

# Eosinophilic Granulomata in a 4 Year Old Girl – A Rare Condition

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**Abstract** Langerhans' cell histiocytosis (LCH) is a destructive osseous lesion characterized by eosinophils and histiocytes. Eosinophilic granulomata is a localized and mildest form of LCH. A four year old girl presented with mild pain and mobility of prematurely seen mandibular permanent first molar and primary molars on the right side of mandibular region. The gingiva in the involved area was inflamed. A well-defined radiolucency approximately 2cm in diameter with scooped out effect was seen in panoramic radiograph. Extraction of the involved teeth and surgical curettage of the lesion was done under general anesthesia. Obtained bony tissues were histopathologically examined which revealed histiocytic infiltration, further the condition was confirmed by immunohistochemistry. The child underwent chemotherapy and steroidtherapy and responded good.

**Keywords:** *histiocytosis, eosinophilic granulomata*

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## 1. Introduction

Langerhans' cell histiocytosis (LCH) was formerly known as histiocytosis X refers to a group of conditions characterized by the uncontrolled stimulation and proliferation of a normal antigen-processing cell, the Langerhans' cell. [1] These Langerhans' cells can be identified by the presence of characteristic organelles, the Birbeck granules seen under electron microscopy and they are positive for S-100 antigen and CD1a surface antigen [2].

LCH is classified into three groups [3] - unifocal, multifocal unisystem, and multifocal multisystem. Unifocal LCH, also called eosinophilic granuloma is a slowly-progressing disease characterized by an expanding proliferation of Langerhans cells in various bones. Eosinophilic granuloma arises predominantly as a solitary bony lesion with mandibular involvement. [4] Multifocal unisystem LCH is characterized by fever, bone lesions and diffuse eruptions on the scalp and in the ear canals, known as the Hand-schuller-christain triad. Multifocal multisystem LCH, also called as Letterer-siwe disease, is a rapidly-progressing disease in which Langerhans cells proliferate in many tissues. Oral lesions may be the earliest manifestation of the condition, and in many cases, the mouth may be the only site of involvement. [5,6] The mandible was more involved than the maxilla and the posterior region was the predominant site. [7] LCH usually affects children between 1 year and 15 years old, with a peak incidence between 5 years and 10 years of age with a male sex predilection [8].

The purpose of this report is to describe a case of unifocal bony Langerhans' cell histiocytosis with mandibular

involvement and to discuss the appropriate diagnosis and management of such cases seen in children.

## 2. Case Report

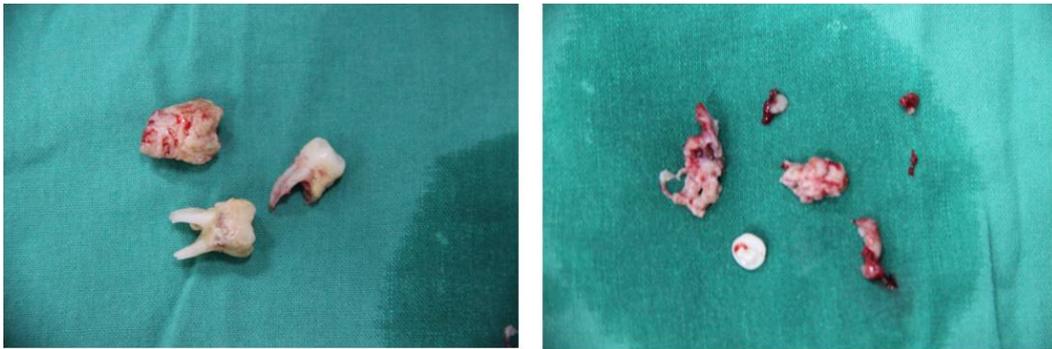
A 4-year old girl reported to the Department of Paediatric dentistry, Rajarajeswari Dental College and Hospital, Bangalore, India with a chief complaint of food lodgement and dull pain while chewing in lower right back teeth region since 5 months and completely stopped chewing since 1 month due to the mobility of the teeth at the involved side. The child was alert, moderately built and had no relevant medical history. Extraoral examination showed presence of a mild swelling present in the right side of angle of the mandible region along with palpable submandibular lymph nodes. Intra-oral examination showed marked mobility of mandibular right primary molars and permanent first molar, which was seen prematurely and mesially drifted due to severe bone loss. Gingiva of the involved area was inflamed and had a granular texture and bled on probing. There was also inflammation and recession of the gingiva of maxillary primary molars on the right side. The oral hygiene status of the involved side was poor. The clinical appearance of the rest of the dentition appeared normal. Haematological picture was within normal limits with elevated ESR.

A panoramic radiograph showed a large radiolucency with scooped-out effect in the mandible extending from the distal surface of the first primary molar to the distal surface of the permanent first molar involving the premolar germs (Figure 1). These teeth appeared 'floating in space'. There was no expansion of the buccal and lingual cortical plates. A complete skeletal survey revealed no involvement of the other bones. The

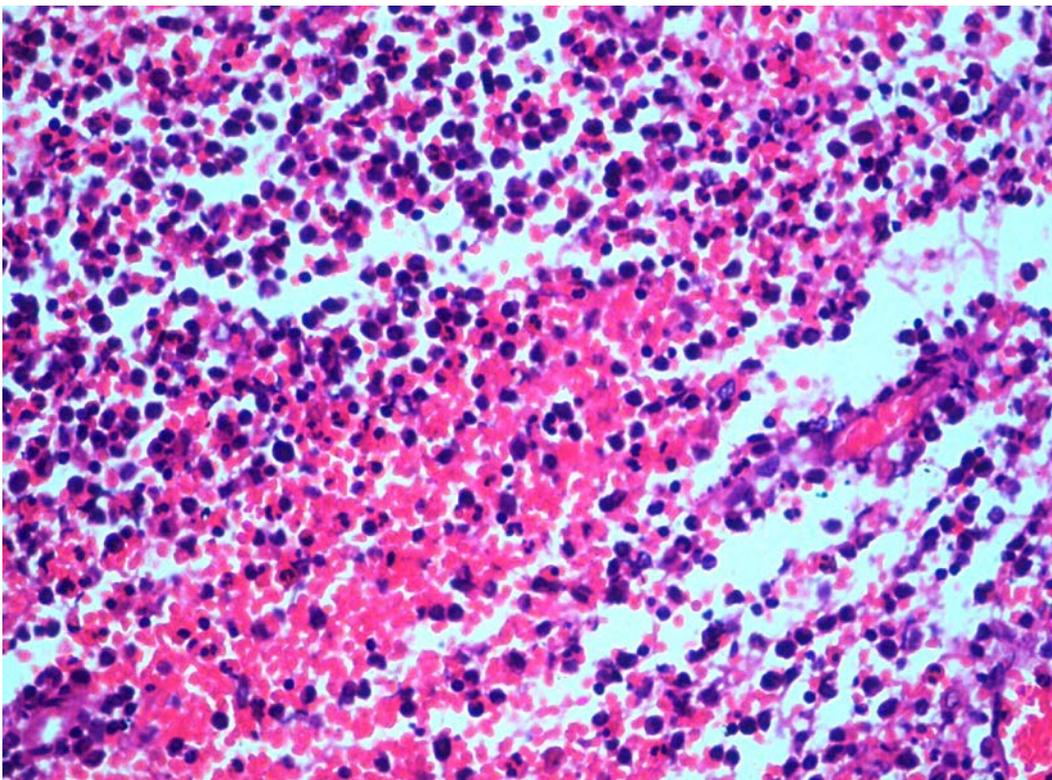
provisional diagnosis of eosinophilic granulomata was made based on the above mentioned findings.



**Figure 1.** Preoperative radiograph illustrating scooped out radiolucency in the involved side



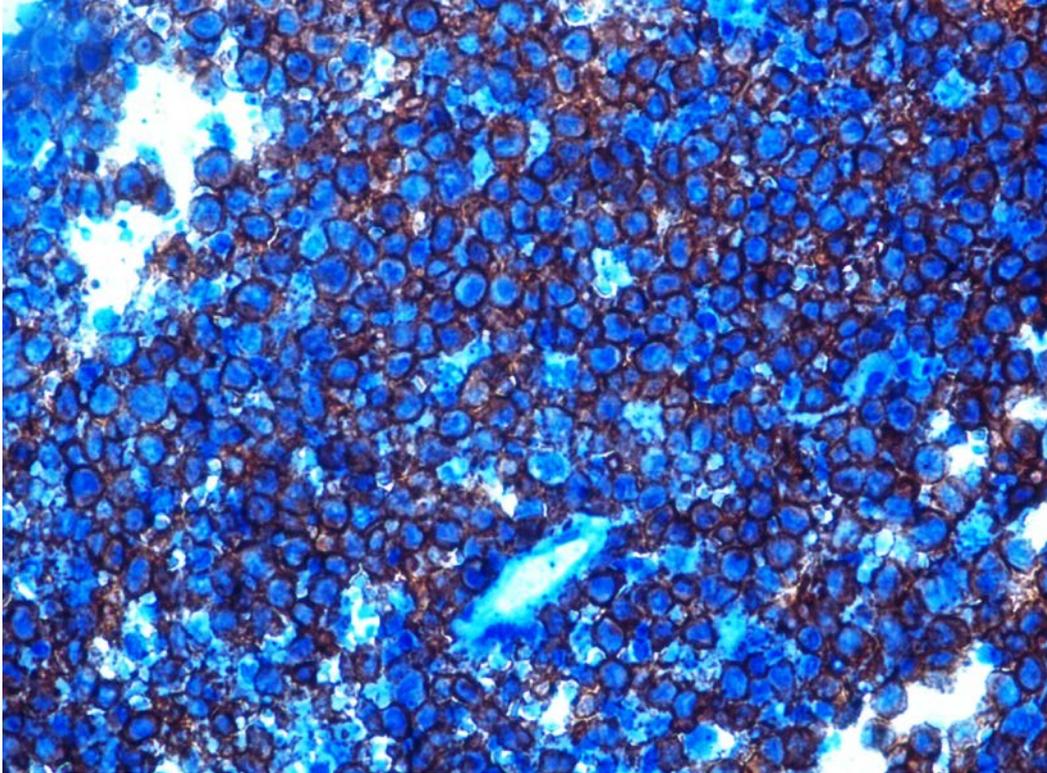
**Figure 2.** Extracted teeth and Bony tissues obtained after surgical curettage



**Figure 3.** Histopathological picture illustrating increased eosinophilic infiltration

Subsequently, under general anaesthesia an open biopsy was carried out with curettage of the abnormal bony tissue and extraction of four teeth (Figure 2), the primary first molar, primary second molar and the developing second premolar and permanent first molar. Histopathological examination revealed chronic inflammatory cells, a mass of histiocytic cells and numerous eosinophils suggestive of

Langerhans' cell histiocytosis (Figure 3). Immunohistochemistry was performed and the cells responded to CD1a surface antigen marker giving a confirmatory diagnosis of LCH (Figure 4). The patient responded well for chemotherapy and steroid therapy. Postoperative radiographic evaluation after 4 months showed good bone healing (Figure 5).



**Figure 4.** Immunohistochemistry positive for CD1a surface marker



**Figure 5.** post operative radiograph illustrating good bone healing

### 3. Discussion

Generally superior than adults, children possess a very good host immune response and when this itself turned against a child, makes the condition fatal if not intervened on time. Unifocal LCH, eosinophilic granuloma is a solitary lesion predominantly involves bone, typically the

skull, vertebrae, pelvis and the jaws, with posterior region of mandible as a common site of bone destruction. [4] Multifocal LCH infiltrate various organs, such as the bone, skin, liver, spleen, lung and brain [8].

LCH is a very rare disease in head and neck region as the etiology and pathogenesis still remain unclear. Varieties of etiological factors have been proposed including immunologic reactions, viruses, bacteria and

genetic influences. Possible development of LCH under the influence of colony stimulating factor (GM-CSF), interleukin-3 and tumor necrosis factor-alpha have also been suggested, and recently, cytogenetic studies have proposed the role of tumor suppressor genes (p53), oncogenes (c-myc, h-ras), growth factors, cell surface immunologic markers and apoptotic factors in LCH as well [9].

The Langerhans cells are immunocomponent cells originate from the bone marrow and then migrate into the epithelium to perform the function of antigen recognition and presentation. [10] The early diagnosis of unifocal LCH with mandibular lesion in this patient was facilitated by clinical and radiological evaluation. The patient's oral findings includes pain and mobility of the teeth with evident bone loss in the mandibular posterior region.

In this case the radiographical evaluation revealed a large radiolucency, well circumscribed and sharp unilocular image. Complete radiographic survey is required to substantiate the condition as unifocal or multifocal LCH. Dagenais et al have delineated seven different radiographic characteristics of the jaw lesions of Histiocytosis X, including solitary intraosseous lesions, a multiplicity of alveolar bone lesions, a well defined periphery, a scooped-out effect in the alveolar process, sclerosis in the alveolar bone lesions, periosteal new bone formation and slight root resorption [4].

The microscopical aspects of the lesion, which showed histiocytes, typical feature of LCH and immunohistochemical positivity to CD1a and S-100, confirmed the diagnosis. Immunohistochemistry has been employed as an ancillary tool for the confirmation of LCH, since the presence of Birbeck granules in proliferating cells helps to distinguish the latter from other histiocytic lesions [11].

Eosinophilic granulomata is usually misunderstood as periodontal disease. The pattern of bone loss is quite unlike the phenomenon of "floating teeth" which has been well-documented as occurring in histiocytosis X [12].

Rapidly progressive periodontitis has been claimed to be the form of periodontitis that is seen in individuals who have systemic diseases such as Down's syndrome, Chediak-Higashi syndrome, insulin-dependent diabetes and Papillon-Lefevre syndrome. [13] Microbiological evaluation provides an essential aid to differentiate between periodontal disease and eosinophilic granulomata. High proportions of Gram-negative anaerobic rods, including black pigmented Bacteroides, Fusobacterium, Capnocytophaga, spirochetes and motile rods are typical for a periodontal pocket flora in untreated periodontitis [14,15].

Surgical management of jaw lesions in the young patient is complicated by the presence of developing teeth which may easily be damaged or inadvertently removed. It has been stated that not all teeth involved by the disease require removal but teeth with marked mobility or with periapical lytic lesions should be sacrificed. [8] As in this case the primary first molar, primary second molar and the developing second premolar and permanent first molar teeth was extracted inevitably.

The management of LCH involves a wide spectrum of treatment modalities which includes surgical curettage together with radiotherapy and chemotherapy. [16] Surgical trauma and removal of the primary lesion may lead to an increased number of circulating lesion cells

which accelerates micrometastases. [17,18,19] So to avoid future recurrence and taking the young age of the child into consideration adjuvant chemotherapy was preferred with vinblastin and oral prednisolone.

## 4. Conclusion

The prognosis of patients with unifocal LCH, eosinophilic granulomata is excellent. Nevertheless, LCH can be unpredictable and it is mandatory that all cases are subject to a careful follow-up programme, so as to identify any signs of local recurrence or dissemination [7].

The aetiology of the disease should be better understood for definitive treatment programmes. The most beneficial factors towards patient care are early detection of the disease and appropriate referral for the treatment and follow-up measures.

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