

## “Eosinophilic granuloma” Mono ostotic type in a child

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**Abstract:** Eosinophilic granuloma (EG) is a solitary, non-neoplastic proliferation of histiocytes. EG is part of a spectrum of Langerhan's cell histiocytosis, formerly known as histiocytosis X. The male to female ratio is two to one. It occurs most commonly in children aged 5 to 10 and is uncommon in blacks. EG is normally symptomatic, local pain, swelling and tenderness are common. We report this case of Eosinophilic granuloma (EG) Mono ostotic type, in a 8 years female child, because of its rarity.

**Key words to paper:** Eosinophilic granuloma, Monoostotic, Bone lytic lesion, Histiocytosis X.

### I. Introduction

Eosinophilic granuloma (EG) is the benign form of three clinical variants of langerhans cell histiocytosis (LCH). The other two variants are Letterer-Siwe disease and Hand-Schüller-Christian disease. The term, “eosinophilic granuloma” was first introduced by Lichtenstein and Jaffe in 1940[1]. EG is characterized by single or multiple skeletal lesions, and predominantly affects children, adolescents, and young adults. Solitary lesions are more common than multiple lesions. When multiple lesions occur, the new osseous lesions appear within one or two years. Any bone can be involved; the more common sites include the skull, mandible, spine, ribs, and the long bones.[2–4] and it makes up 60-80% of all cases of Langerhan's cell histiocytosis. EG consists of sheets of Langerhan's cells. These cells are derived from the mononuclear cell and dendritic line precursors and are found in the bone marrow. The cell is identifiable under the electron microscope as the Langerhan's cell has racket shaped cytoplasmic inclusion bodies called Birbeck's granules. Also present in the lesion are varying amounts of lymphocytes, polymorphonuclear cells, eosinophils and giant cells. Early lesions have many Langerhan's cells and eosinophils. Older lesions have fewer cells and much fibrous tissue. The cause of EG is unknown and speculated to be either infection or immunological. EG has got a good prognosis and may spontaneously regress; it is extremely radiosensitive.[5]

### II. Case Report

A female child presented with pain and swelling in the upper part of right thigh since 6 months. There was no history of trauma. The results of the hematological investigations were within normal limits. Pelvis radiograph revealed a single punched out area of bone destruction with sharp margins in the metaphyseal area of right femur[Figure 1]. A chest radiograph and ultrasonography of the abdomen did not reveal any abnormality. MRI sections of the thigh Showed well defined altered signal intensity, mono ostotic lesion involving metaphysis of right femur and shows well defined margin measuring 30x22x18 mm with no cortical break/internal bony fragments/calcification/periosteal reaction seen.

We received multiple curettings from lytic lesion for histopathological examination. H&E sections revealed sheets of large cells with moderate cytoplasm and round to oval nuclei with nuclear grooving and indentations. These cells are mixed with clusters of histiocytes, eosinophils and few multinucleated cells[Figure 2]. Sections were subjected for CD1a and S100 immuno histochemistry markers and both of them were positive [Figure 3,4].

### III. Discussion

Eosinophilic granuloma is a benign disorder that affects children and young adults, particularly males. Solitary EG accounts for the majority of LCH cases, usually involving bone and less commonly the lymph nodes, lung or skin [7]. The solitary bone lesion may be asymptomatic, or it may cause bone pain because of the expansion of the medullary bone; pathological fractures may ensue.[2,8]. The distinctive morphological lesions of the entire group of Langerhans histiocytosis disorders consist of expanding erosive accumulations of histiocytes, usually within the medullary cavity.

The clinical and radiographic findings are often not specific enough to determine the diagnosis. Cytology is very helpful in arriving at the diagnosis of eosinophilic granuloma of the bone[7]. Morphologically the key feature is the identification of Langerhans cells with characteristic grooved, folded, indented nuclei in the appropriate milieu that includes variable numbers of eosinophils and histiocytes including multinucleated forms, often appearing similar to osteoclasts or touton like giant cells, neutrophils and small lymphocytes[7,9]. The concentration of the eosinophilic infiltrate varies from scattered mature cells to sheet-like masses of cells. Occasionally, areas of bone necrosis may interrupt the cellular infiltrate. The foamy cells may also be amassed in clumps, which are of no clinical significance because these clumps represent phagocytosis of lipid debris.

Any bone can be involved. The skull, long bones of the upper extremities and the flat bones are affected in descending order of frequency[7]. Solitary lesions are more common than multiple ones. When the lesions are multiple, new osseous lesions occur within one or two years but the condition is still classified as EG. Radiologists need to be aware that additional EG of the bone occurring as long as four years after initial diagnosis, should be interpreted as a localized form of Langerhans cell histiocytosis. This differentiation is important because the prognosis is more favourable with focal disease with multifocal disseminated disease, which involves organs other than the skeletal system. Similar lesions may occur within the lungs, skin, and stomach, either as a unifocal lesion or as part of multifocal disease. Lung involvement occurs in 20% of the patients with EG and in an older group (age, 20–40 years). Lung involvement has a strong association with smoking. Diffuse pulmonary infiltrates may be a manifestation of a covert osseous EG[5]. In 50–75% of the patients, the disease is monostotic and skull involvement is seen in 50% of the patients. The disease has the ability to regress spontaneously and is significantly radiosensitive. The prognosis of eosinophilic granuloma has been found to be good[10].

According to Teplick and border only 5.0 percent occur in persons over 30 years of age and 75 per cent of the lesions are solitary. They also state that 59 per cent of the lesions involve the skull, ribs and femurs, although any bone may be involved [6]. O’Neill et al., in their review of 189 patients found that 119 of the cases were males and 48 females.

#### **IV. Conclusion**

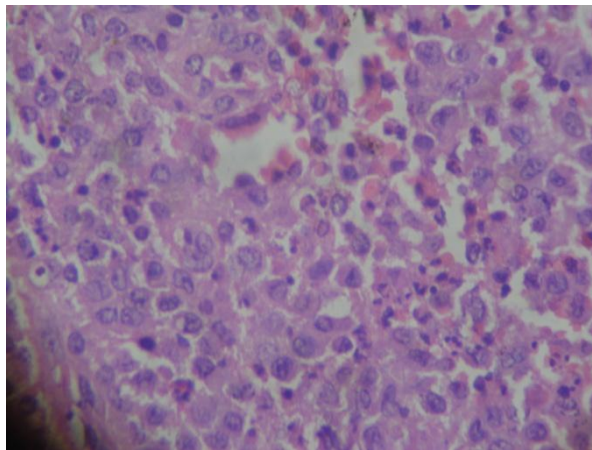
Eosinophilic granuloma is a rare benign disorder that affects children and young adults, particularly males. We present this case in a female child involving single bone.

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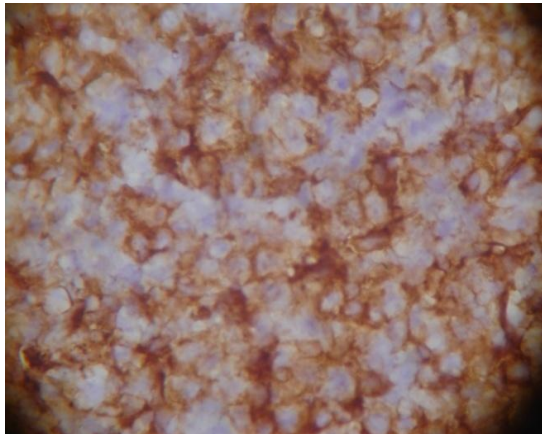
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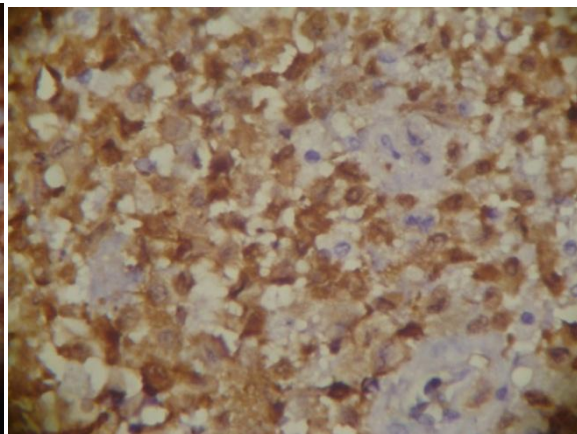
**Figure [1]** X ray – lytic lesion in the metaphysis of Right Femur.



**Figure [2]:**H&E Section shows Langerhans cells and eosinophils.



**Figure [3]:** CD1A-Positive.



**Figure [4]:** S100-Positive.