University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2018, Vol. 4(1)

EOSINOPHILIC GRANULOMA OF FEMUR WITH PATHOLOGICAL FRACTURE - A CASE REPORT

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Abstract : Introduction- Langerhans cell histiocytosis of bone (eosinophilic granuloma) is a benign neoplasm characterized by localized proliferation of predominantly eosinophils andhistiocytes. A high index of suspicion is required for making an early diagnosis and initiating appropriate treatment1. 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 withhistory 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 withopen 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 : langerhanscell 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[2][3]. Localised LCH of bone is a benign tumour like condition which is characterized by a clonal proliferation of Langerhans type histiocytes [3][4], 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[5]. .

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 (***fig 1***) 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 (***fig 2***) and other long bones (***fig 3***) showed no other similar lesion in bones. USG & CT abdomen , CT thorax showed no evidence of malignancy. we did MRI (***fig 4***) 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 (***fig 5***). 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 (***fig 6***) was done.

Post operatively a second cycle of radiotherapy of 8 Gy in 4 fractions was given.

DISCUSSION :

The term eosinpophilic granuloma was coined by Lichenstein and Jaffe in 1940. The skeletal system is the commonest site of involvement of Langerhans cell histiocytosis, and in for 60 - 80% of cases is the only organ system involved. It primarily occurs in older children and young adults, with a male preponderance with a Male to Female ratio of 2:1. The most common locations are skull [8]: 49%, pelvis: 23%, femur: 17%, ribs: 8% (most common in adults), humerus: 7%, mandible: 7%. The initial differential diagnoses considered on the basis of the patient's history and the appearance of the lesion included Langerhans cell histiocytosis (LCH), osteomyelitis, and malignancy. The radiologic appearance of solitary LCH has a wide spectrum; however, less aggressive forms of LCH typically appear as a punched out lytic lesion that may have some degree of periosteal reaction and cortical thickening[6]. Malignancy cannot be excluded on the basis of the imaging findings alone; therefore, biopsy was necessary. In our patient, LCH was the most likely diagnosis based on the radiologic and clinical features.

The tentative diagnosis was confirmed at open surgical biopsy, and histologic work-up was performed to exclude malignancy and infection. The clinical picture may be similar to that produced by osteomyelitis with pain at rest (and at night), fever, and local signs of inflammation[7] . Patients may have one or many lesions.

In flat bones the lesions are well circumscribed,punched out,purely lytic lesions[7].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[7], 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[7]. 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.

For symptomatic lesions, if the diagnosis is established by needle biopsy, the lesion may be injected with methyl prednisolone during the same procedure.

Similarly, if the diagnosis is established by open biopsy, the lesion can be curetted 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[9].

Harpreet Singhand Satnam Kaur reported a unifocal granuloma of femur due to langerhan cell histiocytosis in a 8 yr old child[1]. Christine M. Pui and Harry E. Jergesen reported femoral involvement langerhancell histiocytosis following total hip arthroplasty in 2011[10].

CONCLUSION :

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 .

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,in 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

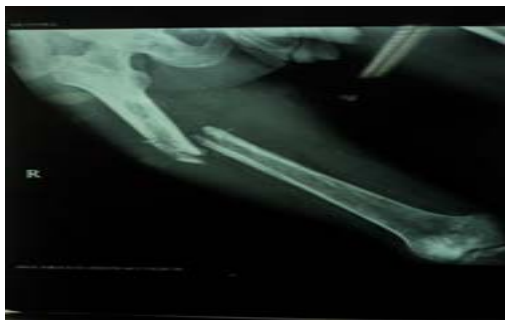

Fig 2-X ray of skull

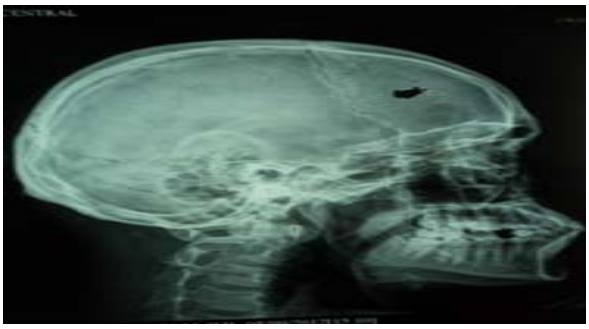

Fig 3-X ray of long bone (humerus)

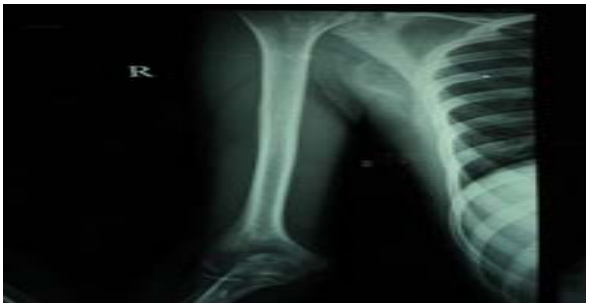

Fig 4-MRI of affeceted femur

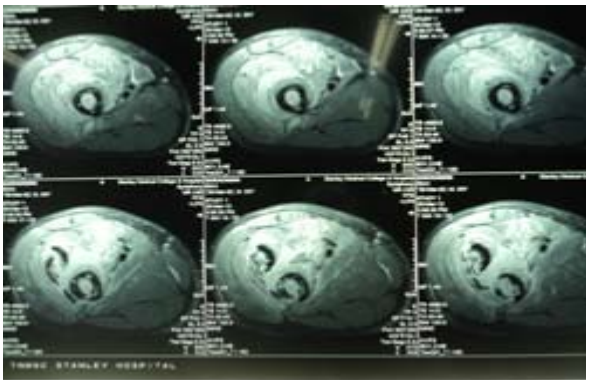

Fig 5-Fracture treated with biopsy and external fixation

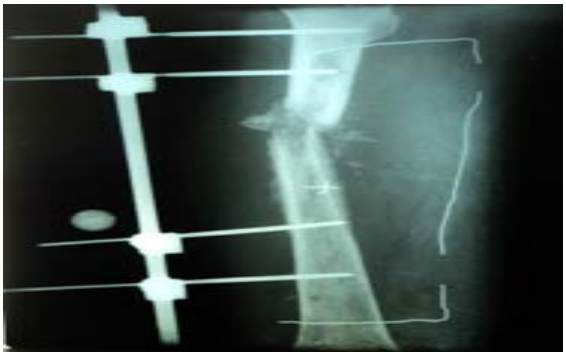

Fig 6-X rays showing definitive fixation with interlocking nailing

Fig 7-X rays showing definitive fixation with interlocking nailing