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CASE REPORT

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A case report of adult Langerhans cell histiocytosis and review of the literature

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Abstract

Langerhans cell histiocytosis (LCH) is an uncommon proliferative disease with an unknown cause. Its clinical manifestations vary and can involve a variety of organs. To diagnose LCH, radiographs, histopathological and immunohistochemical findings are essential. The gold standard for a definite diagnosis is positive CD1a/CD207 and S100 in the immunohistochemical results. Different treatment plans are available for patients struggling with LCH. To our knowledge, the LCH incidence rate is about 8.9 in one million children and 1–2 cases in one million adults. Our case shows the importance of early diagnosis of this rare condition for the prevention of any further spreading. This case report is about a 35-year-old male patient struggling with diabetes insipidus with a chief complaint of tooth mobility. Based on his X-ray radiographs, osteolytic lesions were found. A biopsy was performed, and due to histopathological and immunohistochemical findings, it was diagnosed as a case of LCH in adults. This paper shows that although the incidence rate of LCH is rare in adults, dealing with a patient struggling with diabetes insipidus and osteolytic bone lesions with an unknown cause, LCH must be taken into consideration. Due to the fact that LCH's first manifestations are mostly first oral, its good prognosis in the early stages, and if it progresses, it can turn fatal, it is important that dentists are aware of this disease, its clinical manifestations, and patient management. In case of suspicion, X-ray examination, biopsy, histopathological, and immunohistochemical exams must be performed.

KEYWORDS

adult, diabetes insipidus, eosinophilic granuloma, histiocytosis, Langerhans cells, osteonecrosis

1 | INTRODUCTION

Langerhans cell histiocytosis (LCH) is an uncommon disease with an unknown cause; it is featured by exceeding and acute proliferation of Langerhans cells derived from bone marrow and mature eosinophils in the skin (erythematous and crusted macules, papules, or nodules, with or without ulceration, or petechiae, or seborrhea-like picture[1]), bone, lymph nodes, and other organs of the body.[2–4]

LCH commonly involves the head and neck, especially bones of the skull and jaws (most commonly in

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the gingiva and hard palate).[5,6] Clinical features of LCH vary; they are mixed and can be from a single lytic lesion of bone with a favorable prognosis to a form that is fatal, disseminated, and leukemia-like (Figure 1). LCH mostly involves bone but can be found in almost all organs of the body.[3] Clinically, affected patients show polyuria, polydipsia, and other symptoms such as skin rash, lymphadenopathy, gingival hypertrophy, disturbed balance, and memory problems. The oral manifestation of LCH is an ulcero-proliferative lesion with bleeding gingiva, mobile teeth, and the presence of pain in the area. The symptoms of LCH mostly happen in the mouth prior to any other place in the body. Thus, a thorough examination of the oral cavity and considering a differential diagnosis is important to diagnose LCH at its early stages.[4,7]

LCH is mostly diagnosed by biopsy and microscopic examination of the lesion.[8] Its conventional histopathology shows a destructive lesion with mononuclear cells. These cells contain intended nuclei and have the phenotype of dendritic antigen-presenting Langerhans cells. LCH is also characterized by the infiltration of lymphocytes and eosinophilic granulocytes forming pseudo-abscesses. Definitive diagnosis of LCH based on its histopathology needs S100 and CD1a positive immunohistochemistry or the presence of Birbeck granules under electron microscopy.[4,7] Other indicating factors that can lead to a specific diagnosis of LCH are: Langerin (CD207), Vimentin, CD45, ecto-ATPase, and simultaneous expression of CD68, MIB-1, peanut agglutinin, and placental-like alkaline phosphatase.[2]

The stage of this disease should be determined by a skeletal X-ray examination of the bones. Based on the results and symptoms of the patient, further examinations may be necessary. For the treatment of adults struggling with LCH, different treatment plans are available based on the organs this disease has involved, its clinical stage, age of the patient, and dysfunction of the affected organs. Treatment options are such as watchful waiting, surgical excision and curettage, intralesional corticosteroid injection, Mechlorethamine, Hydrochloride aqueous solution, Thalidomide, irradiation, chemotherapy, immunomodulation, and transplantation (in advanced stages of the disease). The stage of the disease and its prognosis depends mostly on the patient, disease, and factors that are related to the therapy. Multisystem diseases' result is worse than single-system diseases but in the early stages of the limited type of disease, the prognosis is favorable (usually without the need for systemic treatment).[4,7,9] Usually, for the localized form, such as an isolated bone lesion, a minor treatment like biopsy or curettage (radical excision is discouraged) may be needed. For non-symptomatic patients, due to the possibility of spontaneous resolution and healing of the lesion, the "wait and see" approach is also possible. Systemic therapy is useful for multifocal/multisystem LCH and reduces its mortality and morbidity rate. Because of the rarity of LCH, it is still questionable which treatment pathways are the best for different clinical situations.[3,4,7,9,10]

'Single-system disease'	
Localized (single site)	Monostotic bone involvement
	Isolated skin involvement
	Solitary lymph node involvement
Multiple site	Polyostotic bone involvement
	MFB [mutifocal bone disease] (lesions in two or more different bones)
	Multiple lymph node involvement
CNS-risk lesions: involvement of facial bones, sinuses, the maxilla, or anterior or middle cranial fossa (temporal, mastoid, sphenoidal, ethmoidal, zygomatic, orbital bones) with intracranial tumor extension. Not included are vault lesions.	
'Multisystem disease'	
'Low-risk group'	Disseminated disease (multiple organs involved) without involvement of risk organs[1]
'Risk group'	Disseminated disease with involvement of one or more of the risk organs[1]

[1]Risk organs include the haematopoietic system, lungs, liver, and spleen.

FIGURE 1 Classification of Langerhans cell histiocytosis.[7]

As LCH is relatively rare, limited data are available. LCH occurs in male predilected children. Based on the English literature, the incidence of the disease is 8.9 in one million children and 1–2 cases in one million adults.[4]

As LCH is rare in adults and its first manifestations are mostly in the oral cavity, it is important for dentists to diagnose the lesions of this disease in the early stages and treat the patient to prevent the disease to progress.

2 | CASE PRESENTATION

A 35-year-old man without any significant familial or psycho-social adverse history was referred to Shiraz Oral and Maxillofacial Medicine Department. He had a chief complaint of a mobile tooth, first recognized 6 months before his appointment, in the right posterior area of his mandible and the will to extract the tooth. He gave us the history of the extracted mandibular second molar due to its mobility. He was under treatment with antidiuretic hormone replacement spray (DDAVP Nasal Spray [desmopressin acetate nasal spray]) due to his polyuria (the result of struggling with diabetes insipidus). He complained of skin pruritus with rashes on several parts of his body (especially his legs). As seen in Figure 6, skin rashes were erythematous crusted macules.

After signing the written consent form and performing several clinical examinations, other mobile teeth were observed as well, especially in the molars.

His cone-beam computed tomography (CBCT) and orthopantomography (OPG) (Figure 2) showed "floating in

air" teeth in the lesion with no detectable root resorption, a well-defined radiolucency extending from the mesial aspect of the right mandibular canine to the distal aspect of the right mandibular first molar (scooped out periphery), presence of buccal and lingual cortices and alveolar crest destruction, inferior alveolar canal within the lesion via loss of superior cortex, mandibular canal displacement, lamina propria loss, no reactive changes in the surrounding of bone structure in the mandible and severe destruction of the alveolar process in the molar region on both sides (extending to maxillary tuberosity), loss of posterior aspect of maxillary sinus floor on both sides (especially on the left), and severe mucosal thickening of both maxillary sinuses in the maxilla. Magnetic resonance imaging (MRI) (Figure 3) of the patient as well showed severe mucosal thickening of the maxillary sinuses (both sides, especially the left sinus) on both T1- and T2-weighted images. The mandibular area of the patient is not clear in MRI images; therefore, no significant result can be concluded for the mandible from MRI. Due to his CBCT and OPG radiographs, a differential diagnosis for LCH and squamous cell carcinoma was considered, and aggressive forms of periodontal disease were ruled out. The blood test results of the patient showed high triglycerides, low hematocrit and mean corpuscular hemoglobin (MCH), and high aspartate transaminase (AST) and alanine transaminase (ALT) levels which suggested that he was anemic and had liver disorders. As he was struggling with diabetes insipidus, despite polyuria, his urine test was normal, and there was no evidence of glucose in the tested urine. Due to this polyuria and his undertreatment diabetes insipidus, and oral cavity health condition, a disease that can affect several organs and has a variety of symptoms, were suspected.

Eventually, the mandibular right second molar was extracted and an excisional biopsy in the mandibular right premolar and molar area was performed (around the 4th, 5th, and 6th teeth). After that, the specimen was placed in a formalin solution and the biopsied lesion (from the end of the tooth's root and in the extracted tooth socket) was sent to Shiraz Oral & Maxillofacial Pathology Department for a histopathologic examination.

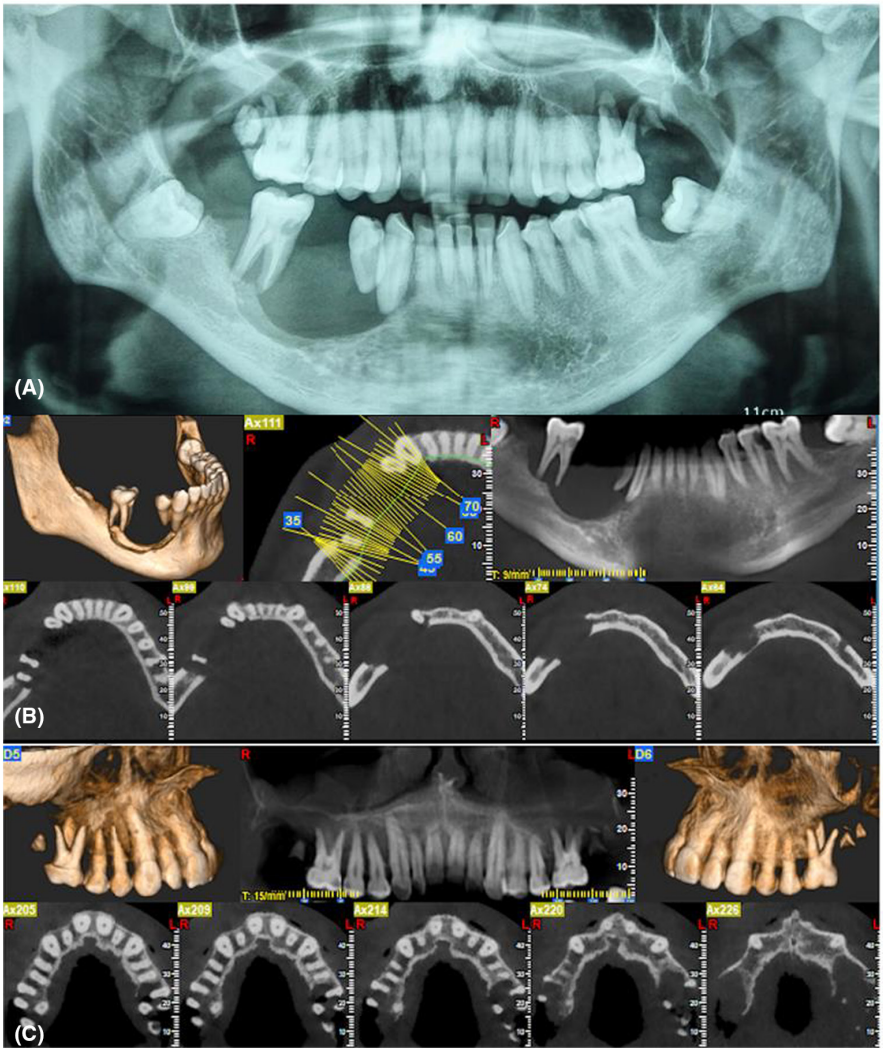

FIGURE 2 The baseline CBCT shows bone resorption ("scooped out" periphery) in the right mandible (from right canine to right first molar) (B), and the left and right molar regions of the maxilla extending to the maxillary tuberosity (C) in a reconstructed panoramic view and 3D-reconstruction. In Figure (A), bone resorption of the mentioned areas can be observed in a panoramic radiograph.

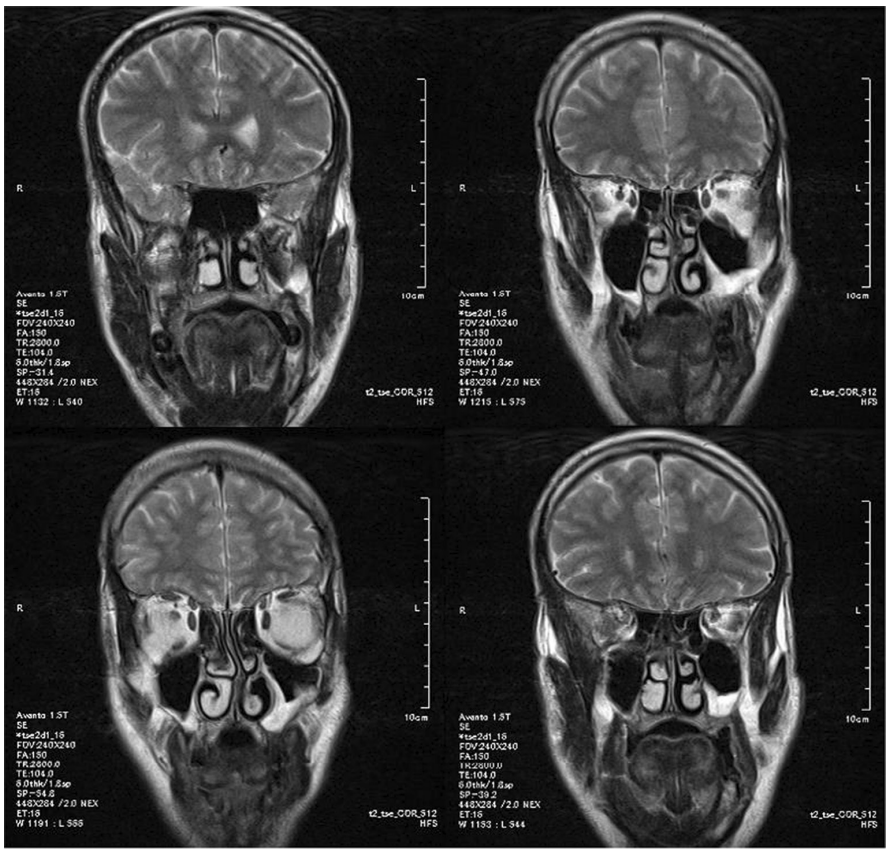

FIGURE 3 MRI images of the patient showing severe mucosal thickening of maxillary sinuses (both sides, especially the left sinus) on both T1- and T2-weighted images. Mandibular area is not clear in images.

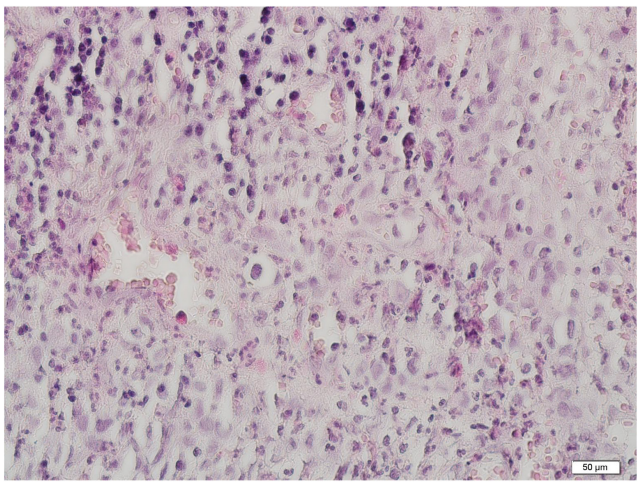

FIGURE 4 400x H&E LCH biopsy: the proliferation of histiocytes and eosinophil infiltration is seen in histologic findings.

Macroscopic examination revealed that the specimen consisted of a piece of irregular creamy-brown elastic tissue, measuring 1.8 × 0.9 × 0.7 cm.

Microscopic examination (Figure 4) showed that the sections contained para-keratinized stratified squamous epithelium with exocytosis and intracellular edema. The connective tissue demonstrated diffused and severe infiltration of chronic inflammatory cells with sheets of histiocytes and numerous scattered eosinophils. Hemorrhaging areas, Russell bodies, bacterial colonies, and a focal area of giant cells were also seen.

Based on the histopathological findings, it was suggested that our patient's condition had been LCH, but for a definite diagnosis, immunohistochemistry (IHC) evaluation (detecting the presence or absence of CD1a and S100) was necessary. His IHC report showed that the specimen was positive for CD1a, CD68, S100, and Ki-67 (in 10% of cells). Therefore, the immunohistochemical profile of the patient with formalin-fixed paraffin-embedded tissue of the mandibular lesion showed Langerhans cell histiocytosis.

The patient was under treatment with prednisolone for 45 days. The results of the patient's follow-up (after 45 days) (Figures 5 and 6) are as follows: his teeth showed less mobility in comparison to the last check-up, he was still using DDAVP Nasal Spray, rashes and skin pruritus lowered. The patient will start chemotherapy for the treatment and will be under supervision for complete management.

3 | DISCUSSION

Paul Langerhans first described the Langerhans cell in 1868.[9] Langerhans cell histiocytosis (LCH), named in

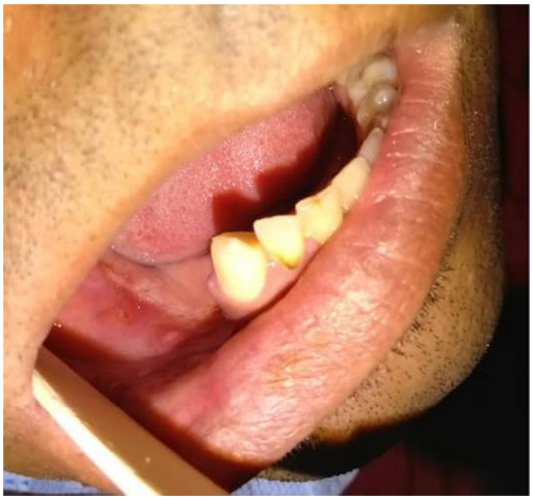

FIGURE 5 Clinical presentation of the posterior part of the right mandibular ridge, 45 days after the biopsy.

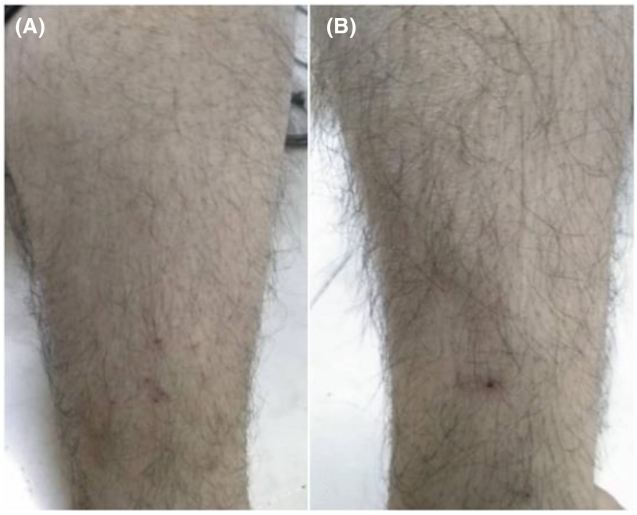

FIGURE 6 Clinical presentation of rashes (erythematous crusted macules) on the right (A) and left (B) legs of the patient with the sense of pruritus in those areas on the 45th day of follow-up.

1987, is a group of hyperplastic cellular diseases.[2] LCH is a rare disease; thus, it may be at a high risk of being under or misdiagnosed. For this reason, when an unclear osteolytic bone lesion is presented, LCH must be kept in mind.[10] Its pathological characteristic is the proliferation of myeloid dendritic cells reminiscent.[11] The proliferation of histiocytes and other inflammatory cells is observed in LCH. This can cause accumulation and pathological dissemination of histiocytes; therefore, the destruction of soft and hard tissues.

Although LCH was previously classified into three groups (eosinophilic granuloma, Hand–Schüller–Christian disease, and Letterer–Siwe disease), it is currently classified, based on the involved organs, into two groups: localized (single-system disease) and disseminated (multisystem disease).[4,7] The single-system class can again be divided into single or multiple sites. In multisystem LCH, two or more organ systems can be involved (with or without organ dysfunction); it can have low- and high-risk subdivisions based on the involvement of high-risk organs such as the spleen, lung, liver, and hematopoietic system. The single-system LCH, which was named "eosinophilic granuloma" in the past, is the benign form of LCH. Due to patient's age, the location of involvement varies. Primary sites for children are the skull, femur, ribs, vertebra, and humerus, and for adults: jaw, skull, vertebra, pelvis, extremities, and ribs.[11]

The etiology of this disease is controversial, but several theories have suggested the relation of infection (e.g., with the Epstein–Barr virus), environment, immunology, genetics (gene mutation and chromosomal instability), smoking (especially for isolated pulmonary LCH), and neoplastic process with LCH.[7,9–12] In this disease, lesional tissue cells have clonal proliferative characteristics, which may indicate that the innate lesion of LCH involving the patient is a tumor. Some scholars have considered LCH as a reactive and hyperplastic disorder; whereas, others have mentioned that LCH can be related to cytokine mediation, immunologic derangement, and viral infections.[2]

This rare condition can occur in all ages, but it mostly happens in children from 1–3 years of age. To this day, more than 50% of cases have been reported to age less than 10 years. Studies have shown that almost 30% of LCH cases have been in adults, but due to LHC's rareness, the incidence rate of LCH in adults is 0.18/100,000, whereas, 2–5 cases of this disease have been reported per one million children. Diabetes insipidus (DI) was the most common endocrine problem in the review of the English literature (present in 5%–50% of patients struggling with LCH). DI happens in 5% of patients simultaneously with the original disease and occurs in 25–33.3% of patients within 5 years of diagnosis.[3,4,7,9–11]

LCH can be self-limiting or locally recurrent, but systemic and high-risk cases may show a high rate of mortality. Due to the facts mentioned and that its first manifestations are mostly first orally, a dentist needs to diagnose this condition at its early stages, as soon as possible, and perform the optimal management and follow-ups based on international guidelines.

The symptoms of LCH in adults are based on the pattern of the organs it involves. The most common symptoms include local pain, weight loss, and fever. Diabetes insipidus, because it involves the pituitary gland, is one of the most important indicators of LCH.[7] The clinical manifestations of this disease can be different because of the differences in the age of onset, the Langerhans cells, and involved tissues and organs' proliferation rate. It can be easily misdiagnosed clinically. Therefore, bone marrow

TABLE 1 A review of LCH cases in the English literature.

Author	Age/sex	Symptoms, signs, and diseases	Microscopic features	IHC	Treatment
Lian C. Shenzhen[1]	34/M	"Pain, axillary ulcers, weakness, polyuria, alopecia, sexual dysfunction, weight increment, chest congestion, mastauxy, polydipsia, red rash"	"Granulocytes with vocules containing toxic particles, erythrocytes with polychromatic erythroblast with proliferating expanding pale area"	CD1a+ CD68+	CHOPE oral isotretinoin the operation of: "debridement + free skin transplantation + thigh skin graft operation"
Kim S. Busan[5]	35/M	"Pain, focal tenderness in the 6th left rib"	"Histiocyte proliferation, eosinophil infiltration"	CD1a+ S100+	Biopsy
Scolozzi P. Switzerland[2]	42/—	"Chronic fatigue, memory impairment, diminished libido, impotence, hair loss, gynecomastia, polyuria, polydipsia, panhypopituitarism, central DI"	"Gliosis with non-specific perivascular chronic inflammatory infiltration, diffuse infiltration of large pale-staining mononuclear cells with indistinct cytoplasmic borders and indented vesicular nuclei with a histiocytic appearance, infiltration composing a large number of eosinophils, plasma cells, and lymphocytes"	CD1a+ S100+	Stereotaxic biopsy
Luz J. Zurich[6]	46/M	"Tympanic effusion, right-sided hearing loss, DI centralis"	"Respiratory mucosa fragments with chronic and partially purulent inflammation with focal eosinophilia, mononuclear histiocytic cells with irregular nuclei admixed with small lymphocytes and eosinophilic granulocytes"	CD1a+ Langerin (CD207)+ S100+	Tooth extraction Chemotherapy with Cytarabin
Christopher Z. Florida[9]	54/M	"Acute low back pain, flank pain, constipation, left buttock pain radiating into left thigh"	"A lesion composed largely of lymphocytes and macrophages admixed with oval-shaped cells with grooved or indented nuclei with fine chromatin, inconspicuous nucleoli, slightly eosinophilic cytoplasm, occasional plasma cells and rare eosinophils in a background of fibrous stroma"	CD1a+ S100+ Langerin+	Open biopsy

Abbreviations: CHOPE, cytoxan, adriamycin, vincristine, prednisone, and etoposide; DI, diabetes insipidus; M, male.

and histopathological exams have to be performed if necessary. Most mouth-related findings include gingival enlargement, oral ulcers, and tooth mobility; thus, in cases where unclear osteolytic lesions in the jaw or diabetes insipidus centralis with unknown etiology are observed, LCH must be kept in mind, and a biopsy must be performed for a definite diagnosis.[10] LCH lacks pathognomonic characteristics radiographically and can show the same features as radicular cysts, periodontal disease, osteomyelitic lesions, and malignancies. LCH lesions are mostly seen as sharp and punched-out radiolucencies. When alveolar involvement is extensive, teeth can appear as "floating in the air".[3,4]

The prognosis of this disease can be based on the age of the patient, the number of organs that are involved, and the stage of the functional lesion. The prognosis of a single organ is proven to be better than multiorgans. Adult LCH has mostly shown a better prognosis; although, the prognosis in patients with lung, liver, spleen, and bone marrow involvement is poor; in these cases, the response to early treatment is relatively poor as well.[2]

Several cases of adult LCH have been reported until this day. We reviewed 5 cases as described in Table 1. They aged from 34–54 years, and all cases (instead of Scolozzi's[3] in which the gender of the patient was not mentioned), similar to our case, were males.

In common with Laz, Lian, and Scolozzi, and in contrast to Kim and Christopher, our patient had diabetes insipidus and was under treatment for this disease. In our case, Lian's showed high ALT and AST levels in the blood test. In Christopher, Scolozzi, Lian, Kim, Luz, and our case, inflammatory cell infiltration had been observed in microscopic evaluation. Kim, Scolozzi, Christopher's case, and our patient went under biopsy performance as a treatment and for histopathologic and IHC diagnosis. Unlikely, Luz's was treated with 12 cycles of chemotherapy and Lian's with combined treatment which is mentioned in Table 1; although, they both underwent biopsy for a definite diagnosis. Luz, Kim, Scolozzi, like our case, showed bone resorption and osteolytic radiolucent lesions in the bone. Similar to Luz, Scolozzi, Kim, Christopher, and Lian, our case showed positive CD1a and S100 in the IHC tests. Moreover, the IHC of our patient and Lian's were positive for CD68 and Ki-67 (in Luz's case, 20% of cells expressed the cell proliferation marker Ki-67).

The limitations of our study included the patient's busy schedule (therefore, a hard to follow-up condition) and lack of genetic examination.

As LHC's first manifestations are mostly oral and its prognosis in the early stages is well, it is important that this disease is diagnosed at early stages (before it progresses and turns fatal). When facing a patient with osteolytic lesions of the jaw with an unknown etiology or a patient with diabetes insipidus centralis that cannot be explained by any disease or disorder, dentists must keep LCH in adults in mind as a possible cause.[13] The differential diagnosis for LHC is other cutaneous histiocytosis, bone metastases, or malignant tumors. For its diagnosis, a biopsy of the lesion must be performed. The gold standard for its definite diagnosis is the positive result of CD1a and S100 in the IHC test. Different treatments are available for this disease (mentioned previously in this paper), and each of them may be used based on different factors of the patient, environment, and the disease. After the treatment, periodic follow-ups should be considered.

4 | CONCLUSION

LCH is a rare condition (especially in adults) with ulcerative and osteolytic bone features, related to the proliferation of histiocyte-like cells. Several factors are related to the occurrence and progression of this disease, but its etiology is still unknown. Lesions that clinically look similar to each other may be mistaken for each other. Therefore, it is important for us to know the clinical, radiological, histopathological, and immunohistochemical characteristics of each condition to diagnose the disease fast and correctly. Accurate diagnosis, choosing the best path of treatment for our patient's condition, and careful follow-ups are mandatory to provide successful patient management and prevent any progression in the patient's condition. Our case adds to the few cases of LCH in adults which have been reported in the English literature. It is essential to document new cases of LCH in adults in the English literature so that better and more accurate treatments will be introduced to prevent any neglect and further damage that they may cause.

AUTHOR CONTRIBUTIONS

All authors contributed to the study conception and design. Material preparation, data collection, and analysis were performed by Dr. Fatemeh Lavaee, Dr. Ali Dehghani Nazhvani, and Aylar Afshari. The first draft of the manuscript was written by Aylar Afshari and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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The authors have no conflicts of interest to declare that are relevant to the content of this article.

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Not applicable.

CODE AVAILABILITY

Not applicable.

ETHICS APPROVAL

Not applicable.

CONSENT TO PARTICIPATE

Available if requested.

CONSENT FOR PUBLICATION

Not applicable.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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