Asian Journal of Dental Sciences

3(4): 8-11, 2020; Article no.AJDS.60334

Langerhans Cell Histiocytosis the Great Mimicker, Awareness in Oral Diagnosis and Management

Muhammad Hilmi Bin Zainal Ariffin[1*], Nor Hidayah Reduwan[1], Wan Nur Hazirah[1] and Faizal Hafez Hidayat[1]

[1]Faculty of Dentistry, UiTM Sungai Buloh Campus, Selangor, Malaysia.

Authors' contributions

This work was carried out in collaboration among all authors. Author MHBZA involved in the clinical management. Author NHR involved in radiographical assessment. Author NHR involved in histological analysis. Author FHH as the supervisor. All authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Ketij Mehulić, University of Zagreb, Croatia.
Reviewers:
(1) Cláudia Roberta Leite Vieira de Figuiredo, Universidade Federal da Paraíba, Brazil.
(2) Montserrat Reyes, University of Chile, Chile.
(3) Daniel Andres Rappoport Wurgaft, University of Chile, Chile.
Complete Peer review History: http://www.sdiarticle4.com/review-history/60334

Case Study

Received 15 June 2020
Accepted 21 August 2020
Published 01 September 2020

ABSTRACT

Aim: Langerhans cell histiocytosis or LCH is an atypical cancer originating form histocytes. Dental manifestation may mimic aggressive form of periodontitis with gingival overgrowth and bleeding, advance alveolar bone loss as well as generalized mobility of tooth. The ambiguity of its clinical features is one of the factors that makes identifying the disease a difficult task.

Presentation of Case: A 30 years old Malay man came with complain of gingival swelling and bleeding on the upper right quadrant associated with bad breath, scarce saliva, generalized tooth mobility, and whitish slough on the gingiva. The condition however does not present with pain or any systemic condition. The patient claimed to be systemically healthy. Intraoral examination displayed a diffused, erythematous swelling over attached gingiva on the maxillary posterior buccal region bilaterally along with sign of loss of pyramidal shape and ulceration with a punched-out profile covered by necrotic slough.

Discussion: Diagnosis of LCH is done with the aid of immunohisto chemical analysis of CD1a and S100. The clinical course of LCH is highly unpredictable and irregular, improvement noticeable with conventional periodontal treatments in controlling the oral manifestations of LCH on early course of the disease.

Conclusion: Proper multidisciplinary approach is essential in the management for the benefit of the patient in such rare entity.

**Corresponding author: Email: curvelist0202@gmail.com;*

Keywords: Periodontitis; periodontal medicine; langerhans cell histiocytosis; diagnosis; management; multidisciplinary.

1. INTRODUCTION

Langerhans cell histiocytosis or LCH is an atypical cancer originating form histocytes, a family of dendritic cells that has a role in the body immunity. Adult-onset LCH incidence is extremely rare, reporting 0.07 cases per million [1]. The abnormal proliferation of CD1a positive Langerhans cell is the main characteristic of LCH. These abnormal multiplying of LCH causes accumulation of the cells to specific part of the body [2]. Bone is the commonest organ to be involved. The most frequent site is on the skull (57%), this is then followed by vertebrae (30%) and the maxilla and mandible (26%) [3]. Dental manifestation may mimic aggressive from of periodontitis with gingival overgrowth and bleeding, advance alveolar bone loss as well as generalized mobility of tooth [4]. The ambiguity of its clinical features is one of the factors that makes identifying the disease a difficult task.

2. PRESENTATION OF CASE

A 30 years old man came with complain of gingival swelling and bleeding on the upper right quadrant associated with bad breath, scarce saliva, generalized tooth mobility, and whitish slough on the gingiva. Patient claimed to be systemically healthy. Intraoral examination displayed a diffused, erythematous swelling over attached gingiva on the maxillary posterior buccal region bilaterally along with sign of loss of pyramidal shape and ulceration with a punched-out profile covered by necrotic slough (Fig. 1). Miller Grade III mobility was noted for majority of the tooth. The results of the baseline periodontal examination revealed a mean probing depth (PD) was 4.4 mm, with a maximum value of 10 mm. 114 out of 168 sites (67%) had a PD of ≥4 mm.

Orthopantomogram was taken (Fig. 2) showing extensive irregular pattern of peri radicular lesion and perforated floor of the right maxillary sinus. On the mandible demonstrating scoop-out pattern of lesion bony resorption with the preservation of alveolar crest distal to 34 and intact furcation alveolar segment of 36.

No abnormalities were detected in both the biochemical and laboratory tests showing no other involvement except for oral region. Through the case presentation, differential diagnosis of, 1) Generalized Aggressive Periodontitis and 2) Necrotizing Ulcerative Periodontitis.

Biopsy including both hard tissue and soft tissue on the subsequent review were done due to recurrence of the ulceration and was verified for CD1a and S100 immunohistochemistry test (Fig. 4). The neoplastic cells exhibited nuclear groove or indented vesicular nuclei with prominent nucleoli and indistinct cell border (Fig. 3). Patient is diagnosed with Periodontitis as a manifestation of systemic disease (Langerhans cell histiocytosis).

The management consist of oral hygiene protocol with full mouth scaling and root debridement with antibiotic adjunct of a combination of Amoxicillin 500 mg and Metronidazole 400 mg administered for one week, along with the use of chlorhexidine mouthwash 0.2% and chlorhexidine gel for 2 weeks. Initial clinical improvement was seen (Fig. 5), patient is then planned for local steroid injection of affected site under local anesthesia.

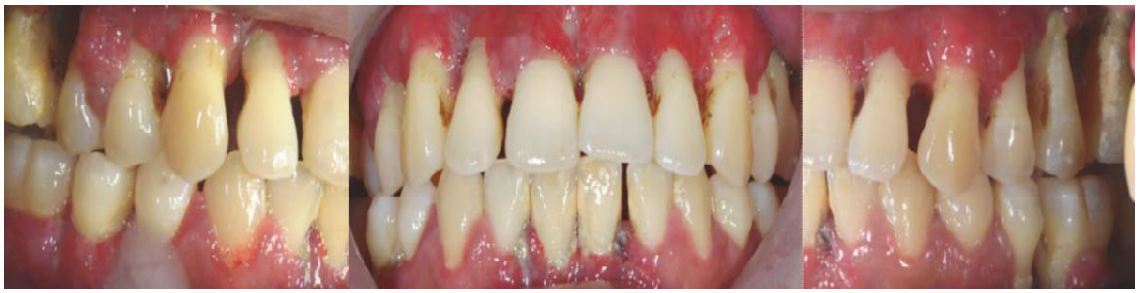

Fig. 1. Ulceration with a punched-out profile covered by necrotic slough

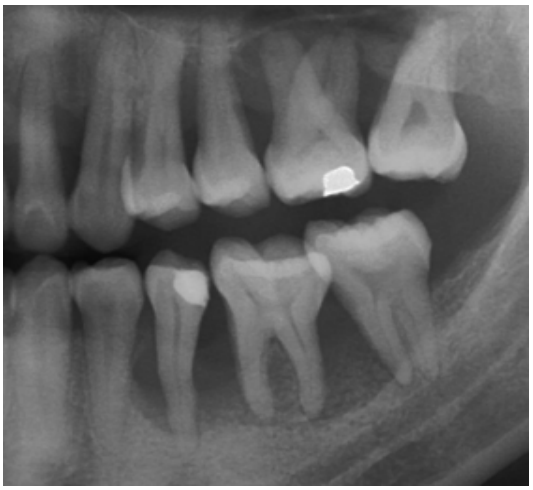

Fig. 2 Scoop-out pattern of lesion bony resorption with the preservation of alveolar crest distal to 34

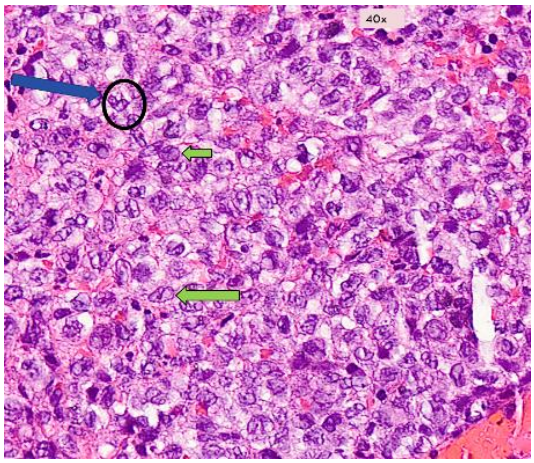

Fig. 3. Histology of Buccal Gingiva 36, Pale staining histiocytic cells exhibiting nuclear groove & indented nuclei with indistinct cell border (coffee bean/kidney shaped)

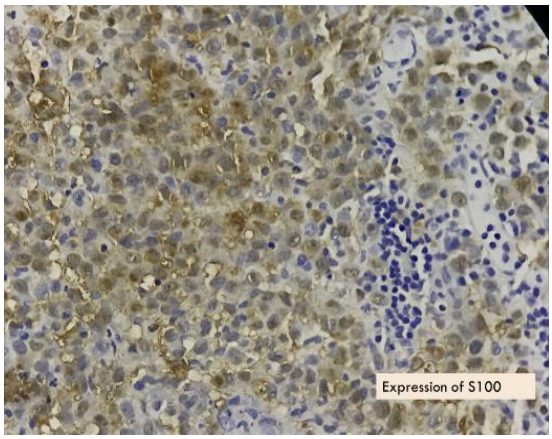

Fig. 4. Histiocytic cells demonstrate diffuse strong positivity for CD1a and focal positivity for S100 (dark stained)

3. DISCUSSION

Final Diagnosis was made with the aid of immunohistochemical analysis. The hallmark of LCH is via assessment of S100 and CD1 on tissue biopsy [5], in relation to the case soft tissue and hard tissue biopsy reveals presence of focal area of diffused sheets of large pale staining histiocytic cells. The clinical course of LCH is highly unpredictable and irregular, with regards to the case improvement were noticed with conventional periodontal treatments in controlling the oral manifestations of LCH on early course of the disease.

Curettage of lesion bearing site in cases of small lesions of 2 cm or less may be sufficient leading to consequent healing making further intervention irrelevant. For massive lesion, total curettage is not recommended [6]. In cases of multiple system LCH, the principal management based is the utilization of a combination of chemotherapy regime of Vinblastine, bleomycin,

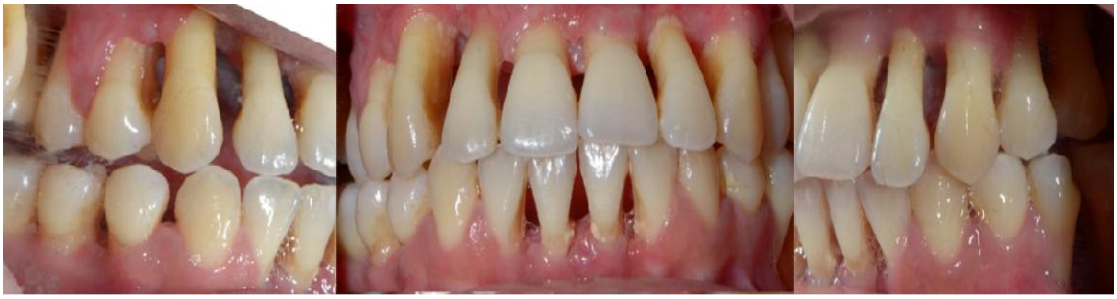

Fig. 5. Initial clinical improvement

methotrexate (VBL) 6 mg/m2 intravenously and a weekly oral bolus of 40 mg/m^2 daily prednisolone for the total period of 6 weeks [7].

4. CONCLUSION

Dental practitioners in dealing with daily patients should be aware of any clinical signs that hint of deviation of common clinical findings. The ambiguity between clinical manifestation of systemic diseases and common oral disease may lead to under diagnosis in such cases. Proper multidisciplinary approach is essential in the management for the benefit of the patient in such rare entity.

CONSENT

Authors affirm that written informed consent has been obtained from the patient for the use of publication in this case report along with accompanying images.

ETHICAL APPROVAL

As per university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Goyal G, Shah MV, Hook CC, et al. Adult disseminated Langerhans cell histiocytosis: Incidence, racial disparities and longterm outcomes. Br J Haematol. 2018;182(4):579–581.
2. Willman CL, et al. Langerhans'-cell histiocytosis (histiocytosis X)-- A clonal proliferative disease. N. Engl. J. Med. 1994;331:154–160.
3. Al-Ammar AY, Tewfik TL, Bond M, Schloss MD. Langerhans' cell histiocytosis: paediatric head and neck study. J. Otolaryngol. 1999;28:266–272.
4. Hernández-Juyol M, Boj-Quesada JR, Gallego-Melcon S. Oral manifestations of Langerhans cell histiocytosis. Case study of a two-year-old boy. Med. Oral. 2003;8: 19–25.
5. Hammouri EH, Sweidan HA, AShokaibi O, Al Omari L. Langerhans cell histiocytosis: A case report with oral manifestations and the role of pediatric dentists in the diagnosis. Clin Case Rep. 2020;8:545–549.
6. Haupt R, et al. Langerhans cell histiocytosis (LCH): Guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years. Pediatr. Blood Cancer. 2013;60:175–184.
7. Tazi A, et al. Vinblastine chemotherapy in adult patients with langerhans cell histiocytosis: A multicenter retrospective study. Orphanet J. Rare Dis. 2017;12:95.

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/60334