



## Case Report

# An Unusual Case of Solitary Swelling over Forehead: Think Eosinophilic Granuloma

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## Abstract

*Langerhans cell histiocytosis is a rare disorder in children. Eosinophilic granuloma is the benign variant of this disease. We report the case of eosinophilic granuloma in a 7 years old male child who presented with right sided solitary swelling over right side of forehead associated with mild dull pain. Plain radiography revealed a solitary gap in frontal bone, computed tomography scan gave a better delineation. Fine needle aspiration cytology was done and histopathology clinched the diagnosis of eosinophilic granuloma. Single boggy swelling over scalp bone of a male child should raise the suspicion of eosinophilic granuloma.*

**Keywords:** *Eosinophilic granuloma; Langerhans cell; Aspiration cytology.*

## Introduction

There are three clinical variants of langerhans cell histiocytosis (LCH): Letter-Siwe disease, Hand-Schuller-Christian disease and Eosinophilic granuloma (EG). Among these three forms EG is the benign form. The term, "eosinophilic granuloma" was first introduced by Lichtenstein and Jaffe in 1940<sup>[1]</sup>. EG is characterised by single or multiple lesions commonly involving long bones, skull, spine, ribs, mandible etc.<sup>[2]</sup> and predominantly affects children, adolescents, and young adults. Solitary lesions are more common than multiple lesions. We present such a case of eosinophilic granuloma in a 7 years old male child

who presented with slightly painful forehead swelling.

## Case Report

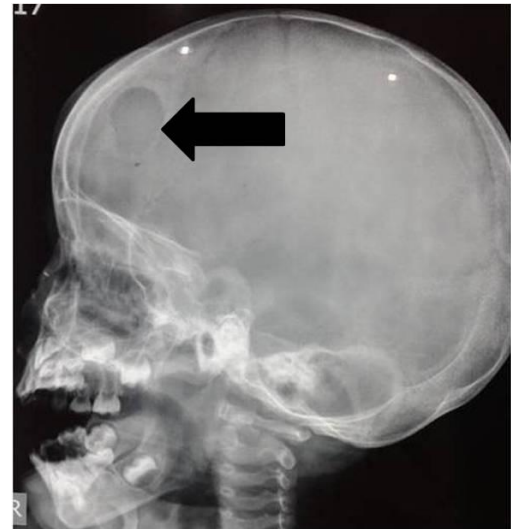
A 7 years old male child presented to the Department of Pediatric Medicine with complaint of swelling over right side of forehead [Figure 1]. It was insidious in onset and gradually progressive, parents first noticed it 8 months back. The patient also complained of dull continuous pain associated with the swelling for the last 2-3 months. On inspection, it was a 6 cm X 5 cm, swelling with no sinus, venous prominence or scar mark or any feature of inflammation and skin

change. On palpation, a swelling was felt in the frontal region of the skull which was soft in consistency with minimal tenderness over the swelling. The bone seemed to give way in the central part. There was no history of trauma, seizure, visual disturbance, vomiting etc. associated with the lump. Routine hematological investigations were within normal limits. Systemic examination of the whole body was also within normal limits.

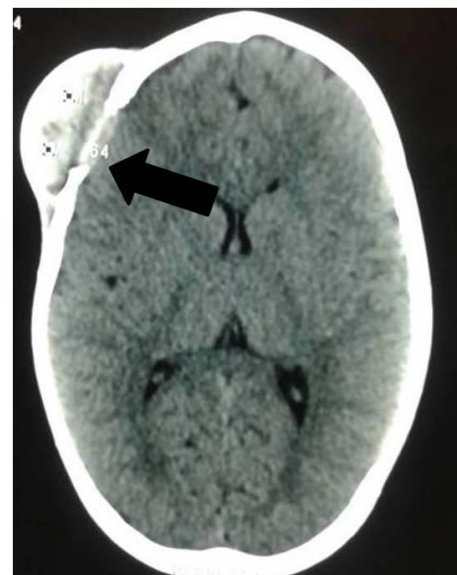
Skull radiograph (lateral view) revealed a single punched out area of bony dehiscence with sharp margins in the frontal region [Figure 2]. A chest radiograph and ultrasonography of the abdomen did not reveal any abnormality.

Axial computed tomography (CT) sections of the cranium showed an osteolytic lesion in the right side of frontal bone [Figure 3]. Enlarged film showed both inner and outer tables of the skull were eroded. Contrast study did not reveal any additional anomaly.

Fine needle aspiration of the lesion revealed sheets of isolated eosinophils intermixed with macrophages and giant cells. Macrophages contained ingested debris and red blood cells [Figure 4]. Eosinophilic granuloma of the frontal bone was diagnosed on the basis of cytological and radiological findings.



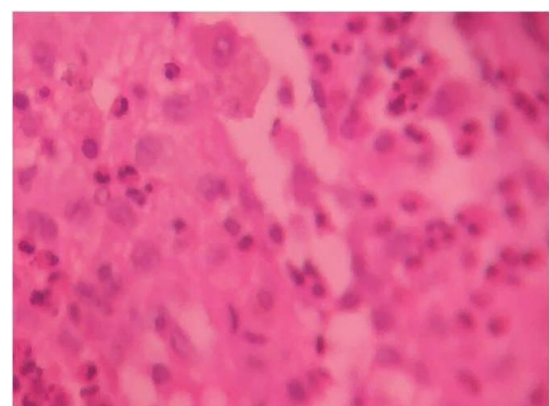
**Figure 2:** Plain lateral radiograph showing solitary lytic lesion of frontal bone



**Figure 3:** Axial CT scan brain showing the swelling and gap in the right side of frontal bone



**Figure 1:** Large oval shaped swelling over right side of forehead



**Figure 4:** Histopathology examination of FNAC tissue (Giemsa stain) showing sheets of isolated eosinophils intermixed with macrophages and giant cells (Langerhans cell histiocyte).

## Discussion

Langerhans cell histiocytosis is a rare disorder with an incidence of approximately 1 to 200,000 children. Eosinophilic granuloma represents less than 1% of bone tumours. Most commonly it affects children with certain predilection to males (male:female ratio=2:1) [3]. Solitary EG accounts for the majority of LCH cases, usually involving bone and less commonly the lymph nodes, lung or skin [4]. In 50–75% of the patients, the disease is monostotic and skull involvement is seen in 50% of the patients [5]. Langerhans histiocytosis group of disorders are distinguished by expanding erosive histiocytes within medullary cavity.

The lesion can be asymptomatic or it may be associated with pain due to expansion of medullary bone or rarely pathological fractures [6]. Also there may be tenderness, fever or frank swelling like in our case. Usually these lesions start to resolve after three months but sometimes it may take even two years. Depending on the location, EG may cause neurological symptoms such as numbness, limping, fracture, loosening of teeth, otitis media [7] or exophthalmos. Hematoma after a mild injury is a common finding of EG involving skull [8]. Routine haematological examination may reveal leucocytosis and eosinophilia in 7% of EG patients, erythrocyte sedimentation rate remains above normal limits [9].

Radiological investigations are necessary to note the activity and nature of the tumour. Plain radiographs are used to see the size and borders. Cortex of affected bone may become thin, eroded or sometimes thickened due to new bone formation. Lesions involving skull may be single or multiple, punched out osteolytic oval defects with regular or irregular borders. CT and MRI are needed to know exact size and borders as well as condition of surrounding tissues [10]. Radionuclide bone scan reveals an enhancing mass and detect other foci or recurrence points. Ultrasound is only used for guided biopsies.

The clinical and radiographic findings are usually non-specific to determine the diagnosis. Cytology

is very helpful in arriving at the diagnosis of eosinophilic granuloma of the bone [4].

Morphologically the key feature is the identification of langerhans cells with characteristic grooved, folded, indented nuclei with presence of variable numbers of eosinophils and histiocytes including multinucleated forms, often appearing similar to osteoclasts or touton like giant cells, neutrophils and small lymphocytes [4,11].

Eosinophilic granuloma does not lead to malignant transformation. If it expands elsewhere but bones then is called Hand Schuller Christian disease and may manifest as diabetes insipidus, cerebellar, hypothalamic and with other central nervous system symptoms [12]. The disease has the ability to regress spontaneously and is significantly radiosensitive. The prognosis of eosinophilic granuloma has been found to be good [5].

## Conclusion

This is one of the rare cases of eosinophilic granuloma of the skull developing in a pediatric patient. It is important to include eosinophilic granuloma in the differential diagnosis of bone lesions of scalp in children because of the possible expansion of the disease if untreated. We suggest surgical curettage and plastic bone transplant if not resolved on its own and radiotherapy following curettage as principal treatment. A follow up of a year is necessary for probable recurrence.

## Acknowledgement

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## Abbreviations

EG: Eosinophilic Granuloma;  
LCH: Langerhans Cell Histiocytosis;  
CT: Computed Tomography;  
MRI: Magnetic Resonance Imaging;

**Source of Support:** Nil

**Conflict of Interest:** None declared.

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