

# Solitary Extragnathic Langerhans Cell Histiocytosis – A Rare Case

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## ABSTRACT

Langerhans cell histiocytosis (LCH), mainly affects the skull, vertebrae, ribs and mandible in children and the long bones of adults. Symptoms range from none to pain, swelling and tenderness over the site of the lesion. This disease presents oral manifestations which can sometimes be the first expression of the condition. It occurs in three forms namely eosinophilic granuloma in which isolated or multiple bones are involved, But has a good prognosis whereas other variants Hand-Shuller-Christian disease (chronic dessiminated variant) and Letterer-Siwe disease (acute dessiminated form) have poor prognosis. Occasionally only soft tissues are affected without bony involvement. Males are more commonly affected than females. This article describes a rare variant of eosinophilic granuloma of labial mucosa without bony involvement.

**Keywords:** Birbeck granules, Eosinophilic granuloma, Histiocytes, Langerhans cells, Ulcer

## CASE REPORT

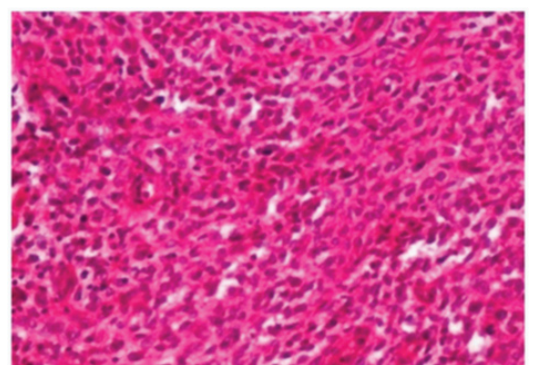
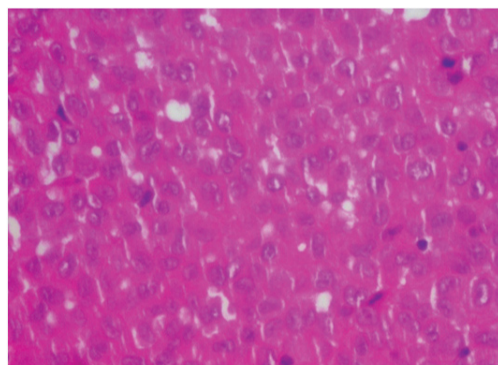
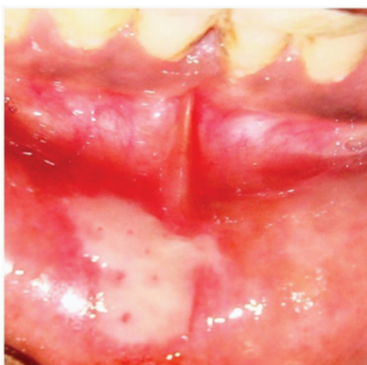
A 50-year-old male patient reported to the Department of Oral Medicine and Radiology of SVS Institute of Dental Sciences, Mahbubnagar with a chief complaint of painful ulcer in the labial sulcus of mandible with a duration of two months. On examination, the ulcer was measuring about 0.8x1 cm approximately. The floor of ulcer was granulomatous with irregular margins and undermined edges [Table/Fig-1]. No other abnormality was detected on general and oral examination. Clinically, the patient was otherwise normal. Incisional biopsy was done and the specimen was sent to our department. H & E stained sections revealed an admixture of cell population comprising of histiocytes containing abundant eosinophilic cytoplasm and eosinophils with bilobed, spectacle shaped nuclei. Cells with relatively pale cytoplasm containing vesicular nuclei with indentations imparting coffee-bean shaped appearance were also seen which are presumably Langerhans cells [Table/Fig-2]. This group of cells comprised a majority of cell population along with plasma cells and lymphocytes. Numerous blood vessels lined by plump endothelial cells and extravasated RBCs were also noted. The intervening delicate connective tissue showed haphazardly arranged collagen fibers with few fibroblasts and fibrocytes [Table/Fig-3-5]. Keeping in view the above findings, a diagnosis of eosinophilic granuloma was given. Subsequently, cephalograms and other radiographs were advised which did not reveal any skeletal involvement nor CT scan of the body revealed any involvement of systemic organs. In view of all the above findings, a final diagnosis of solitary extragnathic Langerhans cell histiocytosis

was arrived at and the patient was referred to Department of Oral and Maxillofacial Surgery for surgical excision.

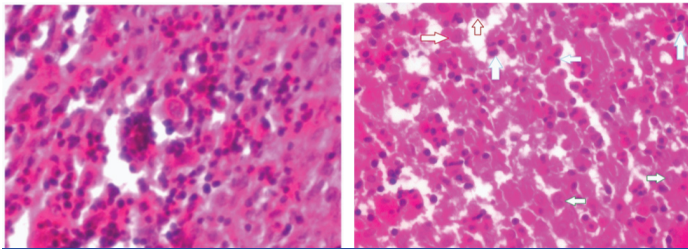
## DISCUSSION

Langerhans cell histiocytosis (LCH) is a group of disorders containing three clinical variants: eosinophilic granuloma, Hand-Shuller-Christian disease and Letterer-Siwe disease [1]. Though the latter two are more generalized and serious forms, eosinophilic granuloma is localized form of LCH generally presenting as a monostotic lesion affecting any of the flat or long bones [2]. Soft tissue involvement might occur affecting mainly the lymph nodes, lungs and mucous membranes subsequently. However, on rare occasions, only soft tissue involvement mostly presenting as palatal or gingival ulcerations without the involvement of underlying bone have been reported [3,4]. To the best of our knowledge, only six cases have been reported where they presented as ulceration in the soft tissues overlying bone [5]. But the present case is quite different from those reported and is probably the first of its kind where it has occurred in the lip, a soft tissue that has no bone within it. Secondly, 90% of cases reported are in children below the age of 10 years [2] and in the present case, the patient is an elderly individual in the sixth decade.

Clinically, a variety of lesions may present as an ulcer. But careful biopsy and histological examination may help us in their accurate diagnosis. In the present case histologically it was observed that there was an inflammatory process with presence of abundant proliferation of eosinophils, histiocytes and Langerhans cells. On



**[Table/Fig-1]:** Granulomatous ulcer in labial sulcus **[Table/Fig-2]:** Sheets of Langerhans cells with coffee bean shaped nucleus (40x) **[Table/Fig-3]:** Sheets of inflammatory cells with predominantly delicate CT background (10x)



**[Table/Fig-4]:** Admixture of cell population comprising à Histiocytes + Eosinophils (20x)  
**[Table/Fig-5]:** In addition to chronic inflammatory cell population à Cells with relatively pale cytoplasm containing vesicular nuclei with indentations imparting "coffee-bean" shaped appearance à Langerhans' cells (green arrow), histiocytes (red arrow), eosinophils (blue arrow) (40x)

extensive search of literature for lesions containing eosinophils and histiocytes, a variety of lesions such as eosinophilic ulcer of oral mucosa, atypical histiocytic granuloma (AHG), angiolymphoid hyperplasia with eosinophilia (ALHE), Kimura disease and LCH particularly eosinophilic granuloma were considered [6,7].

The presence of Langerhans cells with characteristic cleaved or coffee bean shaped nuclei is hallmark of LCH which was evident in our case. However, positivity with CD1a and S100 protein would have helped in confirming the diagnosis [8,9]. However, no such definitive diagnosis could be arrived at due to inadequacy of the tissue and the patient's refusal for further investigations, a detailed follow up could not be carried out.

In our case, a diagnosis of eosinophilic granuloma was made at by excluding Hand-Schuller-Christian disease which shows a classical triad of punched out bone destructions in skull, uni/bilateral exophthalmos and diabetes insipidus with or without manifestations of dyspituitarism such as polyurea, dwarfism/infantilism. Letterer-Siwe disease was also excluded as it is an acute fulminating disorder which invariably occurs in infants usually before the age of three years along with diffuse involvement of visceral organs [10].

The patient was referred to Department of Oral and Maxillofacial Surgery for surgical excision of the lesion. In case of LCH, the prognosis largely depends on the involvement of multiple organ systems. However, eosinophilic granuloma is a chronic localized

form, the prognosis is good. It has also been suggested that the patients must be evaluated regularly during the first six months for any bone or visceral involvement and that long term follow up is important to rule out the possibility of recurrence [11,12].

## CONCLUSION

Keeping in view the clinical manifestation of present case, though it is tempting us to suggest that LCH should also be included in the differential diagnosis of long standing, non-healing ulcers like non-healing diabetic ulcers, chronic ulcers due to trauma, necrotizing sialometaplasia, tuberculosis, deep fungal infections or ulcers in carcinomas occurring in oral cavity, it is difficult to decisively comment on the same till few more cases with similar features are reported in future.

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**FINANCIAL OR OTHER COMPETING INTERESTS:** None.

Date of Submission: **Dec 19, 2014**  
 Date of Peer Review: **Feb 04, 2015**  
 Date of Acceptance: **Feb 18, 2015**  
 Date of Publishing: **Mar 01, 2015**