A Case of Epulis Granulomatosa With Clinical and Radiological Findings Resembling Langerhans Cell Histiocytosis

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ABSTRACT

Aim Langerhans cell histiocytosis (LCH) is a rare disease characterized by the proliferation of Langerhans cells. The differential diagnosis of LCH from epulis granulomatosa may be more difficult, given that the major symptoms of LCH are swelling and a floating tooth appearance. The purpose of this study was to provide a case of epulis granulomatosa that resembled LCH in the jaw bones based on clinical and radiological symptoms, as well as to warn against the possibility of misdiagnosis.

Case Report Smooth-surfaced, lobular, erythematous, sessile, and fibrotic tissue hyperplasias were seen in the maxilla anterior edentulous area and the mandible's left posterior region. The patient's radiographic findings revealed extensive alveolar bone destruction in the maxilla and mandible, as well as severe bone destruction in the posterior area of the left mandible, consistent with a floating tooth. Under local anesthetic, an excisional biopsy of the lesion was performed in the left posterior region of the mandible. Histopathological examination revealed that the patient had inflammatory fibrous tissue hyperplasia.

Discussion The clinical symptoms of LCH patients vary depending on their location and degree of involvement. When completing a full mouth examination on a patient, it is critical to detect soft tissue abnormalities as well as provide an accurate diagnosis and treatment plan.

Conclusion LCH, together with surrounding inflammatory alterations, should be considered in the differential diagnosis of osteolytic lesions of the jaw.

Keywords Differential diagnosis, Epulis granulomatosa, Histopathologic examination, Langerhans cell histiocytosis, Oral diagnosis

Introduction

Langerhans cell histiocytosis (LCH), formerly known as "histiocytosis X," is a rare condition characterized by strong and aberrant proliferation of bone marrow-derived immature myeloid dendritic cells-Langerhans cells (LCs) in the skin, bone, lymph nodes, and other organs (1,2). LCH is more prevalent in children with a male predisposition. The disease's incidence is reported to be 8.9 cases per million in children and 1-2 cases per million in adults (3, 4). LCH may form in a wide range of tissues including bone, lung, liver, skin, or endocrine systems, lymph nodes, neurological, and digestive systems (5, 6). In half of the LCH patients, bone involvement is seen, particularly in the mandibular region (5). LCH causes osteolytic lesions in these individuals, which are manifested by pain, edema, and tooth mobility (7,8).

LCH is formed by the clonal growth of immunophenotypical and functionally immature LCH cells, as well as eosinophils, macrophages, lymphocytes, and, on rare occasions, multinucleated giant cells. Histiocytosis X, eosinophilic granuloma, Letterer-Siwe illness, and Hand-Schüller-Christian disease are all names for LCH. Nevertheless, the recommended nomenclature is LCH since the diseased histiocyte common to all of these diagnoses was found to include typical Birbeck granules using electron microscopy (9, 10). There is controversy about whether clonal proliferation of LCH cells is caused by malignant transformation or by an immune trigger. Regardless of the mechanism causing clonal proliferation, the main therapy, if indicated, includes chemotherapeutic drugs (11).

Epulis is a clinical word that refers to a reactive localized connective tissue growth in the gingiva, the specific histological basis of which is uncertain. Epulis can affect individuals of any gender or age, however it is more frequent in women and young people (12). While epulis is classified differently in the literature, the most frequently recognized classification divides it into three major categories based on tissue origin: granulomatous epulis, fibrous epulis, and giant cell epulis. Granulomatous epulis, also known as gingival pyogenic granuloma, lobular capillary haemangioma of the gingiva, and epulis granulomatosa, is a smooth or lobulated exophytic lesion with a deep red or purplish color. Local irritants such as calculus, hormonal factors, certain medicines, and poor oral hygiene may all play a role in the development of granulomatous epulis (12, 13).

The treatment of granulomatous epulis is determined by the clinical symptoms. When the lesion is tiny, painless, and bleeding-free, removal of the causative irritants, clinical observation, and follow-up may be indicated. Although conservative excision, which reaches down to the periosteum and reserves the teeth, is the standard therapy, intrusive resection, which involves removing the neighboring teeth, should be performed to treat the vast lesion with significant loose teeth or the recurring lesion (13). Given that the main signs of LCH include edema and a floating tooth appearance, differentiating LCH from epulis granulomatosa

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may be more challenging. The aim of this case report was to provide a case of epulis granulomatosa in the jaw bones that resembled LCH based on clinical and radiological symptoms, as well as to warn against misdiagnosis.

Case Report

A 55-year-old female patient presented at the Department of Oral and Maxillofacial Radiology at Marmara University Faculty of Dentistry with complaints of teeth mobility and difficulty in chewing. The medical history was not significant for any medical conditions other than hypertension. Smooth-surfaced, lobular, erythematous, sessile, fibrotic tissue hyperplasias were seen in the maxilla anterior edentulous region and the maxilla left posterior region of the patient (Figure 1a, b). Panoramic radiography and Cone Beam Computed Tomography (CBCT) images revealed widespread alveolar bone destruction in the maxilla and mandible, as well as extensive bone loss in the left mandible posterior area consistent with the floating tooth. (Figure 2, 3).

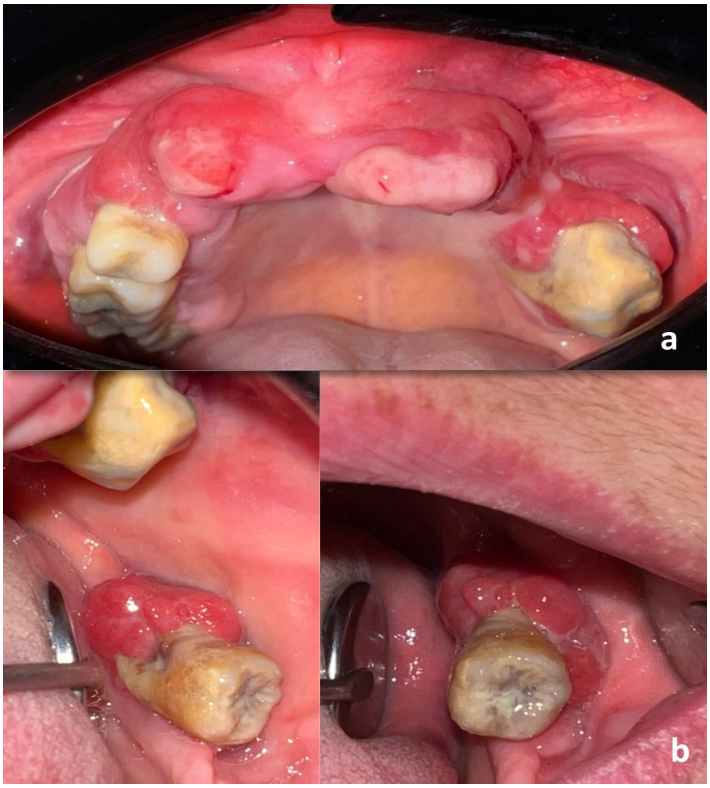

Figure 1: Smooth-surfaced, lobular, erythematous, sessile, fibrotic tissue hyperplasias were observed a) in the maxilla anterior edentulous region and b) the left posterior region of the maxilla.

Excisional biopsy of the associated lesion with tooth number 38 was performed under local anesthesia in the left posterior area of the jaw, and bleeding control was maintained. Histopathological examination revealed acanthosis and papillomatous tissues in the stratified squamous epithelium covering the surface, as well as a fiber-rich connective tissue with intense lymphocyte and plasma cell infiltration beneath it. Histopathologically, the diagnosis is inflammatory fibrous tissue hyperplasia. When the patient arrived at the control, he showed improvements in condition week after excision (Figure 4). The patient is currently stable and asymptomatic for oral lesions, with no new complaints of tooth mobility or discomfort.

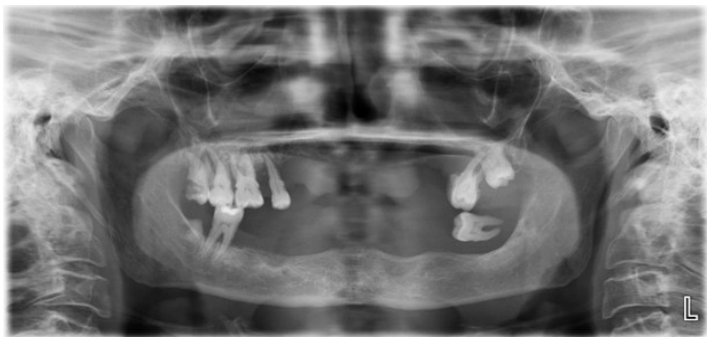

Figure 2: Panoramic radiography revealed widespread alveolar bone destruction in the maxilla and mandible, as well as extensive bone loss in the left mandible posterior area consistent with the floating tooth

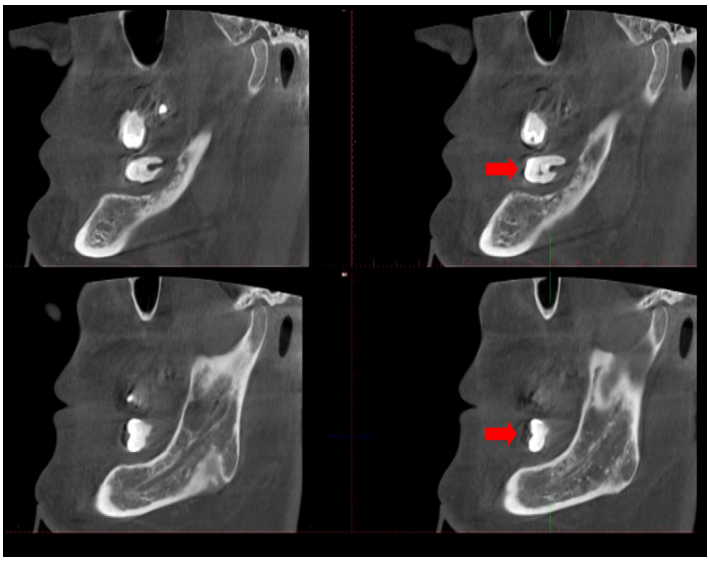

Figure 3: Extensive bone loss in the left mandible posterior area consistent with the floating tooth (red arrow) in CBCT.

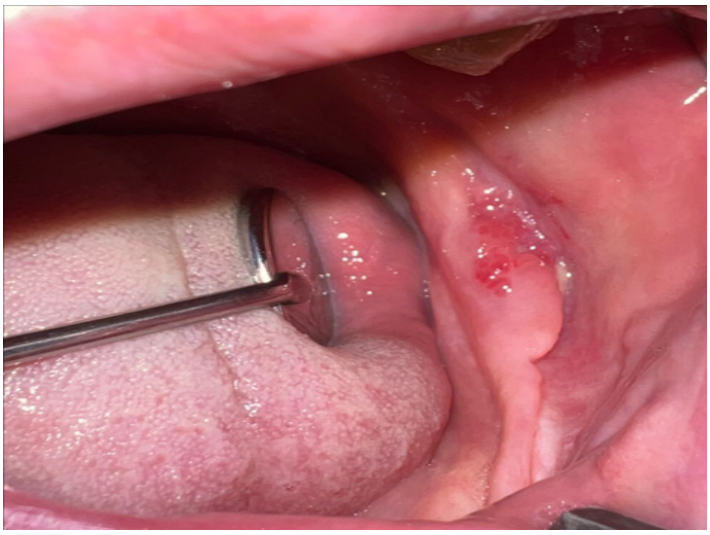

Figure 4: Recovery after excision of the lesion.

Discussion

The term histiocytosis X was used to refer to a group of clinopathologic diseases defined by the proliferation of histiocyte-like cells. Langerhans cells have been identified as the characteristic histiocytic cells present in this lesion, and the disorder is now known as LCH (1-3, 10)

LCH can affect any bone, but mainly the skull (1-3, 14, 15). It is distinguished by a single or several lytic bone lesions (14, 16, 17). Lesions may grow in size and number or merge, and oral alterations are frequently the first clinical manifestation of the disease (15, 17). This circumstance highlights the critical function of dentists in diagnosis and treatment. The mandible is impacted more frequently than the maxilla, and posterior parts are afflicted more frequently than anterior regions (14, 17, 18). LCH lesions in the alveolar bone are often numerous, whereas intraosseous lesions are single (14, 18). In the affected regions, clinicians may notice pain, sensitivity, gingival necrosis, and swelling (2, 3, 18, 19).

Clinical-radiological-pathological evidence should be used to determine the diagnosis (1-3). The characteristic presentation of LCH in the jaws frequently leads to tooth loosening or early exfoliation, as well as premature eruption of permanent teeth. In these circumstances, a differential diagnosis should include juvenile or diabetic periodontitis, hypophosphatasia, leukemia, cyclic neutropenia, and metastatic malignant neoplasms. Periapical lesions may resemble a periapical cyst or granuloma. Isolated radiolucent lesions in the jaws should be distinguished from odontogenic tumors and cysts, and many well-circumscribed radiolucencies may indicate multiple myeloma (10, 17-19). LCH's primary diagnostic imaging modalities are computed tomography and magnetic resonance imaging. In order to make a definite diagnosis, a biopsy and an immunohistochemistry investigation are performed (14, 20). As compared to computed tomography, CBCT is a favored imaging tool for evaluating bone destruction in these patients due to advantages such as low radiation dosage, good resolution, less time and lower cost (20-22).

67% of oral symptoms involve bone tissue, specifically the posterior mandible (5, 23-27). They might be isolated or many in numbers, and they can range in severity from mild to severe. Periodontal bone loss appears as "floating teeth" on X-rays. Soft tissue lesions can be gingival ulcerations or enlargements, as in the present case (23, 25). Periodontal disease and squamous cell carcinoma are major differential diagnoses for alveolar lesions. The differential diagnosis of intraosseous lesions includes metastatic malignant neoplasms and malignant tumors (28). In general, both benign and malignant bone lesions of the oral cavity and soft tissue should be evaluated in the differential diagnosis (29).

When observed in the oral cavity, LCH presents a substantial difficulty for the dental professional since some clinical characteristics of the disease mirror more frequent disorders such as periodontal disease, malignancies, and granulomatous or ulcerative lesions (2, 6, 25). As a result, the clinical and radiological results of the reported patient revealed advanced periodontitis, which may have suggested near-total extraction. Therefore, reevaluating the patient and performing an incisional biopsy for histological analysis assisted the authors in avoiding disease misdiagnosis and perhaps incorrect therapy.

LCH is treated by surgical curettage and bone grafting, as well as low-dose radiation, chemotherapy, and local steroid treatment into the lesion (15, 22). The treatment technique is governed by the patient's age, the location of the lesion, the number of implicated bones and lesions, the size of the lesion, and the disease's natural course (22, 30). Since jaw lesions have a low recurrence probability, surgical treatment is typically favored (1). After therapy, the patient should be regularly monitored for an extended period of time. The prognosis is determined by the patient's age and the number of organs affected. Those with LCH who show the first signs of the disease at a young age have a worse prognosis than children who get LCH later in life (19-25).

Conclusion

The clinical symptoms of LCH patients vary depending on their location and degree of involvement. When completing a full mouth examination on a patient, it is critical to detect soft tissue abnormalities as well as provide an accurate diagnosis and treatment plan. LCH, together with surrounding inflammatory alterations, should be considered in the differential diagnosis of osteolytic lesions of the jaw.

Declarations

Author Contributions: *Conception/Design of Study- F.N.P.; Data Acquisition- S.Y.U., G.K.; Data Analysis/Interpretation- F.N.P. , S.Y.U., V.O.; Drafting Manuscript- G.K., S.Y.U.; Critical Revision of Manuscript- F.N.P.; Final Approval and Accountability- F.N.P.; Material and Technical Support- V.O.; Supervision- F.N.P.*

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