Abstract: Introduction- Langerhans cell histiocytosis of bone (eosinophilic granuloma) is a benign neoplasm characterized by localized proliferation of predominantly eosinophils and histiocytes. A high index of suspicion is required for making an early diagnosis and initiating appropriate treatment. Different forms of treatment have been reported to give satisfactory results. We report a case of eosinophilic granuloma of femur in a 30 year male presenting with pathological fracture. Case presentation: A 30 year male patient presented with pain over right thigh with history of trivial fall and fracture of shaft of femur. He had pain in his right thigh for 6 months duration. On investigating, X ray showed osteolytic lesions and periosteal reaction with pathological fracture in proximal metaphysis and diaphyseal region. He was treated with open biopsy and external fixation. Biopsy revealed langerhan cell histiocytosis of femur for which he was treated with radiotherapy. External fixator was removed, later interlocking nailing was done. Post operatively he was given a second cycle of radiotherapy. Conclusion: Patient symptomatically improved after the initiation of treatment. LCH is an uncommon disease and presenting with pathological fracture is a rarity. Literature search revealed no case has been reported so far with this kind of presentation. Keyword: Langerhan cell histiocytosis, eosinophilic granuloma, osteolytic lesion, nailing

INTRODUCTION:

Langerhan cell histiocytosis is a proliferative disorder of the Langerhans cells, which are dendritic cells that normally populate the skin, mucosal surfaces, lymph nodes and other tissues where they function as specialized antigen presenting cells. The clinical spectrum of Langerhans cell histiocytosis (LCH) is wide, ranging from potentially lethal leukaemia like disorder to a solitary lytic lesion of bone. Localised LCH of bone is a benign tumour like condition which is characterized by a clonal proliferation of Langerhans type histiocytes, and is commonly referred to as eosinophilic granuloma. The bones which are the most commonly involved are the skull, the pelvis, and the diaphysis of long bones. The systemic condition includes Hand-Schüller- Christian disease with a triad of exophthalmos, diabetes insipidus and osteolytic lesions of the skull, and Letterer-Siwe disease with hepatosplenomegaly, lymphadenopathy, skin rash, fever, anaemia and thrombocytopenia. A variety of forms of treatment has been reported for solitary eosinophilic granuloma of bone, including observation, injections of steroid, local excision and curettage with or without bone grafting, chemotherapy and irradiation. All of these treatments are reported to give satisfactory results with a recurrence rate of less than 20%.

CASE PRESENTATION:

A 30 year male presented to our department with complaints of pain and deformity over right thigh after a trivial fall. Previously, he had pain on and off during night, often associated with fever for which he had been taking medications in the form of analgesics. On examination, patient general condition was stable, right thigh showed swelling over middle third, tenderness and abnormal mobility. X ray of right thigh showed pathological fracture in diaphysis with osteolytic lesions and periosteal reaction in both diaphysis and metaphyseal region. Upper tibial pin traction was applied. Except for a mild increase in ESR other blood parameters were normal. Peripheral smear showed microcytic hypochromic anaemia. Bone marrow study showed mild increase in eosinophilic precursors and no abnormal histiocytes. X ray of skull showed pathological fracture in diaphysis with osteolytic lesions and periosteal reaction in both diaphysis and metaphyseal region. Upper tibial pin traction was applied. Except for a mild increase in ESR other blood parameters were normal. Peripheral smear showed microcytic hypochromic anaemia. Bone marrow study showed mild increase in eosinophilic precursors and no abnormal histiocytes. X ray of skull and other long bones showed no other similar lesion in bones. USG & CT abdomen, CT thorax showed no evidence of malignancy. We did MRI of the thigh showed fracture of proximal third of femur, with surrounding fluid collection and soft tissue edema and periosteal thickening around the fracture site. We proceeded with open biopsy and external fixation. Histopathological examination showed bony trabeculae separated by loosely arranged sheets of inflammatory cells, predominantly macrophages, eosinophils and giant cells, suggestive of langerhan cell histiocytosis. Post operatively, patient was given radiotherapy of 12 Gy in 6 fractions. After 1 month external fixator was removed and interlocking nailing was done.
asymptomatic, no treatment is necessary because lesions have been noted to regress spontaneously. In spine it presents with marked flattening of vertebral body or vertebra plana. The involvement of long bones are presented as expansile lytic lesions with soft tissue mass and laminated periosteal reaction.

Microscopically, the diagnosis is made by the identification of Langerhans cells. The cells stain positively for S-100 protein. The lesion also contains multinucleated giant cells and other inflammatory cells, including clusters of eosinophils. Electron microscopy may identify characteristic organelles in the Langerhans cell cytoplasm called Birbeck granules.

CT and MRI shows cortical erosion and soft tissue involvement and aid in biopsy and surgical planning. Bone scan may be helpful in identifying additional lesions.

Biopsy is required to make the diagnosis. Recommended treatments have included steroid injections, radiation therapy, and curettage with or without bone grafting. If a lesion is small and asymptomatic, no treatment is necessary because lesions have been noted to regress spontaneously.

Similarly, if the diagnosis is established by open biopsy, the lesion can be curedtted during the same procedure. Also it responds well to low dose radiotherapy which is usually reserved for inaccessible areas.

The differential diagnosis which has to be considered are Osteomyelitis, Round Blue Cell Tumours, Metastases, Primary Bone Tumours, Lymphoma / Leukemia. On reviewing the literature, in journal of orthopaedics and sports physical therapy, 2011 eosinophilic granuloma was reported with hip pain in a 33 year old patient.

Langerhan cell histiocytosis has many appearances that depend on the stage of disease, and it may mimic other conditions. Biopsy is required to confirm the diagnosis in all patients suspected of having this disease. In pathological fractures of long bones, internal fixation by interlocking nailing is a viable option which gives good results. Treatment depends on the stage of the disease. Small lesions can be treated with injection of methylprednisolone or it can be completely curetted with or without bone grafting. For bigger lesions, with impending fracture, an accessible bone like femur, has to be treated with radiotherapy and fracture stabilized with external fixator or internal fixation. For in accessible lesions, it can be exclusively treated with radiotherapy alone. In our case, since the lesion is bigger involving the diaphysis of femur, we have given radiotherapy combined with stabilization of fracture. The overall prognosis for skeletal lesions is excellent with a very low rate of local recurrence and few complications.

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Fig 1-X ray of femur showing osteolytic lesions and fracture.
Fig 2 - X-ray of skull

Fig 3 - X-ray of long bone (humerus)

Fig 4 - MRI of affected femur

Fig 5 - Fracture treated with biopsy and external fixation

Fig 6 - X-rays showing definitive fixation with interlocking nailing

Fig 7 - X-rays showing definitive fixation with interlocking nailing