International Surgery Journal
Anehosur V et al. Int Surg J. 2026 May;13(5):868-872
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902
DOI: https://dx.doi.org/10.18203/2349-2902.isj20261191

Case Report

Bilateral synchronous presentation of Langerhans cell histiocytosis of maxilla and mandible: a rare case report and literature review

Venkatesh Anehosur[1]*, Akshay A. Byadgi[1], Sudeep Narayana B.[1], Parinitha S. Sangam[2], Rani Hephzibah[2]

[1]Department of Oral and Maxillofacial Surgery, SDM Craniofacial Research centre, SDM College of Dental Sciences and Hospital, A Constituent Unit of Shri Dharmasthala Manjunatheshwara University, Dharwad, Karnataka, India
[2]Department of Pathology, SDM College of Medical Sciences and Hospital, A Constituent Unit of Shri Dharmasthala Manjunatheshwara University, Dharwad, Karnataka, India

Received: 17 March 2026
Accepted: 17 April 2026

***Correspondence:**
Dr. Venkatesh Anehosur,
E-mail: venkyrao12@yahoo.co.in

ABSTRACT

Previously known as histiocytosis X, Langerhans cell histiocytosis (LCH) is a rare haematological condition that primarily affects new-borns and young children. The uncontrolled activation and proliferation of normal antigen-presenting cells, specifically Langerhans cells, are the defining features of this condition. A positive immunohistochemistry result for CD1a/CD207 and S100 is the gold standard for a conclusive diagnosis. There are various therapy options available for individuals with LCH. Due to its comparatively low occurrence, little information is known about the epidemiology of LCH; cases are estimated to occur 3-5 times annually per million population. In this report, we have highlighted a case of 35-year-old adult who reported to the department with Bilateral synchronous gingival enlargements in maxilla and mandible. On clinical, radiological, histopathological (Incisional biopsy) and immunohistochemical examinations revealed a diagnosis of LCH. even though LCH incidence in adults is uncommon, LCH must be taken into account when evaluating multiple osteolytic bony jaw lesions in young adults that have an unclear aetiology. So here we present a rare case of Bilateral synchronous presentation of LCH of maxilla and mandible in adult with its management.

Keywords: Langerhans cell histiocytosis, Immunohistochemistry test, Antigen presenting cells

INTRODUCTION

Langerhans cell histiocytosis (LCH), previously known as histiocytosis X, is a rare haematological condition that primarily affects new-borns and young children. LCH is characterized by the proliferation of abnormal Langerhans cells, which are specialized dendritic cells responsible for antigen presentation. Due to its rarity, there is limited data available on the disease.[1] LCH is more commonly diagnosed in male children and usually considered to be a disease of childhood, with a peak incidence from 1 to 3 years.[1,2] The disease presents a wide clinical spectrum, ranging from isolated lesions in a single organ to multisystemic involvement, which can be life-threatening.[3] Despite advances in understanding LCH's pathophysiology, the aetiology remains unclear, and the disease is often categorized as both inflammatory and neoplastic. The role of the MAPK/ERK pathway, specifically BRAF mutations, has been highlighted in recent years, providing potential avenues for targeted cell therapy.[4] LCH commonly involves the head and neck, especially the bones of the skull and jaws, with the gingiva and hard palate being the most frequently affected. Clinical features of LCH vary; they can present as a single lytic lesion of bone with a favourable prognosis or as a more severe, disseminated form resembling leukaemia.[4] Depending on the organs involved, LCH has been categorized into a localized (single-system disease) and a disseminated form (multisystem disease). The pattern of organ involvement

differs significantly between patients suffering from single-system disease and multisystem disease. Single-system disease accounts for approximately one-third of patients. Up to 20-30% of adult patients with LCH present with isolated pulmonary involvement.[5]

This case report highlights Bilateral synchronous presentation of LCH of maxilla and mandible in adult with its clinical presentation, diagnostic approach, and treatment course. And also emphasized on importance of early diagnosis, individualized treatment, and long-term follow-up to improve patient outcomes.

CASE REPORT

A 35-year-old male patient was referred to our tertiary care centre for an opinion regarding unresolved non healing irregular gingival growth in upper and lower jaw since last 6 months. Clinical examination revealed history of pain which was intermittent, dull aching, non-radiating, progressive, nocturnal type which aggravated on having spicy food with Burning sensation. Patient was on medications for gastroesophageal reflex disease since the past 10 years (Medication unknown). History of betel quid chewing present once in 2 days for 6-7 years. extraction history noted in relation to mandibular left first molar due to periodontal issues and extraction was uneventfull.

Extra orally no abnormalities detected added to that no lesions noted over the other parts of the body. No cervical lymphadenopathy noted bilaterally. On intra oral examination, multiple ulcerations noted over maxilla and mandible. Ulcerative lesion noted in relation to the extracted mandible alveolar socket region measuring approximately 2×2 cm extending over the buccal sulcus region and lingual extension noted. Erythematous mucosa noted over the palatal aspect in relation to maxillary right molars, measuring approximately 1.5×1 cm, and maxillary left molar area, measuring approximately 2×1 cm. The lesion was tender on palpation and rough in consistency. No active bleed/bleed on palpation was noted. Grade II mobility teeth noted with maxillary first molars (Figure 1).

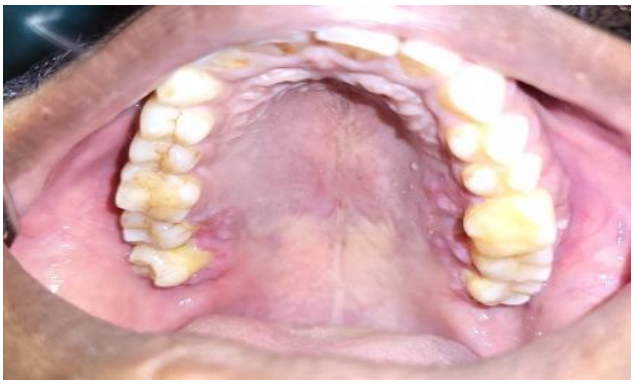

Figure 1: Clinical presentation of LCH on both right and left posterior maxilla showing diffused lesions.

Routine orthopantomography (OPG) radiograph, and routine blood investigations were found to be in normal range and tested negative for HIV, HBsAg, and HCV. After signing the written consent form, an incisional biopsy was performed from multiple sites intraorally and sent for histopathological examination. Microscopic examination revealed that the cells display elongated bland nucleus with longitudinal grooves, inconspicuous nucleoli and abundant pale eosinophilic cytoplasm. Few cells show multinucleation were present. Eosinophilic infiltration noted amidst these cells with the formation of eosinophilic abscesses focally. Multiple sections from excised lesion showed Sheets of large, round to oval cells with nuclear grooves (coffee bean nuclei), abundant eosinophilic cytoplasm, minimal nuclear atypia, admixed with eosinophils. Cells were seen infiltrating the deeper stroma (Figure 2).

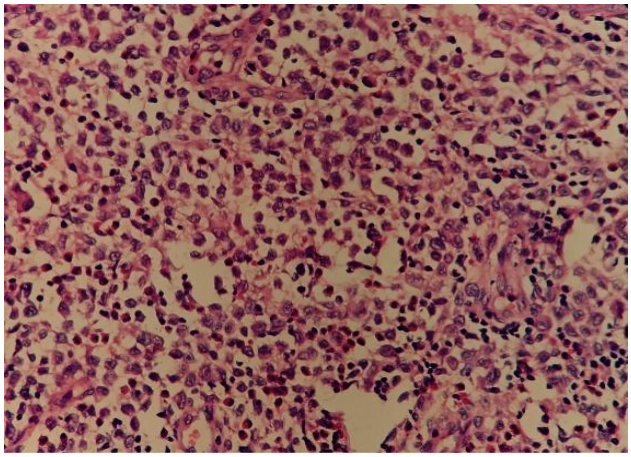

Figure 2: Microscopy showing sheets of large, round to oval cells with nuclear grooves abundant eosinophilic cytoplasm, minimal nuclear atypia admixed with eosinophils (×400, H and E stain).

Based on the histopathological findings, it was diagnosed that the patient has a condition called “LCH” (Note: multiple lesions suggestive of Hand-Schuller-Christian disease). Later on, patient was suggested for immunohistochemistry (CD1a and S100) for confirmation, which was found to be positive for both markers. Therefore, the immunohistochemical profile of the patient with mandibular lesion confirmed the diagnosis of LCH.

Meanwhile patient underwent full volume CBCT for further evaluation. CBCT suggested that, a well-defined radiolucency extending from the distal aspect of the left mandibular first molar to the mesial aspect of the left mandibular second molar (scooped out appearance), buccal, lingual cortices and alveolar crest destruction noted, loss of superior cortex, lamina propria and the absence of reactive changes in the surrounding mandibular bone structure noted. Significant thickening of the mucosa in both maxillary sinuses was noted. Substantial loss of the posterior aspect of the maxillary

sinus floor on the left side, significant damage of the alveolar process in the molar region on the left side (extending to the maxillary second molar) (Figures 3 and 4).

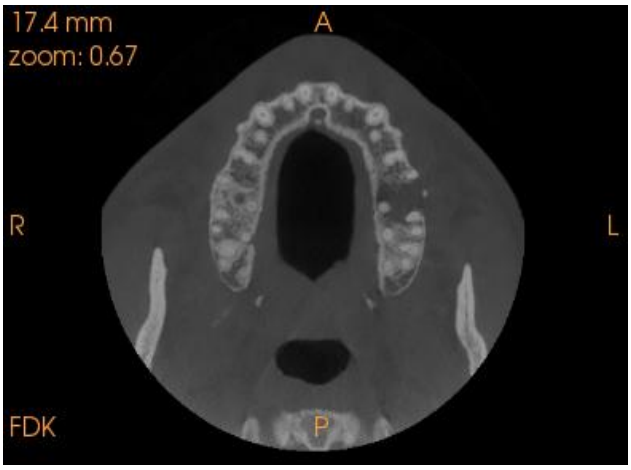

Figure 3: Osteolytic lesion in right and left posterior aspect of maxilla.

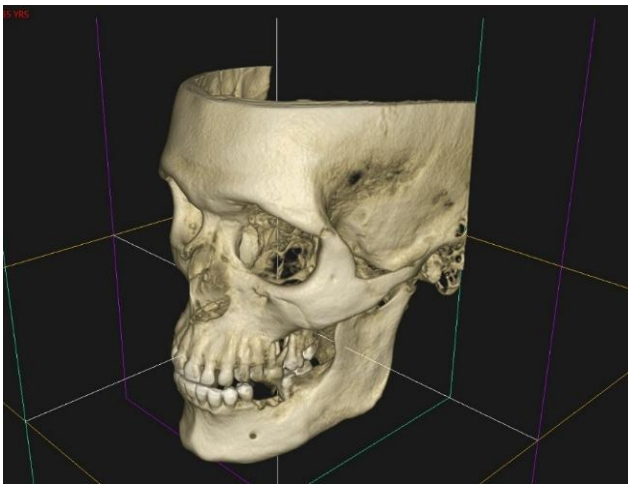

Figure 4: Bony erosion noted in the left manibular region.

For further treatment planning and the patient was advised to undergo a PET-CT scan which suggested that in the head and neck, focal increased tracer uptake is noted in the subtle hypodense lytic lesion in the left upper alveolus in the mid part (maxilla on left side), SUV max 8.1. No focal FDG avid areas noted in the rest of the maxilla and mandible, with evidence of subtle hypodensity in the left mandible. No discrete enhancing lesion or focal abnormal FDG uptake seen in paranasal sinus/oropharynx, nasopharynx, hypopharynx, brain, thorax, abdomen pelvis and rest of the body.

After all the routine investigations, the patient was planned for surgical intervention under general anaesthesia. Wide excision of the lesion with 1 cm clearance and closure done using local buccal pad of fat on right and left posterior maxilla, and buccal advancement flap on mandibular region (Figures 5). The patient recovered well during the post-operative period at hospital for 3 days and was discharged. During which patient received IV Antibiotics of Amoxycillin with clavulanic acid (1.2 gm BD) and INJ Metronidazole 500 mg TID. Further which patient regular follow-up was done, healing was satisfactory with no evidence of recurrence at Six months (Figure 6).

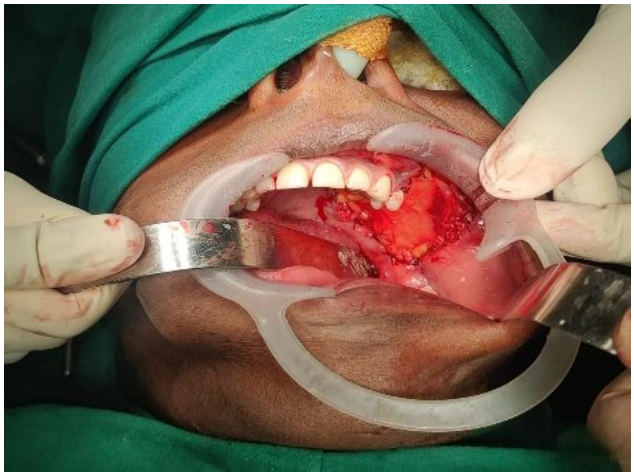

Figure 5: Intraoperative wide excision of the lesion and reconstruction with buccal pad of fat.

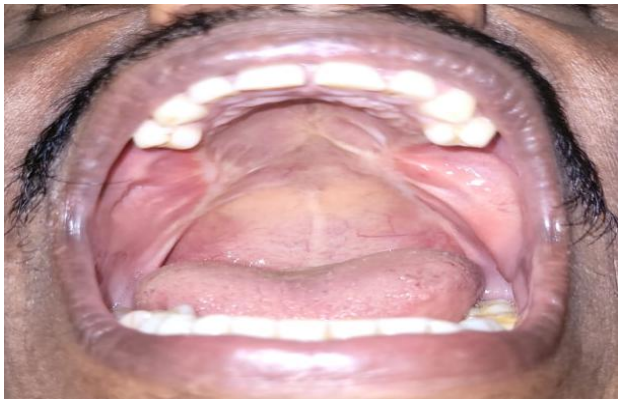

Figure 6: Healing after 6 months follow-up.

DISCUSSION

LCH, formerly known as histiocytosis X (HX), refers to a group of hyperplastic cellular disorders with an unknown origin. In 1987, the Histiocyte Society officially adopted the name LCH.[6,7] LCH is currently classified into three clinical subtypes using a widely accepted classification system: acute disseminated LCH, chronic multifocal LCH, and chronic focal LCH. However, an alternative and more commonly used classification categorizes LCH into two main forms localized and disseminated based on the extent of lesions: Localized (LCH) involves a single skin lesion without organ involvement. Presents as isolated bone lesions, which may occur with or without diabetes insipidus (DI), regional lymph node

enlargement, or skin rash. Includes multiple bone lesions, affecting more than one bone or multiple sites within a single bone, potentially accompanied by DI, nearby lymph node involvement, or rash. Disseminated (LCH) are characterized by involvement of internal organs, with or without DI, regional lymphadenopathy, or rash, but without functional impairment of the lungs, liver, or hematopoietic system. A more severe form includes internal organ involvement along with dysfunction of vital systems such as the lungs, liver, or bone marrow.[8]

LCH can occur at any age, though it most commonly affects children between the ages of 1-3. In adults, the incidence is relatively rare, estimated at 1-2 cases per million individuals. The disease exhibits a male predominance, with a reported male-to-female ratio of approximately 2:1. Despite ongoing research, the exact cause of LCH remains unclear. However, advances in molecular biology and genetics, along with more in-depth clinical studies, have led to a better understanding of its nature. LCH is now considered a reactive hyperplastic disorder, with its pathogenesis closely linked to chromosomal instability and specific gene mutations. The presence of clonal proliferation of affected cells supports the view that LCH originates from a neoplastic process.[9]

The diagnosis of LCH depends on clinical manifestation, iconography examination, lesion histopathology and electron microscopy. Pathological examination includes print and pathological biopsy and immunohistochemical examination. Pathological materials can be taken from the rash, lymph nodes, the tumour or the local lesions. The nucleus was folded within a groove, making it looks like coffee beans. Eosinophils and other inflammatory cells was sporadic round in shape. The gold standard of diagnose is to find Birbeck particle by electron microscopy.[10]

The most common clinical manifestation involves solitary or multiple bone lesions frequently involving the skull, ribs, vertebrae, and jaws. Skull and femoral lesions are often found in children younger than age 10, and patients older than age 20 more often have lesions in the ribs, shoulder girdle, and mandible. The most common presentation of oral signs and symptoms are intraoral mass, pain, gingivitis, loose teeth, mucosal ulcer, impaired healing, and halitosis, of which few were noted in this case. As oral lesions in LCH are often intraosseous and can resemble various other clinical conditions, a thorough differential diagnosis is essential. Conditions to consider include giant cell granuloma, granulomatous diseases, osteomyelitis, and odontogenic tumours.[11,12]

Regarding the location, the majority of the cases of LCH usually affect the posterior jaw regions, while anterior regions are less affected.[13] In this case report, the posterior hard palate region was also predominantly affected. In a case series presented by Eckardt et al 10 patients with LCH, seven had LCH manifestations in the maxillofacial region, four of which had only a single oral lesion; predominant maxillofacial lesions were located in the posterior mandible.[14] Oral soft tissue lesions are most commonly found in gingiva and hard palate. The floor of the mouth, maxillary sinus, and buccal mucosa each account for less than 10% of lesions of LCH. Radiographically, advanced and progressive alveolar bone destruction in LCH often results in the characteristic "floating teeth" appearance.[15] However, this feature was observed in this report over the left upper molar region.

Treatment of LCH remains controversial due to high variation in clinical features and the absence of standard diagnostic and evaluation criteria. Surgical curettage is commonly mentioned as the elective treatment for individual bone lesions in the jaws. However, chemotherapy or radiation therapy are advised if surgery cannot reach the location.[15,16] Intralesional corticosteroid agents may be effective in some patients with localized lesions (e.g., prednisolone 20-30 mg/day for 2-4 weeks and then followed by tapering of the dose). Multisystemic disease needs systemic chemotherapy. The most common agents used in different combination regimens and several cycles are corticosteroids, vinblastine, etoposide, cytarabine, 6-mercapto purine, methotrexate, 2-chlorodeoxyadenosine, cyclosporine, thalidomide and others. A combination of vincristine and prednisone seems to reduce the risk of recurrence.[17] Since it was isolated to the oral cavity, in this case report we have performed surgical resection with 1cm clear margins and curettage followed by closure using the local flaps.

For LCH, the prognosis is generally favourable. However, the prognosis is poor for young patients (those aged 2 or under) who have organ dysfunction due to a multisystem disease. According to a study done by Baptista AM and Quraishi et al the survival rate for unifocal and multifocal bone involvement was 100% at a median 5-year follow-up, whereas the survival rate for systemic illness was 76.5%. It is crucial to note that this illness has several ramifications that could seriously lower patients' quality of life.[18,19]

CONCLUSION

This case report is being reported as an uncommon and rare in terms of age and condition. The diagnosis of LCH is primarily aided by oral symptoms following which incisional biopsy and definitive immunohistochemistry markers to be done to mark a early diagnosis. These oral symptoms could easily be confused with common dental conditions as periapical and periodontal illnesses. Hence the oral cavity symptoms could be a sign of systemic disorders, the dental surgeon must closely monitor, diagnose, and look into common oral abnormalities to rule out underlying severe systemic involvement like LCH.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Bhargava D, Bhargava K, Hazarey V, Ganvir SM. Hand-Schüller-Christian disease. Indian J Dental Res. 2012;23(6):830-2.
2. Scolozzi P, Lombardi T, Monnier P, Jaques B. Multisystem Langerhans' cell histiocytosis (Hand-Schüller-Christian disease) in an adult: a case report and review of the literature. Europ Arch Oto-Rhino-Laryngol Head Neck. 2004;261:326-30.
3. DM Howarth. Langerhans cell histiocytosis diagnosis, natural history, management, and outcome. Cancer. 1999;85:2278-90.
4. Allen CE, Merad M, McClain KL. Langerhans-cell histiocytosis. N Eng J Med. 2018;379(9):856-68.
5. Stockschlaeder M, Sucker C. Adult Langerhans cell histiocytosis. Europ J Haematol. 2006;76(5):363-8.
6. Lian C, Lu Y, Shen S. Langerhans cell histiocytosis in adults: a case report and review of the literature. Oncotarget. 2016;7(14):18678.
7. Madrigal-Martínez-Pereda C, Guerrero-Rodríguez V, GuisadoMoya B, Meniz-García C. Langerhans cell histiocytosis: literature review and descriptive analysis of oral manifestations. Med Oral Patol Oral Cir Bucal. 2009;14:222-8.
8. Chu T. Langerhans cell histiocytosis. Aust J Dermatol. 2001;42(4):237-42.
9. Campanati A, Simonetti O, Marconi B, Giuliodori K, Ganzetti G, Brandozzi G, et al. Purely cutaneous Langerhans' cell histiocytosis in an adult woman. Acta Derm Venereol. 2009;89(3):299-301.
10. Zhinan Zhang, Ti Shen. Diagnostic and curative standard of blood disease. Edition 3. Beijing: Science Publishing Company. 2007;7:248-53.
11. Neves-Silva R, Fernandes DT, Fonseca FP, Rebelo Pontes HA, Brasileiro BF, Santos-Silva AR, et al. Oral manifestations of Langerhans cell histiocytosis: A case series. Spec Care Dentist. 2018;38(6):426-33.
12. 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.
13. Annibali S, Cristalli MP, Solidani M, Ciavarella D. Langerhans cell histiocytosis: oral/periodontal involvement in adult patients. Oral Dis. 2009;15:596-601.
14. Eckardt A, Schultze A. Maxillofacial manifestations of Langerhans cell histiocytosis: a clinical and therapeutic analysis of 10 patients. Oral Oncol. 2003;39:687-94.
15. Bartnick A, Friedrich RE, Roeser K, Schmelzle R. Oral Langerhans cell histiocytosis. J Craniomaxillofac Surg. 2002;30:91-6.
16. Hartman KS. Histiocytosis X: a review of 114 cases with oral involvement. Oral Surg Oral Med Oral Pathol. 1980;49:38-54.
17. Rao DG, Trivedi MV, Havale R, Shrutha SP. A rare and unusual case report of Langerhans cell histiocytosis. J Oral Maxillofac Pathol. 2017;21(1):140-4.
18. Baptista AM, Camargo AF, de Camargo OP, Odone Filho V, Cassone AE. Does adjunctive chemotherapy reduce remission rates compared to cortisone alone in unifocal or multifocal histiocytosis of bone. Clin Orthop Relat Res. 2012;470:663-9.
19. Quraishi MS, Blayney AW, Breatnach F. Aural symptoms as primary presentation of Langerhan's cell histiocytosis. Clin Otolaryngol Allied Sci. 1993;18:317-23.

Cite this article as: Anehosur V, Byadgi AA, Sudeep NB, Sangam PS, Hephzibah R. Bilateral synchronous presentation of Langerhans cell histiocytosis of maxilla and mandible: a rare case report and literature review. Int Surg J 2026;13:868-72.