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Case Report
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# Oral and Maxillofacial Langerhans Cell Histiocytosis: A Rare Case Report and Literature Review

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## Abstract

Langerhans Cell Histiocytosis (LCH) is a rather rare tumor derived from myeloid dendritic cells and featured by the ontogenesis and dissemination of CD1a+/CD207+ LCH cells affecting oral and maxillofacial region in most cases. We reported a case of rapid progressive LCH involving bilateral mandible, including one relapse in situ in a male adult patient with a history of repetitive dental interventions. No cases have previously been reported in literature associating external trauma history with the acceleration of the disease course. Diagnostic criteria for differentiation between maxillofacial LCH and periodontitis are also presented based on clinical observation and literature review.

**Keywords: Langerhans Cell Histiocytosis (LCH); Trauma; Oral cancer**

## Introduction

Langerhans Cell Histiocytosis (LCH) is a myeloid neoplastic featured by the ontogenesis and dissemination of CD1a+/CD207+ dendritic cells (LCH cells), occurring in adults with an incidence close to 1-2/million [1]. Based on the number of affected organs and systems, LCH can be classified into three distinct categories: The Single-System Single site (SS-s), the Single-System Multi-site (SS-m), and the Multisystem (MS). LCH that affects the oral and maxillofacial regions encompasses a broad spectrum of clinical manifestations, including alveolar or cortical bone absorption, produces ulcers, lymphadenopathies and periodontitis-like lesion [2]. Single or multiple lesions of inflammation, bleeding, gingival recession, dental mobility, as well as an immature loss of teeth are often witnessed [3]. Here we present a rare case of a male adult patient suffering from single system multi-sites maxillofacial LCH involving bilateral mandible who experienced in total thrice onsets, including one relapse in situ in less than two years. We also offered practical diagnostic criteria for differentiation between maxillofacial LCH and periodontitis, which is particularly notable, considering its significance in early diagnosis.

## Case Presentation

A 33-year-old male patient reported a refractory gingival swelling around the second premolar of left mandible, manifesting reddish swollen in periodontal tissue and later developing into nonspecific pain. From 2016 to 2018, no accurate diagnosis was given but repetitive dental interventions were performed as the second premolar of left mandible was eventually extracted without relieving his discomfort. In June, 2018, he consulted the local hospital and initially diagnosed as an odontogenic cyst. During the surgery, the first molar of left mandible was also removed; an incisional biopsy was performed, which consisted of a partial curettage of the lesion and biopsy taken which postoperative pathological diagnosis confirmed as LCH primary onset lesion. In the meanwhile, the third molars of bilateral mandible, both of which suspected as opsigenes pericoronitis for repetitive discomfort, were extracted at the same time. In October, 2018, he underwent a tumidness in the left mandible and was referred to the Department of Maxillofacial Surgery of Nanfang Hospital. Recurrence of the primary lesion in left mandible was confirmed and expanded excision was performed. In November, 2018, he went to the Department of Periodontal Dentistry of Nanfang Hospital with the chief complaint of painful reddish swelling around the second molar in right mandible. We speculate another LCH primary onset occurred in right mandible. A biopsy was taken, and through immunohistochemical test LCH was diagnosed. Expanded mandibular excision was performed. By far, he was closely followed in case of recurrence. Panoramic radiograph was taken regularly and the latest one shot in June, 2019 shows an ideal prognosis. Clinical course of the patient with X–ray panoramic radiograph was presented in Figure 1.

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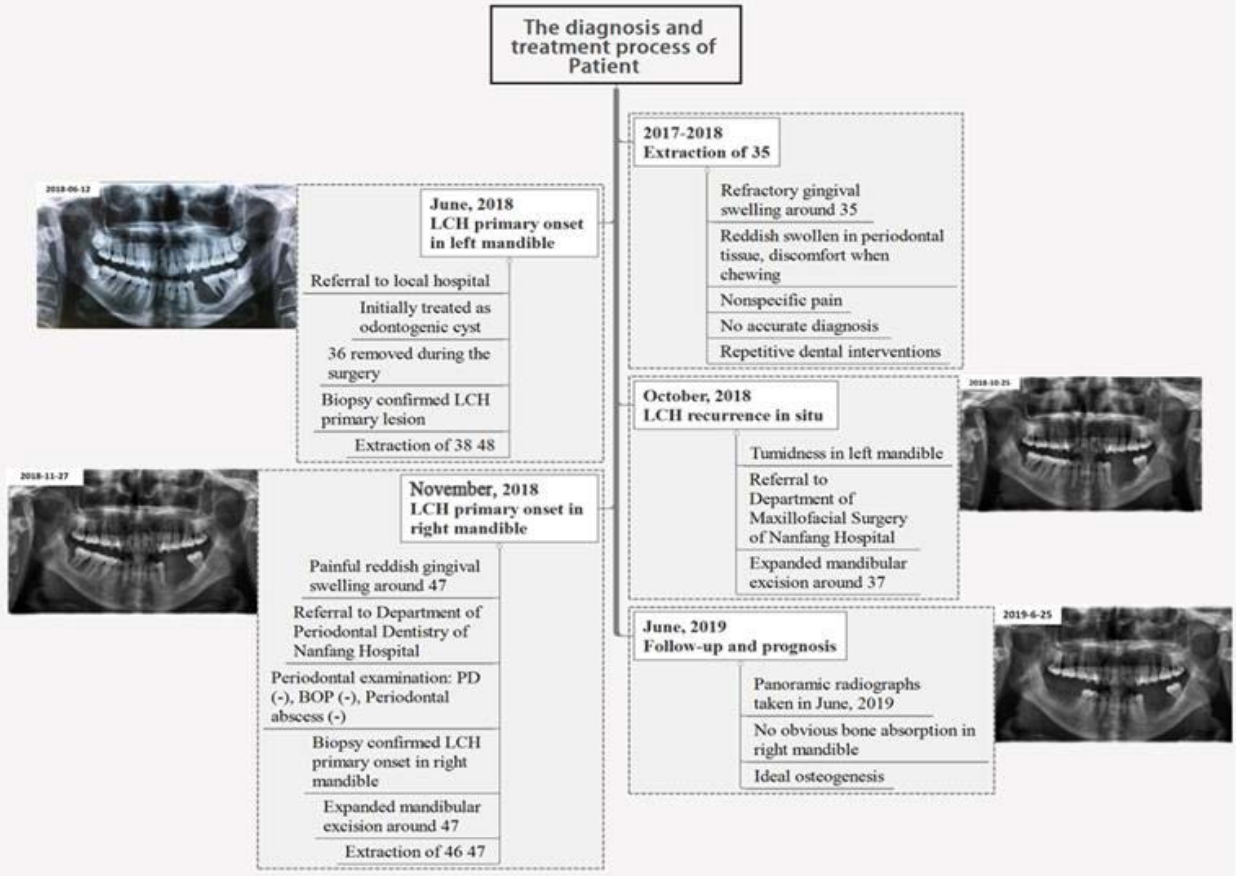


**Figure 1:** Clinical course of the patient with X-ray manifestations.
1) June 2018: panoramic radiography showed that third molars existed in bilateral mandible, anodontia of 35 and arc-shaped bone erosion between 35 and 36. Normal bone substance radiolucency around 47 was observed.
2) October, 2018: 36 had been surgically removed. 38 and 48 were extracted during the first surgery. Radiologic assessment presents arc-shaped bone erosion around 47. Alveolar bone resorption in 47 reached the apical 1/3.
3) November, 2018: Panoramic radiograph shows progressively increased erosive lesion, resulting in a more severe apical alveolar bone resorption around 47.
4) June, 2019: Panoramic radiograph revealed no expansion of bone defect area in the right mandible. Ideal osteogenesis was witnessed in the left mandible.

## Discussion

Oral and Maxillofacial LCH mainly presents as unifocal or multifocal disease affecting oral and maxillofacial region (particularly the posterior maxilla) in nine out of ten cases [1]. However, given the limited number of LCH incidence (approximately 1-2/million in adults [4]), clinicopathological features of oral and maxillofacial LCH of adult patients are seldom reported in the dental literature to the best of our knowledge. Disease history (especially trauma history) has not previously been particularly addressed in LCH treatment, as there is only one children patient aged about ten reported a history of trauma but no correlation was made between [5]. Although a few studies previously reported that LCH can sometimes be self-limited, partial resection could not ensure a steady and favorable prognosis. Song reported a case of orbital LCH relapsed months after biopsy and excision treatment [6]. Trauma history can be an important factor accelerating the clinical disease course of LCH, as supported by the unusually rapid clinical course of our patient. LCH relapsed in primary lesion four months after he received the first surgery in left mandible. Another primary LCH lesion occurred only five months after the third molar tooth extraction, manifesting as an accelerating bone absorption progression around the apical area of the second molar in right mandible. Due to the lack of clinical data and experimental studies, whether external trauma is one of the triggers or contributes to LCH onset remains to be determined. The mechanism is uncertain about whether operative interventions in the physical could pathologically disrupt disease cascade [7]. Overall, this case outlines the importance of considering external injury or trauma history including repetitive dental surgery (tooth extraction, bone excision, periodontal scaling and cyst excision) in the treatment of LCH. Clinically, LCH is difficult to distinguish from periodontitis, acute necrotizing periodontitis, inflammatory condition (sarcoidosis), soft tissue sarcoma and so on [8]. During the treatment of the patient, we retrieved his medical records and found him diagnosed as periodontitis, pulp inflammation and odontogenic cysts by different dental practitioners. Literature showed that there were a number of patients, similar to this patient, were initially referred to periodontal specialists [9]. Oral changes are usually the first, unique manifestations of LCH [10-20]. The differential diagnosis can sometimes be difficult due to rather nonspecific manifestations. Based on our clinical observation and other case reports, we summarized the following diagnostic criteria for differentiation between maxillofacial LCH and periodontitis:

- Both patients with periodontitis and patients with LCH proliferation can present gingival redness and swelling. In periodontal examination, patients with periodontitis manifest deep periodontal pockets. In LCH patients,

however, periodontal probe is often unable to penetrate into the gingival sulcus.

- Alveolar bone resorption occurred in both periodontitis patients and mandibular LCH patients but the former presents as wedge-shaped bone resorption, while the latter presents as a circular-arc bone resorption. Jaw lesions manifest as single lumen radiolucency with circumscribed borderline [11].
- Progression of periodontitis was rather slow compared with LCH patients. In LCH patients, bone defect can be the initial symptom and rapid progression occurred.
- Usually, one of the clinical manifestations of periodontitis is poor oral hygiene condition without systemic inflammatory responses. Patients with LCH, whether adults or children, can manifest as consequent teeth moving, toothache as well as "floating teeth" mobility [21] and accompanied with other systemic complications such as diabetes insipidus, bone defects in other systems [21].

More generally, we suggested that dental intervention towards LCH patient including periodontal scaling, incision or tooth extraction should be performed prudently. Our preference treatment is early diagnosis and expanded incision instead of limited curettage, avoiding minimalist repetitive interventions. In our observation, most patients are free from local recurrence but it is noteworthy that long-term follow-up should be carried especially to LCH patients with external trauma or tooth-extraction history.

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## References

1. Bedran NR, Carlos R, de Andrade BAB, Bueno APS, Romanach MJ, Milito CB. Clinicopathological and Immunohistochemical Study of Head and Neck Langerhans Cell Histiocytosis from Latin America. Head Neck Pathol. 2018;12:431-9.
2. Madrigal-Martínez-Pereda C, Guerrero-Rodríguez V, Guisado-Moya B, Meniz-García C. Langerhans cell histiocytosis: Literature review and descriptive analysis of oral manifestations. Med Oral Patol Oral Cir Bucal. 2009;14(5):E222-8.
3. Su M, Gao YJ, Pan C, Chen J, Tang JY. Outcome of children with Langerhans cell histiocytosis and single-system involvement: A retrospective study at a single center in Shanghai, China. Pediatr Hematol Oncol. 2018;35:385-92.
4. Kobayashi M, Tojo A. Langerhans cell histiocytosis in adults: Advances in pathophysiology and treatment. Cancer Sci. 2018;109(12):3707-13.
5. Almuzayyen A, Elhassan W, Alabbadi M. Intralesional Triamcinolone for Treating Mandibular Langerhans Cell Histiocytosis: A Case Report and Literature Review. Saudi J Med Med Sci. 2019;7:47-50.
6. Song A, Johnson TE, Dubovy SR, Toledano S. Treatment of recurrent eosinophilic granuloma with systemic therapy. Ophthalmic Plast Reconstr Surg. 2003;19:140-4.
7. Woo KI, Harris GJ. Eosinophilic granuloma of the orbit: Understanding the paradox of aggressive destruction responsive to minimal intervention. Ophthalmic Plast Reconstr Surg. 2003;19:429-39.
8. Babu NC, Kumar KV, Juneja NLS. Aggressive periodontitis as a presenting sign of Langerhans cell histiocytosis. Oral Oncology. 2011;47:S92-S.
9. Guimarães LF, Dias PF, Janini ME, de Souza IP. Langerhans Cell Histiocytosis: Impact on the Permanent Dentition After an 8-year Follow-up. J Dent Child (Chic). 2008;75(1):64-8.
10. Shirley JC, Thornton JB. Oral manifestations of Langerhans' cell histiocytosis: Review and report of case. ASDC J Dent Child. 2000;67(4):293-6.
11. Lombardi T, Hauser C, Budtz-Jorgensen E. Langerhans cells: Structure, function and role in oral pathological conditions. J Oral Pathol Med. 1993;22:193-202.
12. Luz J, Zweifel D, Hüllner M, Bühler M, Rücker M, Stadlinger B. Oral manifestation of Langerhans cell histiocytosis: A case report. BMC Oral Health. 2018;18(1):106.
13. Khonsari RH, Ruhin B. Images in Clinical Medicine. Loose Teeth and Excessive Thirst. N Engl J Med. 2016;374:e25.
14. Sherwani RK, Akhtar K, Qadri S, Ray PS. Eosinophilic granuloma of the mandible: A diagnostic dilemma. BMJ Case Rep. 2014;2014.
15. Kannan K, Alwithanani N, Salama M, Kumar M, Uthappa R, Ahamed M. Eosinophilic Granuloma in Jaw Bone: A Pare Pediatric Case Report. Ethiop J Health Sci. 2016;26:487-90.
16. Liu J LD, Zhou Y, Chen Q, Zeng X, Jin X. Langerhans cell histiocytosis with widespread ulcers and masses on the palate. Indian J Dermatol Venereol Leprol. 2014;2014;80:462-3.
17. Tamura R, Maeda S, Terashi H. Reconstruction of a severe mandibular pathological fracture caused by Langerhans cell histiocytosis using a free fibula osteocutaneous flap: A case report. Case Reports Plast Surg Hand Surg. 2018;5:9-13.
18. Terada T. Recurrent multifocal Langerhans cell histiocytosis of the mandible and maxilla in a 46-year-old man: A pathologic case report. Int J Clin Exp Pathol. 2013;6(5):939-42.
19. Cisternino A, Asa'ad F, Fusco N. Role of multidisciplinary approach in a case of Langerhans cell histiocytosis with initial periodontal manifestations. Int J Clin Exp Pathol. 2015;8(10):13539-45.
20. V Y, E SK, S NB, A K, Sekhar MS. Solitary extragnathic langerhans cell histiocytosis - A rare case. J Clin Diagn Res. 2015;9:ZD22-3.
21. Facciolo MT, Riva F, Gallenzi P, Patini R, Gaglioti D. A rare case of oral multisystem Langerhans cell histiocytosis. J Clin Exp Dent. 2017;9(6):e820-e824.