JOURNAL OF WOMEN MEDICAL & DENTAL COLLEGE

Pediatric Mandibular Langerhans Cell Histiocytosis with Maxillary Recurrence: A Case Highlighting Multidisciplinary Management

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Received date: 05-05-2025 **Publication date:** 13-09-2025

Abstract

Histiocytosis X used to be a common name for a diverse group of rare clinicopathologic conditions. Histologic analysis shows these conditions have histiocyte-like cells and different amounts of eosinophils, lymphocytes, plasma cells, and multinucleated giant cells. The histiocytic cells in these lesions are Langerhans cells, so the disease is now called Langerhans cell histiocytosis. This report describes a four-year-old girl who was previously healthy. She was taken to the hospital with a nonhealing ulcer on her mandible. A distinct lytic lesion on her jaw was discovered by radiographic examination. Histopathology and immunohistochemistry are used to confirm the disease. At first, curettage was the only procedure carried out. Two months later, the disease returned in the maxilla, necessitating a second operation and systemic chemotherapy. The patient had no negative side effects from taking the medication as directed. Six months after surgery, imaging and clinical evaluations showed total remission. This case highlights the importance of early diagnosis, careful follow-up, and teamwork in treating pediatric LCH with jaw involvement.

Keywords Gingival ulceration, Pediatric , Histiocytosis X

1. Introduction

Langerhans cell histiocytosis is a rare condition. One distinguishing feature is the abnormal proliferation of Langerhans cells. These cells have specific surface markers like Langerin and CD1a (1). LCH is classified as an idiopathic condition. It involves the clonal proliferation of cells derived from bone marrow that resemble Langerhans cells, including T cells, macrophages, and eosinophils. Mutations in the RAS/MAP kinase pathway cause these changes, which raises the proportion of CD1a and CD207-positive cells (1). In 1868, the Paul Langerhans-named epidermal dendritic cells were discovered. The Birbeck granule is Langerhans cells' ultrastructural characteristic (2). The primary victims of Langerhans Cell Histiocytosis, a rare disease with 0.5–5.4 cases/million people annually that is more common in men (2:1) and has various forms depending on age and system involvement, are children aged 0–15. (3) Classified as dendritic cell histiocytosis, LCH encompasses both localised eosinophilic granulomas and systemic conditions such as Hand-Schüller-Christian and Letterer-Siwe diseases. (4) Mutations in the BRAF or MAP kinase pathways may result from the clonal proliferation of CD1a-, CD207-, and S100-positive progenitor cells in the bone marrow, which can lead to LCH (5). While the 5-year survival rate for multisystem LCH including vital organs is lower (77%), the prognosis for single-system LCH is fairly favourable (100% survival, <20% recurrence in 5 years). (6).

Arriving at a diagnosis of LCH requires a coordinated effort amongst an interprofessional team, beginning with a comprehensive review by a trained pathologist to perform a review of the morphologic immunohistochemical and molecular data on a sample for each individual patient with suspected LCH.

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https://doi.org/10.56600/jwmdc.v3i2.103

This case highlights the challenges in finding and treating recurrent LCH with oral involvement, which is uncommon among pediatric patients

2. Case Report

When she first visited the outpatient department of Oral and Maxillofacial Surgery, Indus Medical College Hospital, Tando Muhammad Khan, Pakistan, the four-year-old girl had no known medical issues, social problems, or family history, presented with a sore on the right side of her mouth had healed three weeks earlier. aEven with the sore, she did not show any systemic symptoms such as fever, weight loss, or general illness. There were no signs of injuries, dental infections, or serious oral health issues. Moreover, no relevant history of drug use was discovered. Intraoral examination revealed a single ulcer on the mandibular ridge with an erythematous base and irregular border. The lesion was tender to the touch, but there were no purulence, soft tissue masses, or obvious swelling. An extraoral examination revealed no cervical lymphadenopathy or facial asymmetry. Biopsy was performed..

On 11/09/2023 an incisional biopsy was done that showed a hyperplastic stratified squamous epithelium showing sheets of cells with grooved nuclei against an inflammatory background of eosinophils. Immunohistochemistry supported the diagnosis plus Langerin, S100, and CDIA positive proteins. (FIG; 1,2,3). Langerin, a Langerhans Cell–restricted protein that induces the formation of Birbeck granules and is constitutively associated with them, is a highly specific marker (Fig 1) . S100 proteins are markers for inflammatory diseases and can mediate inflammation and act as antimicrobials(Fig 2). CD1a is a crucial diagnostic marker for Langerhans cell histiocytosis (LCH). It is expressed on the surface of the abnormal Langerhans cells that characterize the disease(Fig 3). High power view under histopathological section shows sheets of cells with grooved nuclei present against inflammatory background rich in eosinophils, Cells are oval or round with a nuclear groove resembling a coffee bean. rod-shaped inclusions in the cytoplasm known as Birbeck granules (Fig 4)

A panoramic radiograph identified a lytic lesion in the right posterior region of the mandible. The lytic lesion was observed without peripheral sclerosis, which raised suspicion of Langerhans Cell Histiocytosis (LCH). Other differential diagnoses were odontogenic cysts, osteomyelitis, nature bone tumors.

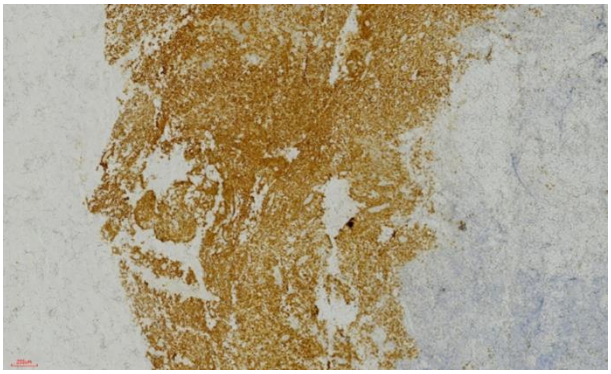

Figure 1: Langerin immunohistochemical marker

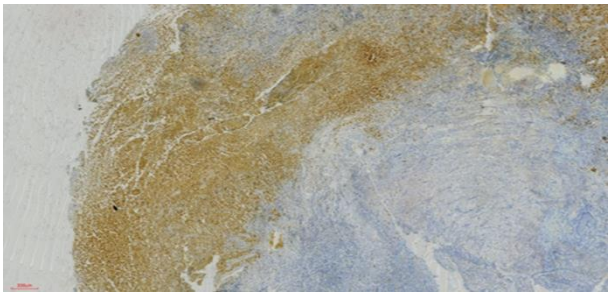

Figure 2: S100 immunohistochemical marker

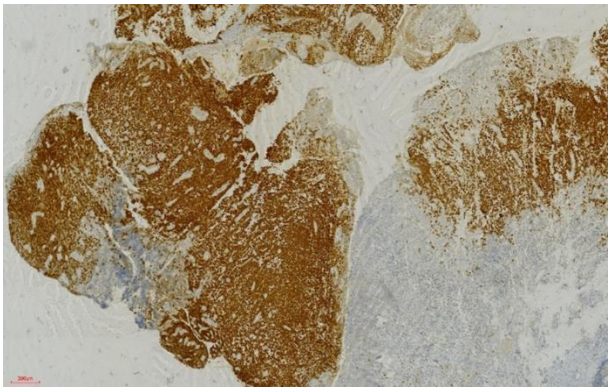

Figure 3: CD1A immunohistochemical marker

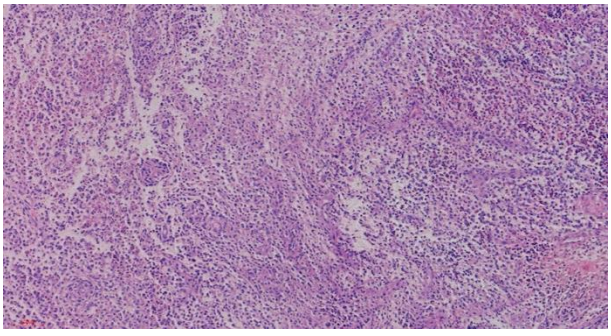

Figure 4: High power Histopathology

showing a Langerhans cell histiocytosis with the characteristic reniform Langerhans cells accompanied by abundant eosinophils. H&E stain.

Langerhans cell histiocytosis diagnosed under histopathology. The diagnosis was simple; quick diagnosis and treatment planning were made possible by imaging and histopathological analysis. The parents of the patient gave her informed consent. The patient's right to privacy was honoured. On September 15, 2023, the patient underwent general anaesthesia for a local aggressive curettage of the mandibular lesion. There were no incidents during the post-operative period. However, the patient had to have a second surgery (under general anaesthesia) on November 15, 2023, because a similar lesion had appeared in the maxilla. On January 12, 2023, chemotherapy began in an effort to stop recurrence. Chemotherapy was well tolerated by the patient, and no side effects were reported. Imaging and clinical exams were performed at later follow-up appointments to gauge the severity of the illness. The patient showed no clinical or radiographic signs of an LCH recurrence six months after chemotherapy ended (01/06/2024). No adverse events or complications were reported during chemotherapy, and the patient was otherwise healthy. (table 1).

Table 1: Results of Chemotherapy

Date	Event
11/09/2023	Incisional biopsy confirming LCH
15/09/2023	Surgical curettage of the mandibular lesion
15/11/2023	Recurrence in maxilla; second surgery
01/12/2023	Initiation of chemotherapy
01/06/2024	Six-month follow-up with no recurrence

3. Discussion

This case shows the difficulties of recurrence of LCH in Paediatric patients. LCH involving the oral cavity is rare but presence of lytic bone lesions or persistent ulcers should include consideration of LCH. Histopathology and immunohistochemistry is vital for diagnosis and using markers like CD1a and Langerin are diagnostically pathognomonic.

In this case, the important lesson using radiology and histology for early diagnosis is shown with the successful diagnosis and management of Langerhans cell histiocytosis (LCH) in a Paediatric patient, including surgical, histopathological and chemotherapy multidisciplinary approach. One significant concern was that the maxillary lesion recurred in two months, which demonstrates the difficulty in controlling the disease and needing treatment. One of the limitations in this case, was that earlier systemic treatment could have resulted in decreased likelihood of recurrence.

This case shows how LCH can present in an atypical manner as a nonhealing mandibular ulcer that otherwise looks like it can be mistaken for osteomyelitis and odontogenic cysts. The sequence of treatment, because of juvenile LCH and the lack of follow-up, local surgical surgery, systemic chemotherapy, adds significance.

The true incidence of Langerhans cell histiocytosis (LCH), which is a rare disease, is not known. A statewide retrospective study in Korea from 1986 until 2010 confirmed that there is an average of about 24 cases/year (603 individuals), median age of diagnosis being 65 months, and a greater frequency in males (1.4:1). The bone is the most commonly involved organ (80%) (7). Research implies that LCH represents a monoclonal neoplasm, although the exact aetiology is unknown because the tumour often has the BRAF V600E mutation, which also governs cell survival, proliferation, motility, and differentiation. In more extensive disease, radiographic imaging will display notable osteolytic lesions and “floating teeth.” The typical presentation may be complaints of mouth pain and swelling. A CT scan may show “punched out” radiolucency, without a corticated border, and periosteal reactions that may confuse LCH with osteomyelitis.

There is considerable difficulty in diagnosing Langerhans cell histiocytosis because numerous common histological features, including lymphocytic infiltration and Langerhans cells also appeared in a number of diseases, including TB and scabies. A diagnosis is accomplished with the collective of clinical, histological, and immunohistochemical elements, all while also noting that there is presence of CD1a and CD207 expressed as surface markers of diseased cells. A lytic ground-glass craze in the jaw is always a combination of more than one differential diagnosis, including ossifying fibroma and Ewing's sarcoma.

For head and neck LCH, often watchful waiting is all that is needed. Many lesions resolve on their own. If there is a need to provide treatment, the treatment is minimally invasive, including topical steroid, minor surgery (curettage) if needed or low dose radiation, but only for progressive disease or when vital structures are at stake. Multidisciplinary management, including regular follow-ups, is critical to ensuring favorable outcomes. (8)

Given the lesion recurrence in the patient, it is possible to deduce that isolated LCH cases may not well to initial local therapy—especially in the juvenile population. This suggests that systemic therapy should be assessed in isolated cases right from the start. Further study is needed to determine if early systemic therapy could reduce recurrence and improve long-term outcomes as suggested in this case.

The aggressive nature of LCH demands multimodal intervention as evidenced by association of recurrence and inadequate initial treatment. This case provides initial support for the proposition that localized LCH can and may not remain localized, but recur later even with fully discussed surgical resection. Systematic assessment and continued follow-up will remain vital for improved patient outcomes.

4. Conclusion

This case demonstrates the importance of early recognition and accurate diagnosis of Langerhans Cell Histiocytosis (LCH) in pediatric patients presenting with persistent oral lesions. Initial biopsy on 11/09/2023 confirmed LCH, and surgical curettage on 15/09/2023 temporarily resolved the mandibular lesion. However, recurrence in the maxilla by 15/11/2023 highlighted the aggressive potential of the disease, prompting initiation of chemotherapy on 01/12/2023.

At six-month follow-up on 01/06/2024, the patient remained disease-free, indicating a favorable outcome with combined surgical and systemic treatment. This case reinforces the need for timely diagnosis, multidisciplinary management (oral and maxillofacial surgeons, pathologists, pediatric oncologists, and radiologists), and careful follow-up to ensure effective control of LCH and prevent recurrence.

Conflict of interest The author declares no conflict of interest.

Acknowledgment We would like to express our sincere gratitude to all individuals who participated in this study.

References

1. Tillotson CV, Reynolds SB, Patel BC, Langerhans Cell Histiocytosis, Salt Lake: StatPearls (Internet)., 2024.
2. K. DM, "Historical perspectives of langerhans cells histiocytosis," hematol oncol clin north Am, vol. 1, pp. 9-21, 1987 MARCH.
3. Christopher R Shea, "Langerhans Cell Histiocytosis," Dermatology, nov 2022.
4. Neville, damm,allen,chi, Langerhans cell histocytosis, elsevier, 2023.
5. Rodriguez-Galindo C, Allen CE, "Langerhans cell histiocytosis," Blood, vol. 135, no. 16, pp. 1319-1331, 2020 april.
6. Hashimoto K, Nishimura S, Sakata N, Inoue M, Sawada A, Akagi M, "Treatment Outcomes of Langerhans Cell Histiocytosis: A Retrospective Study.," Medicina, vol. 57, no. 4, 2021.
7. Kim BE, Koh KN, Suh JK, Im HJ, Song JS, Lee JW, et al, "Clinical features and treatment outcomes of Langerhans cell histiocytosis: a nationwide survey from Korea Histiocytosis Working Party," J Pediatr Hematol Oncol, vol. 36, pp. 125-133, 2014.
8. Steven E. Davis, Dale H. Rice, "Langerhans' cell histiocytosis: Current trends and the role of the head and neck surgeon," ENT journal , vol. 83, no. 5, 2004340-350.