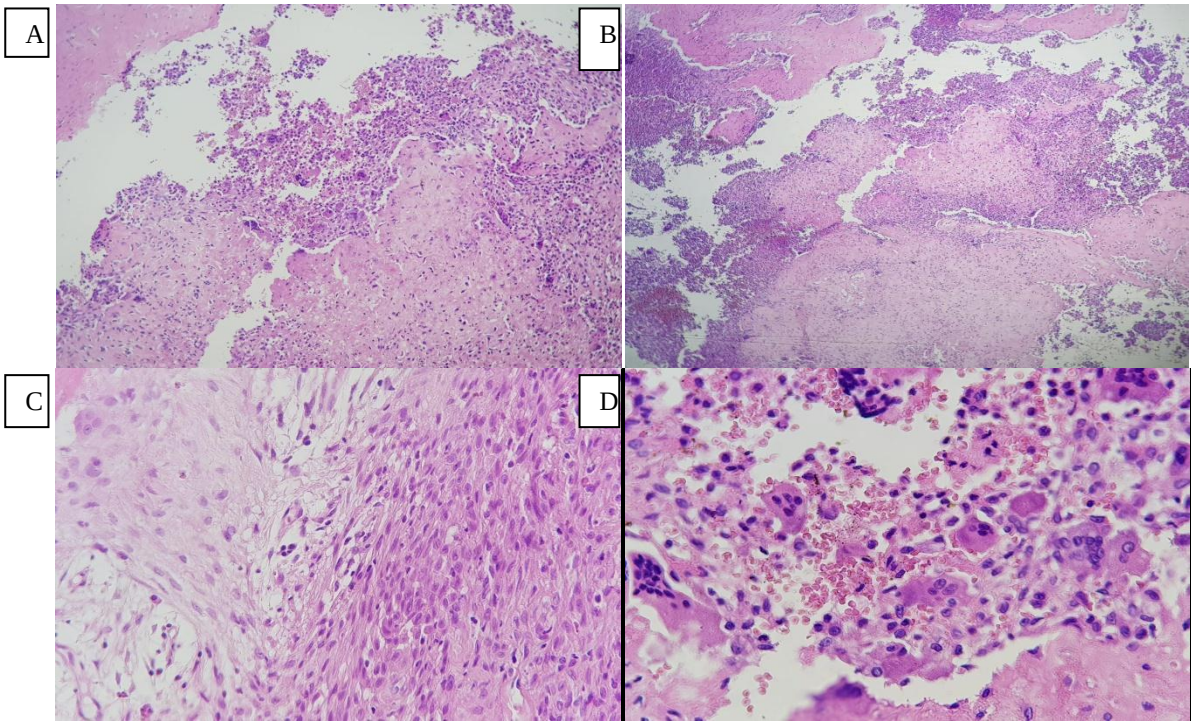

Figure 3:-A+B, Foci of cartilaginous tissue with a chondroblastic proliferation. C,Chondroblastoma is characterized by polyhedral tumor cells with nuclear grooves and eosinophilic cytoplasm mixed with scattered multinucleated giant cells. D, compact rounded tumor cells with bean-shaped nuclei, giant cellsand chondroid matrix.

Discussion:-

Chondroblastoma account for approximately 1% to 2% of all bone tumours and most patients are between 10 and 25 years of age at the diagnosis, with a male predominance (7).

WHO has defined chondroblastoma as 'a benign, cartilage-producing neoplasm usually arising in the epiphyses of skeletally immature patients (8).

Chondroblastomas occur mostly in the second decade of life (1). Males are affectedmore often than females in a ratio of approximately 2 to 1 (9). They usually arise from theepiphyseal plate or the epimetaphyseal region of long bones, although few primary metaphyseal or diaphyseal chondroblastomas have been reported (1). Bones that are most frequently involved are the proximal humerus, distal femur, proximal tibia and the iliac bones (10). Other unusual but classic sites of involvement include the talus, calcaneus, and patella (11). The differential diagnosis of a tumor or tumor-like lesion located in the epiphysis of a long bone includes: giant cell tumor, clear cell chondrosarcoma, osteomyelitis, intraosseous geode, Paget disease and osteochondral lesion (2).

Bone pain is the most common presentation, and may be longstanding, but other more site-specific complaints can also arise (6). Although the clinical presentation is highly variable, local ± gradually increasing ± pain, soft tissue swelling and tenderness are the most frequent presenting complaints. Sometimes pain is related to the nearest joint, with loss of joint function, joint effusion and limitation of movement. Pathological fractures occur (12). Typically on a radiograph a chondroblastoma presents with an eccentrically or centrally located osteolytic lesion with either smooth or lobulated margins with a thin sclerotic rim that involves the epiphysis or other secondary ossification centres (13). In 20% to 25% of the cases metaphyseal involvement is also seen (14). Cortical expansion with erosion and periosteal reaction may be present and occasionally unusual radiological changes are also seen (1).

Grossly, CBT is sharply separated from the adjacent bone and contains a mixture of soft, friable, grey-yellow material and hemorrhage (15). Small calcifications provide a gritty and chalky cut surface. Occasionally, areas of rubbery blue-grey chondroid matrix are seen. Necrosis and hemorrhagic cystic cavities (secondary aneurysmal bone cyst formation), mayalso be present, but usually only comprise a small portion of the tumor (6).

Histologically, the most characteristic finding of chondroblastomas is linear deposition of calcification surrounding individual chondroblasts, creating a chicken wire pattern (16). Chondroblastoma is characterized by a sheetlike proliferation of small to intermediate-sized round polygonal cells. The cytoplasm is eosinophilic, although focally, clear cell change can be seen. The nucleus is centrally placed and relatively large, and often a central, longitudinal nuclear groove (‘‘coffee bean’’ nucleus) can be seen. Nucleoli are small. Cellular atypia with enlarged, irregular, and sometimes hyperchromatic nuclei may be present, especially in tumors located in the skull and facial bones (17). Mitoses are occasionally found, although they are not numerous, with an average count of 1 to 3 mitotic figures per 10 high-power fields. Atypical mitotic figures should not be seen, and if present, tend to exclude CBT from the differential diagnosis (6). Randomly distributed osteoclast-type giant cells are almost always present. Variable area of deposition of chondroid material accompanies the chondroblast as are foci of hemosiderin deposition (6,18). Matrix formation must be seen

to confirm a diagnosis of CBT. Bluish or purple granular calcium deposits are seen in approximately one-third of cases, most commonly in long bone tumors. The calcifications may be seen in the cytoplasm or stroma, where they demonstrate a delicate pericellular lacelike or ‘‘chicken-wire’’ appearance (6). Other features such as tumor necrosis, vascular invasion, cortical breakthrough, and soft issue invasion can also be present in a small percentage of cases. The tumor necrosis is usually composed of bland ghost cells without any inflammation. Concurrently, the cytological features on fine-needle aspiration cytology from the swelling revealed good cellularity smears with fragments of chondroid matrix admixed with multinucleated osteoclast-like giant cells and mononuclear cells with distinct cell borders, round-to-oval nuclei and dense eosinophilic cytoplasm. Immunohistochemical staining shows reactivity of the neoplastic cells for S100 protein and vimentin, although the expression of other antigens has been reported, with cytokeratin being among the most commonly observed (11). New marker, discovered on gastrointestinal stromal tumor 1 (DOG1), also shows positivity in CBT tumor cells and may serve as a specific marker for differentiating CBT from other giant cell–containing bone tumors (19). Sox9 is a transcriptional factor known to be involved in chondrogenesis and also shows positivity in CBT and chondromyxoid fibroma in a nuclear distribution (6). Sjogren et al found recurrent breakpoints at 2q35, 3q2123, and 18q21 and detected rearrangements ofchromosome band 8q21 exclusively in aggressive chondroblastomas (20). Ostrowski et al reported a patient with malignant transformation of a recurrent pelvic chondroblastoma with a p53 mutation (21). There have also been other reports of abnormalities in chromosomes 5, 8, 11, and 17 in patients with chondroblastoma. These recurrent structural chromosomal abnormalities suggest that there may be preferential involvement of these chromosomes in chondroblastoma (11).

Intralesional treatment consisting of curettage, often followed by packing of the cavity with cancellous bone chips (autologous or homologous bone) or local resection (if there is a suspicion of malignancy), have generally been sufficient for successful treatment. In exceptional cases, when a surgical approach was impossible or not accepted, radiotherapy cured the lesion (9).

Local recurrence is the most frequent complication. There is no significant difference in the recurrence rates, regardless of the age or sex of the patient, size of the lesion, amount of calcification or vascular invasion seen on histological examination, duration of followup, or method of treatment (11). Although generally benign, CBT is placed in the ‘‘intermediate, rarely metastasizing’’ category in the 2013 World Health Organization classification of bone tumors (22). Metastasis to the lung, bone, and soft tissue can occur in CBT, although such cases are rare 2% (6). Almost no mortality occurs (0.2%). Spontaneous healing exists (9).

Conclusion:-

Chondroblastoma represents a rare but distinctive cartilaginous bone tumor affecting predominantly children and adolescents. Characteristic features include a small, lytic lesion in the epiphysis of long bones, and the finding of chondroblasts, chondroid matrix, and assorted secondary elements on microscopy. Radiological findings should always be taken into consideration while confirming the diagnosis of chondroblastoma or any other bone and cartilaginous tumours. Long-term clinical follow-up is mandated because of high risk of recurrence and rare occurence of malignant transformation.

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