CASE REPORT

AN UNUSUAL PRESENTATION OF ORAL LANGERHANS CELL HISTIOCYTOSIS AS A PERSISTENT ULCER WITH ALVEOLAR BONE LOSS IN A YOUNG CHILD: A CASE REPORT

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ABSTRACT

Background: Langerhans Cell Histiocytosis (LCH) is a rare clonal proliferative disorder of dendritic cells that predominantly affects children and commonly involves bone. Oral manifestations may be the first or only sign of disease and often resemble common inflammatory or traumatic lesions, leading to diagnostic delay.

Case Report: A 3-year-old male child presented with a persistent ulcer on the lingual alveolar mucosa in relation to the lower left primary molars, associated with localized alveolar bone destruction on radiographic examination. Clinical suspicion of an underlying systemic pathology prompted histopathological and immunohistochemical evaluation, which confirmed the diagnosis of LCH. Multidisciplinary management was initiated, and the child was kept under regular follow-up.

Conclusion: This case underlines the importance of recognizing atypical oral ulcers with underlying bone loss in children as potential manifestations of LCH and highlights the need for early diagnosis and long-term surveillance.

KEY WORDS

Langerhans Cell Histiocytosis; Paediatric oral lesion; Alveolar bone loss; Non healing oral ulcer; Mandibular involvement.

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1. INTRODUCTION

Langerhans Cell Histiocytosis (LCH) is a rare disorder characterized by the clonal proliferation of pathologic Langerhans-type dendritic cells, which originate from the mononuclear phagocyte system[1]. Historically considered a reactive condition, LCH is now recognized as a neoplastic disease driven by activating mutations in the mitogen-activated protein kinase (MAPK) signalling pathway, most commonly involving the BRAF V600E mutation[2]. The reported incidence of LCH is approximately 2-5 cases per million children per year, with a peak occurrence between 1 and 4 years of age[2].

The clinical spectrum of LCH is highly variable and ranges from isolated single-system disease, most commonly affecting bone, to aggressive multisystem involvement with significant morbidity. Skeletal involvement occurs in nearly 80% of paediatric cases, with the skull, long bones, ribs, and jaws being commonly affected sites[7]. Jaw involvement, particularly of the mandible, may lead to early oral manifestations that precede systemic signs, making the oral cavity an important diagnostic site for this condition[5].

Oral manifestations of LCH are diverse and include gingival ulceration, periodontal destruction, tooth mobility, premature exfoliation of teeth, and alveolar bone loss. These features often resemble more common inflammatory, infectious, or traumatic oral conditions, contributing to delayed or missed diagnosis, especially in paediatric patients[4]. Persistent oral ulcers associated with underlying bone destruction should therefore raise clinical suspicion of LCH, particularly when conventional treatment fails to achieve resolution[8].

Radiographically, LCH lesions of the jaws typically appear as well-defined osteolytic areas with loss of lamina dura and rapid alveolar bone destruction, occasionally producing the characteristic "floating teeth" appearance[6]. Although imaging findings are suggestive, definitive diagnosis relies on histopathological examination and immunohistochemical confirmation, with positivity for markers such as CD1a, S-100 protein, and Langerin (CD207) being diagnostic5.

Given the potential for disease recurrence and progression, early recognition and appropriate multidisciplinary management are essential to improve outcomes in affected children. Dental practitioners play a pivotal role in the initial detection of oral manifestations of LCH and in providing long-term follow-up care. This report aims to highlight the importance of recognizing atypical oral presentations of LCH in pediatric patients and underscores the need for thorough evaluation of persistent oral ulcers associated with alveolar bone loss.

CASE REPORT

A 3-year-old male child reported to the Department of Paediatric Dentistry with a complaint of pain associated with a non-healing ulcer in the lower left posterior region of the oral cavity for approximately three weeks [Figure 1]. There was no history of trauma, fever, systemic illness, or similar previous lesions.

Clinical Findings

Intraoral examination revealed an irregular ulcer involving the lingual alveolar mucosa in relation to the lower left primary molars. On palpation, the lesion was tender with mild induration and underlying bony softness, suggestive of deeper involvement. No cervical lymphadenopathy was detected. The associated teeth were non-carious and showed no signs of acute infection.

Radiographic Findings

Orthopantomogram (OPG) demonstrated a localized osteolytic lesion around the lower left primary molars with loss of supporting alveolar bone, a finding commonly associated with LCH-related jaw involvement [Figure 2]. CBCT demonstrating well defined punched out radiolucency with vertical bone loss around primary molars [Figure 3].

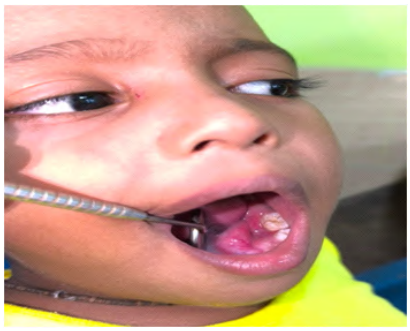

Figure 1: Intraoral pre-operative picture

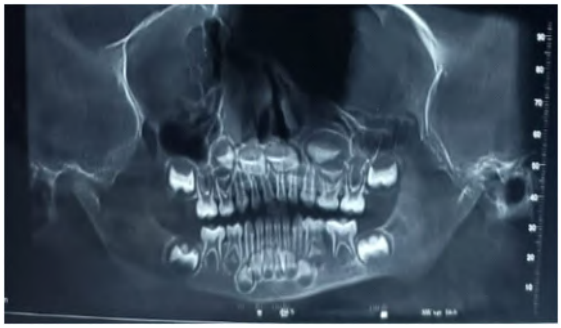

Figure 2: OPG shows Punched out osteolysis with floating teeth

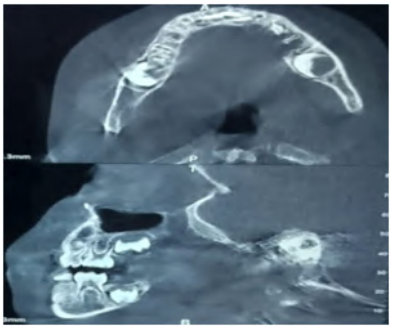

Figure 3: CBCT shows well defined vertical alveolar bone loss around primary molars

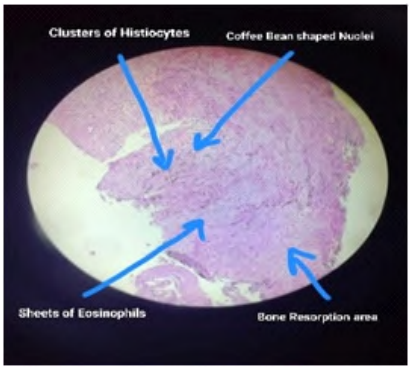

Figure 4: Histopathologic picture of Langerhans cell Histiocytosis

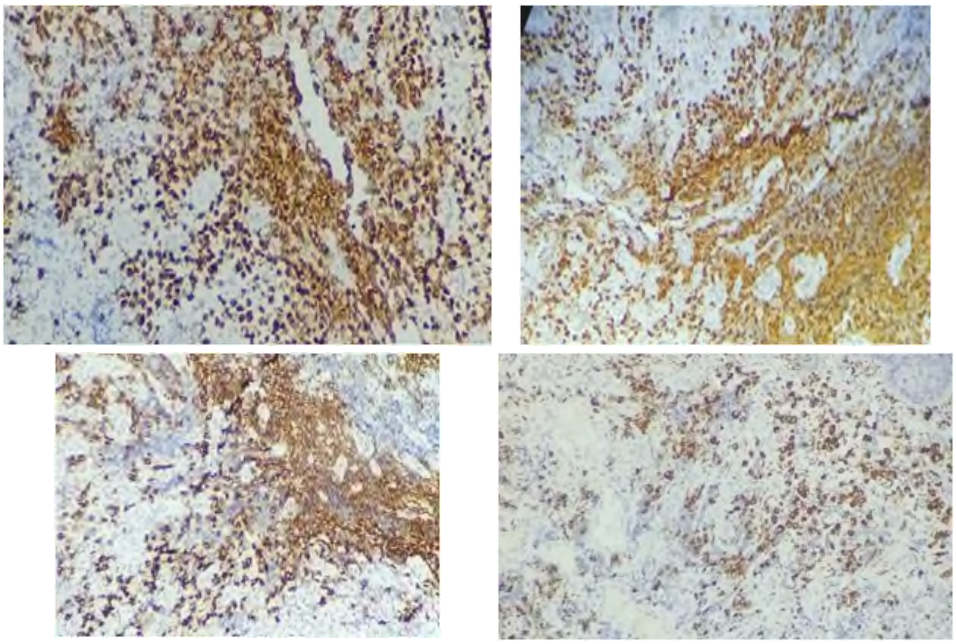

Figure 5: IHC positive for Langerin, CD1a, S100, CD68

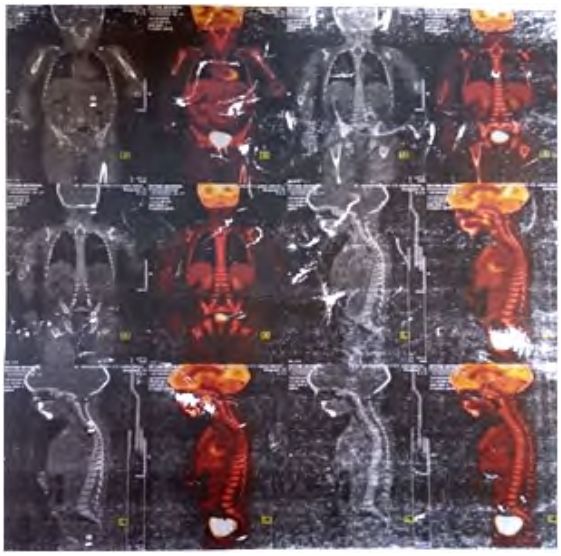

Figure 6: PET CT Scan showing Isolated Mandibular Involvement

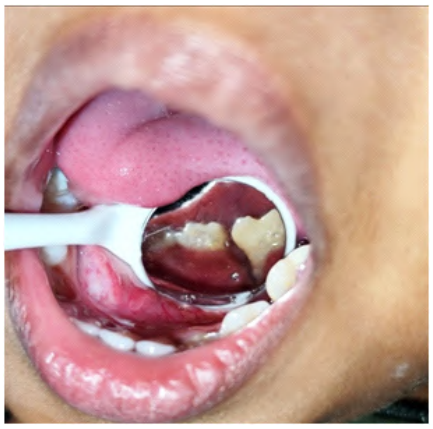

Figure 7: Recurrence of the ulceration at the previous site

Investigations

Based on the clinical and radiographic features, a punch biopsy of the lesion was planned. Haematological investigations revealed elevated serum lactate dehydrogenase (LDH) levels exceeding 450 U/L, indicating increased cellular turnover, which has been reported in active histiocytic disorders.

Histopathology and Diagnosis

Histopathological examination showed sheets of atypical histiocytic cells with grooved, coffee-bean–shaped nuclei, accompanied by a mixed inflammatory infiltrate rich in eosinophils [Figure 4]. Immunohistochemical analysis revealed Langerin, S-100, CD1a, CD68 positivity, confirming the diagnosis of Langerhans Cell Histiocytosis [Figure 5].

Management and follow up

Following diagnosis, the patient was referred for multidisciplinary evaluation involving paediatric oncology and radiology to assess systemic involvement [Figure 6]. Supportive oral care and symptomatic management were provided. The lesion showed initial clinical resolution; however, at the 6-month follow-up, the child presented with reappearance of ulceration at the same site along with radiographic evidence of renewed alveolar bone loss, necessitating further oncologic management and continued close surveillance [Figure 7].

DISCUSSION

Langerhans Cell Histiocytosis (LCH) is an uncommon disorder characterized by clonal proliferation of pathologic Langerhans cells and predominantly affects infants and young children[1]. The disease demonstrates a wide spectrum of clinical presentations, ranging from isolated bone lesions to multisystem involvement with potential morbidity. The jaws, particularly the mandible, are among the frequently affected skeletal sites, making oral manifestations an important diagnostic clue in paediatric patients[5].

Oral involvement in LCH may present as gingival ulceration, periodontal destruction, tooth mobility, or alveolar bone loss, often mimicking common odontogenic or inflammatory conditions [4]. Because these manifestations are nonspecific, diagnosis may be delayed, especially when oral lesions are isolated and unaccompanied by systemic symptoms. Persistent ulceration in conjunction with underlying bone destruction should prompt clinicians to consider LCH among the differential diagnoses, particularly in young children[8].

Radiographic evaluation plays a crucial role in identifying jaw involvement. Typical radiographic features include well-defined osteolytic lesions, loss of lamina dura, and rapid alveolar bone destruction, sometimes producing the classic "floating teeth" appearance[6]. Orthopantomogram serves as an effective initial screening modality, while advanced imaging techniques such as CBCT and MRI provide detailed assessment of cortical and marrow involvement and help determine disease extent[3].

Laboratory investigations are generally nonspecific in LCH; however, elevated serum lactate dehydrogenase (LDH) levels may be observed and reflect increased cellular turnover or active disease[2]. Although LDH lacks diagnostic specificity, it may support clinical suspicion when correlated with radiographic and histopathological findings.

Histopathological examination remains the cornerstone for definitive diagnosis, revealing characteristic Langerhans cells with grooved or "coffee-bean" nuclei and an eosinophil-rich inflammatory background. Immunohistochemical confirmation with markers such as CD1a, S-100 protein, and Langerin (CD207) is essential to distinguish LCH from other histiocytic disorders and inflammatory conditions[4,5]. Advances in molecular biology have further established LCH as a neoplastic disorder driven by activating mutations in the MAPK/ERK signalling pathway, explaining its potential for persistence and recurrence[1].

Management of LCH depends on disease extent and severity. Localized bone lesions may respond to limited surgical intervention or low-intensity systemic therapy, whereas multisystem disease requires systemic chemotherapy under oncologic supervision[3]. Regardless of treatment modality, recurrence is a recognized feature of LCH, particularly in osseous lesions, necessitating prolonged clinical and radiographic follow-up[7].

From a dental standpoint, early recognition of atypical oral ulcers and unexplained alveolar bone loss is critical. Dentists play a vital role in initial detection, biopsy planning, and long-term oral care, especially during oncologic treatment. Regular follow-up is essential to monitor for disease reactivation and to manage dental and developmental complications in growing children.

CONCLUSION

This case highlights the diagnostic challenges posed by paediatric oral Langerhans Cell Histiocytosis presenting as a persistent ulcer with alveolar bone loss. Early recognition, appropriate histopathological evaluation, and multidisciplinary management are essential to optimize outcomes and monitor for disease reactivation.

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