

# Eosinophilic Granuloma of the Mandible

Akhilesh Prathap, Ravi Rajan Areekkal, Eapen Thomas, Nithin Pratap, Vinesh Udayakumar

Department of Oral and Maxillofacial Surgery, Pushpagiri College of Dental Sciences, Thiruvalla, Kerala, India

## Abstract

Eosinophilic granuloma (EG) is a rare histiocytic disorder resulting from clonal proliferation of Langerhans cells. Eosinophilic granuloma, the most benign of the three entities of Langerhans cell histiocytosis, may be multiple or solitary. This lesion can affect any bones but commonly involves the mandible when the jaws are affected. It is difficult to make a correct diagnosis on it without proof of a pathological diagnosis, which correlates with the diverse clinical and radiographic presentations of EG in the jaws. This report describes a case of unifocal EG of the mandible occurring in a 6-year-old boy whose initial presentation led to confusion of any solitary bony lesion. A final diagnosis of EG was established after histopathological examination of the biopsy specimen.

**Keywords:** Granuloma, Langerhans cells, monoclonal antibodies

## INTRODUCTION

Langerhans cell histiocytosis (LCH) refers to a relatively rare condition resulting from neoplastic proliferation of Langerhans cells. Eosinophilic granuloma (EG) – a term used synonymously with LCH by the World Health Organization – is a localized form of the disease.<sup>[1]</sup> Lichtenstein and Jaffe in 1940 introduced “EG of bone.”<sup>[2]</sup> It is one of the rarest bone tumors representing <1% of them. In 90% of the reported cases, it appears in children under the age of 10 years. It is, in fact, the mildest form of the histiocytosis-X group of diseases, which also encompass Hand–Schuller– Christian disease and Letterer–Siwe disease. The lesion shows a slight male predilection with male:female ratio 2:1.<sup>[3]</sup> EG affects almost any bones, but of facial bones, mandible is most commonly affected.<sup>[4]</sup>

EG may not present physical signs or symptoms in the clinical observation, and most of the time, it is discovered during routine radiographic examination. Loss of superficial alveolar bone and localized ulcerative lesions are common early forms of the disease.<sup>[5]</sup>

This report presents a case of EG of the mandible occurring in a 6-year-old boy who was admitted with complaints of a gradually increasing swelling on the left side of his face. Radiological investigations showed the presence of a lytic lesion in the mandible. Based on these findings, the

differential diagnoses considered were osteomyelitis and primary bone tumor. However, biopsy from the lesion showed the characteristic features of EG. We highlight this unusual case and emphasize the importance of histopathological examination in the diagnosis of this condition.

## CASE REPORT

A 6-year-old boy patient was referred to the outpatient department of our hospital with complaints of pain and progressively increasing swelling on the left side of the lower jaw. The swelling was noticed one month ago and he was taken to a nearby hospital. He was subsequently referred to us as there was no improvement in his signs and symptoms. A low-grade fever was associated with this right from the time of onset of the swelling; There is no history of trauma of weight loss. During this entire period, the patient had several trials of antimicrobials but without any effect.

**Address for correspondence:** Dr. Akhilesh Prathap,  
Department of Oral and Maxillofacial Surgery, Pushpagiri College of Dental  
Sciences, Medicity, Perumthuruthy, Thiruvalla - 689 107, Kerala, India.  
E-mail: dr.ravi.ra@gmail.com

**Received:** 05-08-2017

**Revised:** 05-09-2019

**Accepted:** 13-01-2020

**Published:** 08-06-2020

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

**For reprints contact:** WKHLRPMedknow\_reprints@wolterskluwer.com

**How to cite this article:** Prathap A, Areekkal RR, Thomas E, Pratap N, Udayakumar V. Eosinophilic granuloma of the mandible. *Ann Maxillofac Surg* 2020;10:254-7.

### Access this article online

#### Quick Response Code:



**Website:**  
www.amsjournal.com

**DOI:**  
10.4103/ams.ams\_139\_17

**Clinical examination revealed a single large**

Swelling measured approximately 5 cm × 4 cm located over the left lower jaw region with extension toward the angle of the mandible [Figure 1].

On palpation, the swelling had diffuse margins, was tender, immobile, and attached to the underlying structures [Figure 2].



Figure 1: Extraoral image

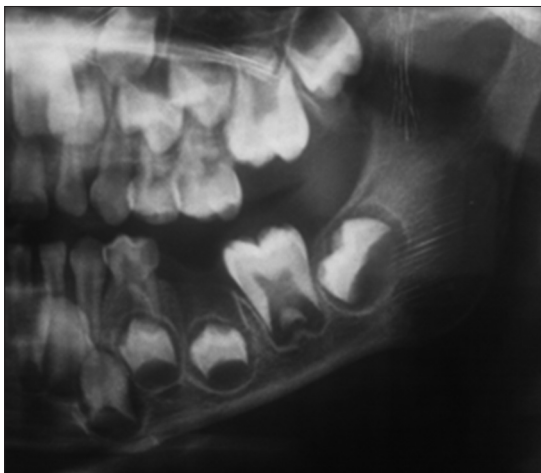


Figure 3: Orthopantomogram showing lesion

No regional lymphadenopathy or hepatosplenomegaly was observed.

Orthopantomogram showed an osteolytic lesion in the left ramus region that was unilocular in appearance. Axial cut and three-dimensional computed tomography were obtained to assess the extent of the lesion [Figures 3 and 4].

Fine-needle aspiration cytology of the lesion remained inconclusive as it showed only mixed inflammatory cells and macrophages. Hence, an excisional biopsy was performed under general anesthesia and the soft tissue and bone chips obtained at the biopsy were sent for histopathological



Figure 2: Intraoral image

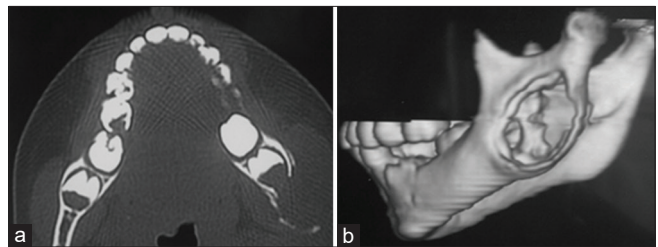


Figure 4: (a) Axial cut computed tomography scan showing bone expansion. (b) Three-dimensional computed tomography showing bone destruction



Figure 5: Intraoral surgical site

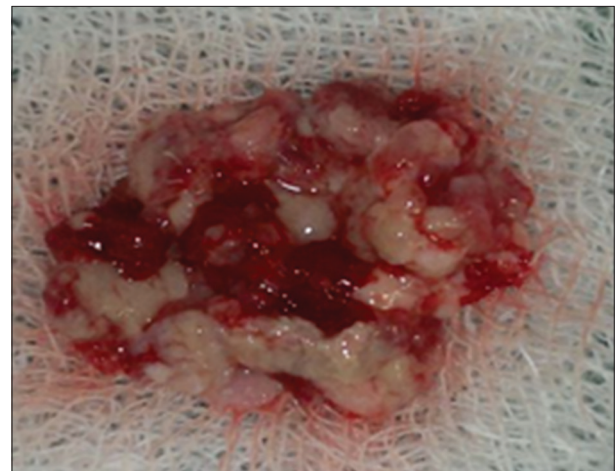
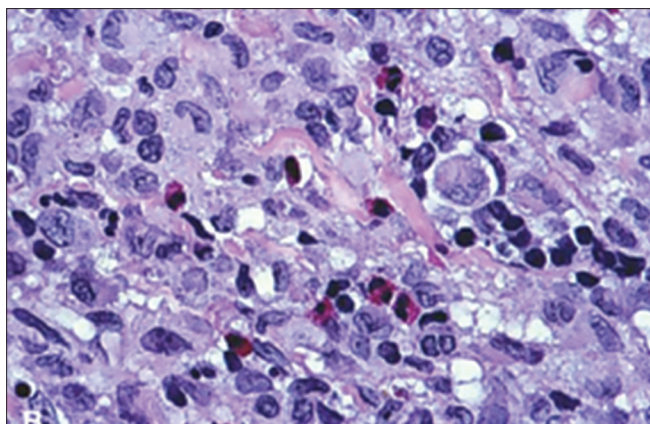


Figure 6: Excised lesion



**Figure 7:** Histopathological view of eosinophilic granuloma

evaluation [Figure 5].

On gross examination, small fragments of whitish soft tissue and bony chips were observed [Figure 6]. The histopathological slide showed features of typical eosinophilic granuloma [Figure 7]. The patient was discharged after an uneventful week of postoperative hospital stay [Figure 8]. A three year follow up showed no recurrence of the lesion after which the patient was lost to followup.

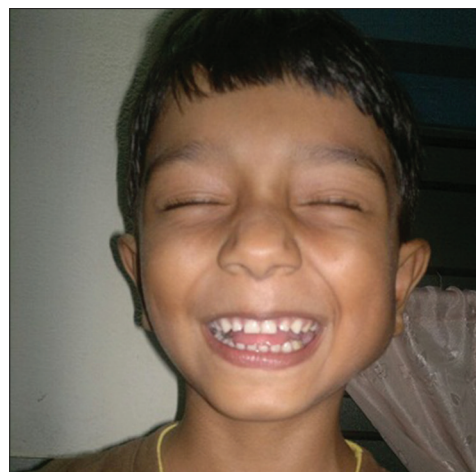
## DISCUSSION

The term “EG of bone” was first suggested by Lichtenstein and Jaffe.<sup>[2]</sup> LCH is a disease of unknown etiology that arises from clonal proliferation of Langerhans cells.<sup>[6]</sup> EG usually presented as a monostotic lesion affecting flat and long bones (70%), the skull bone, jaw bone, and the vertebral spine.<sup>[1]</sup> A variety of etiological factors have been proposed, including immunological reactions, viruses, bacteria, and genetic influences, but definitive evidence is still lacking. Some studies have suggested that the etiology may be related to immunological abnormalities resulting from a suppressor cell deficiency.<sup>[3,4]</sup>

Monostotic as well as polyostotic lesions are two forms in which EG of bone occurs, the former being more common. Bones of the axial skeleton, namely, calvaria (especially parietal bone), jaw bone, vertebral spine, and the pelvis, are usually affected.<sup>[1]</sup> On plain radiographs, EG typically presents as a punched out lesion with reactive sclerosis. A less common finding is one of the permeative patterns with or without periosteal reaction.<sup>[7]</sup>

The tumor material is sterile but there have been reports about the presence of staphylococcus and streptococcus.<sup>[8]</sup> Eosinophils, lymphocytes, fibroblasts, and foam cells may also be found but none of them is pathognomonic but they are suggestive of the diagnosis. This lesion when associated with Langerhans cells called as LCH.

On plain radiographs, jaw lesions have a unilocular radiolucent appearance with well-demarcated borders showing a characteristic scooped-out appearance. Tooth displacement



**Figure 8:** Postoperative view

is associated with about half of the bony lesions. Extensive alveolar involvement causes teeth to appear as if floating in air and root resorption is seen less frequently.<sup>[8]</sup>

Histologically, lesion is composed of Langerhans cell histiocytes, intermediate cells, and interdigitating cells of a dendritic cell lineage, T-cell lymphocytes, eosinophils, and macrophages. The hallmark cell is the Langerhans cell histiocyte. This cell has abundant eosinophilic to amphophilic cytoplasm and a nucleus that appears reniform, deeply indented, or grooved.<sup>[9]</sup>

EG does not lead to malignant transformation. If it extends beyond osseous tissue, it is then called Hand–Schuller–Christian disease and may manifest as diabetes insipidus, cerebellar, hypothalamic, and with other central nervous system symptoms. The prognosis depends on the age of diagnosis and the number of foci.

In general, no treatment is needed for localized osseous EG and often the biopsy is enough to initiate healing. Steroid injection, curettage, excision, or radiation may be necessary depending on the extent of the disease and the symptoms.<sup>[5]</sup> The prognosis is good for localized disease and malignant transformation has not been observed.<sup>[10]</sup> In rare instances, solitary disease may recur rapidly and result in death.<sup>[11]</sup> Plasschaert *et al.*<sup>[12]</sup> found that adults with EG have a higher chance of recurrence compared with children.

## CONCLUSION

As the course of the disease can be unpredictable at times, the potential for unifocal disease to become multifocal should not be underestimated, and therefore, long-term follow-up is mandatory.<sup>[13]</sup> Monoclonal antibodies directed against CD1a or CD207 may evolve as one of the potential treatment modalities in the future.<sup>[14]</sup>

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other

clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

### REFERENCES

1. Young DB, Unni KK. Langerhans cell histiocytosis. In: Fletcher CD, Unni KK, Mertens F, editors. World Health Organization Classification of Tumours. Pathology and Genetics of Soft Tissue and Bone. Lyon: IARC Press; 2002. p. 345-6.
2. Lichtenstein L, Jeffe HL. Eosinophilic granuloma of bone. With report of a case. *Am J Pathol* 1940;16:595-604.
3. Ando A, Hatori M, Hosaka M, Hagiwara Y, Kita A, Itoi E. Eosinophilic granuloma arising from the pelvis in children: A report of three cases. *Ups J Med Sci* 2008;113:209-16.
4. Sai S, Fujii K, Masui F, Kida Y. Solitary eosinophilic granuloma of the sternum. *J Orthop Sci* 2005;10:108-11.
5. Agarwal A, Agrawal GP, Alam S, Husain B. A case of unifocal eosinophilic granuloma of the mandible in an adult female: A case report. *Case Rep Dent* 2012;2012:1-3.
6. Sherwani RK, Akhtar K, Qadri S, Ray PS. Eosinophilic granuloma of the mandible: A diagnostic dilemma. *BMJ Case Rep* 2014;2014. pii: bcr2013200274.
7. Wood NK, Goaz PW. Multiple Separate well Defined Radiolucencies. Ch. 22. Wood and Goaz: 5<sup>th</sup> Edition; 1997. p. 386-9.
8. Fowles JV, Bobechko WP. Solitary eosinophilic granuloma in bone. *J Bone Joint Surg B* 1970;52:238-43.
9. Rajendaran R, Ahamad M. Shafer's Textbook of Oral Pathology. Disease of Bone and Joints. Ch 17. 8<sup>th</sup> ed: Shaefer; 2016. p. 749-53.
10. Watzke IM, Millesi W, Kermer C, Gisslinger H. Multifocal eosinophilic granuloma of the jaw: Long-term follow-up of a novel intraosseous corticoid treatment for recalcitrant lesions. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2000;90:317-22.
11. Ramani P, Chandrasekar T, Baig MF, Muthusekar MR, Thomas S, Manyam R, *et al.* Langerhans cell histiocytosis of the mandible in a six-year-old child. *Indian J Dermatol Venereol Leprol* 2007;73:114-6.
12. Plasschaert F, Craig C, Bell R, Cole WG, Wunder JS, Alman BA. Eosinophilic granuloma. A different behaviour in children than in adults. *J Bone Joint Surg Br* 2002;84:870-2.
13. Dewan M, Al-Ghamdi AA, Zahrani MB. Lessons to be learned – Langerhans' cell histiocytosis. *J R Soc Promot Health* 2008;128:41-6.
14. Hicks J, Flaitz CM. Langerhans cell histiocytosis: Current insights in a molecular age with emphasis on clinical oral and maxillofacial pathology practice. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2005;100:S42-66.