La Prensa Medica Argentina

Case Report

Oro-Periodontal Manifestations of Langerhans Cell Histiocytosis (Eosinophilic Granuloma)

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Abstract

A 25-years-old man suffering from tooth mobility and dental loss was seen at the Clinic of the Department of oral medicine and Periodontology, faculty of dentistry, Mansoura university, Egypt. Investigated revealed clinical and radiographic with radiolucent images massive diffuse alveolar bones around of both molar and premolar areas in the mandible, mirror-like image and the posterior teeth gave an image of "floating teeth.

Taking into consideration, special investigation biopsy sample: were performed revealed a Routine Haematoxylin and Eosin (H and E) stained slides revealed moderately cellular lesion characterized by Langerhans cells accompanied by variable amounts of non-neoplastic reactive mixed inflammatory elements that include eosinophils, neutrophils, macrophages, lymphocytes, and plasma cells.

The Langerhans cells are recognizable morphologically by eosinophilic to clear cytoplasm and characteristically oval grooved "coffee bean" nuclei. This case presents several special features. First, the eosinophilic granuloma (Langerhans cell histiocytosis) has been rarely. Second the association between the periodontal disease and eosinophilic granuloma (Langerhans cell histiocytosis) has been rarely reported in the literature.

Keywords: Periodontal disease; Eosinophilic granuloma; Langerhans cell histiocytosis

Introduction

Langerhans cell histiocytosis (LCH), which was previously termed “histiocytosis X,” is a rare clonal disorder characterized by the proliferation of clonal CD1a-positive immature dendritic cells (LCH cells) in the skin, bone, lymph nodes and other organs. The clinical manifestations of LCH vary from a self- limiting single bone disease to rapidly fatal disseminated disease. However, LCH usually follows a chronic course and reactivations often occur. This can result in permanent consequences, such as orthopedic abnormalities, the development of central diabetes insipidus (CDI), and neurodegenerative central nervous system (ND-CNS) disease. LCH was originally described as three different entities, namely, eosinophilic granuloma, Hand–Schuller– Christian disease, and Letterer–Siwe disease. However, at present, LCH is classified as single-system (SS) vs. multisystem (MS) and unifocal vs. multifocal disease. MS disease is classified into two groups depending on whether risk organs (RO), namely, the liver, lung, spleen and bone marrow, are involved. SS disease, RO-negative MS disease, and RO-positive MS disease are almost equivalent to eosinophilic granuloma, Hand–Schüller–Christian disease, and Letterer–Siwe disease, respectively [1,2].

The purpose of this study is to present a case history of adult patient with Langerhans' cell histiocytosis of the variety eosinophilic granuloma who presented oral lesions.

Case Report

A 25-year-old male patient was seen at the Clinic of the Department of oral medicine and Periodontology, faculty of dentistry, Mansoura university, Egypt with the main complaint of tooth mobility and dental loss.

The past dental history revealed that the patient had a previous minor mandibular trauma in the anterior region about 4 years earlier 2 years before being seen at the periodontic clinic the patient noticed tooth mobility and spontaneous gingival bleeding as well as dental loss during mastication, which caused him to seek dental care. No past medical history. In clinical examination, complete blood picture, liver function, and thyroid function investigation revealed all are normal.

Intraoral examination revealed several teeth loss, severe tooth mobility, deep periodontal pockets in all sextants, an amount of dental plaque, spontaneous gingival bleeding, gingival recession, increased gingival volume.

Panoramic and cone-beam radiographs showed radiolucent images massive diffuse alveolar bones around of both molar and premolar areas in the mandible, mirror like image and the posterior teeth gave an image of floating teeth (Figures 1 and 2)

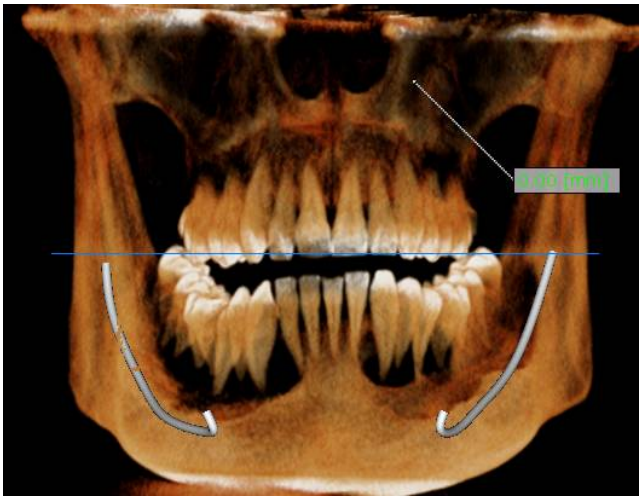

Figure1: Panoramic x-ray.

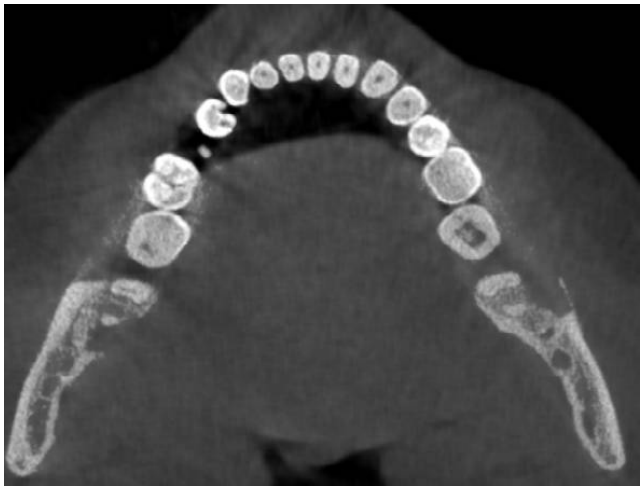

Figure 2: Cone-beam.

In order to investigate a possible genetic influence, the patient was further questioned about the disease occurrence in his family. No familial occurrence was reported.

Microscopic examination of the biopsy sample (incisional biopsy from represented area of the lesion): Routine Haematoxylin and Eosin (H and E) stained slides revealed moderately cellular lesion (Figure 3) characterized by Langerhans cells accompanied by variable amounts of non-neoplastic reactive mixed inflammatory elements that include eosinophils, neutrophils, macrophages, lymphocytes, and plasma cells (Figures 4 and 5).

The Langerhans cells are recognizable morphologically by eosinophilic to clear cytoplasm and characteristically oval grooved "coffee bean" nuclei (Figure 6).

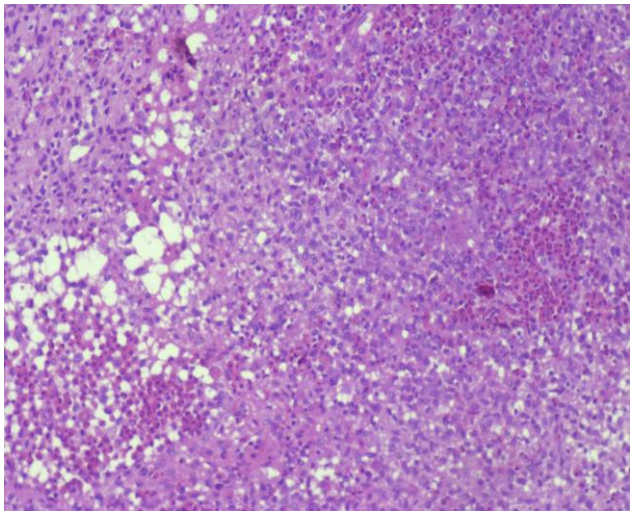

Figure 3: H & E showing moderately to hyper cellular lesion (x20).

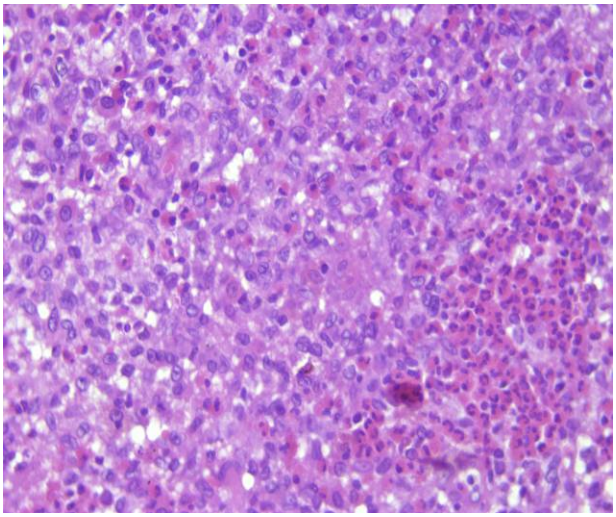

Figure 4: H & E showing moderately to hyper cellular lesion with numerous reactive inflammatory infiltrate as well as larger cells with cleaved nuclei (x40).

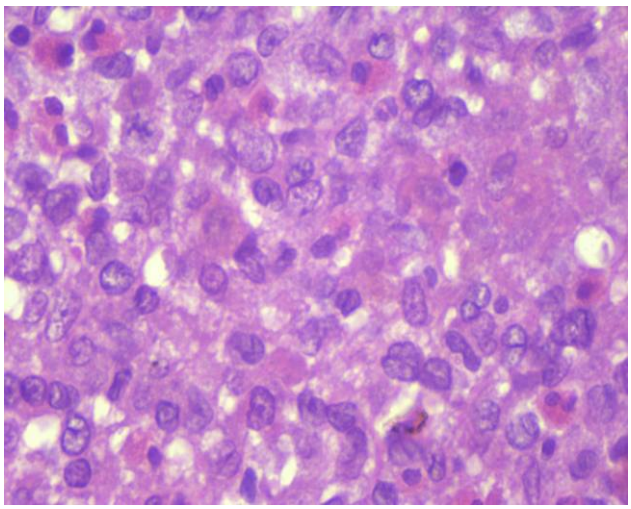

Figure 5: H & E showing numerous eosinophils as well as larger cells with cleaved nuclei (x100).

Immunohistochemical staining: Formalin-fixed paraffin-embedded sections of the lesion were cut into 4 microns thick sections. Antigen retrieval was performed by using EDTA. The slides were then incubated 30 minutes with monoclonal anti-Human CD1a (prediluted, clone 010, Dako), Rabbit Polyclonal anti-S-100 protein (prediluted, iso type N/A, Genemed) and anti-Human CD68 (prediluted, clone KP1, Dako). This is followed by biotinylated secondary antibody and streptavidin-peroxidase conjugate. DAB was used as chromogenic substrate. Brown staining was identified as positive.

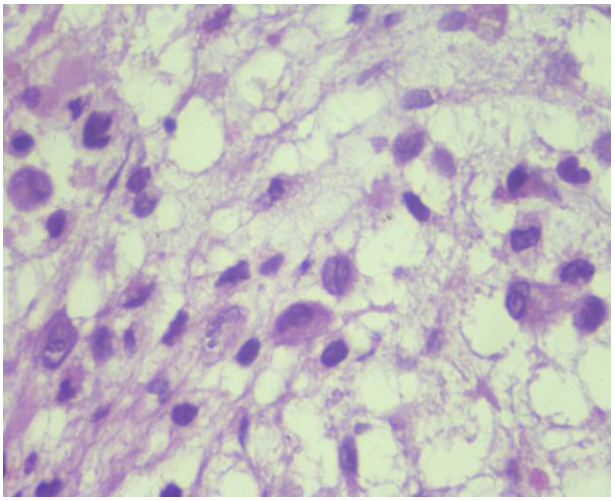

Figure 6: H & E showing cells with eosinophilic to clear cytoplasm and characteristically oval grooved "coffee bean" nuclei.

Interpretation of Immunohistochemical staining: For CD1a, a positive reaction is localized to cell membrane (more specific) (Figures 7 and 8). For CD68, positivity is indicated as a distinct brown membranous staining. For S100 protein, a positive reaction is seen as brown cytoplasmic staining (Figures 9 and 10).

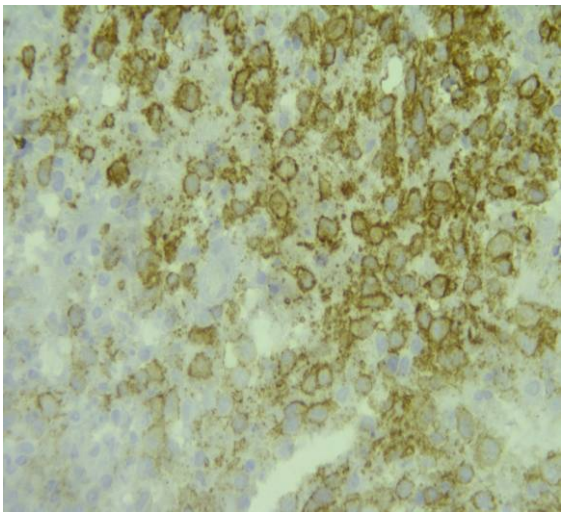

Figure 7: Membranous immunoreactivity of the Langerhans cells for CD1a (x40).

Diagnosis

Dependent on biopsy and Immunohistochemical characteristics which include cell surface CD 1a and S-100, Langerhans cells are prominent. Prognosis is good for the patient with treatment, almost all such patients survive. Morbidity and mortality are increased in patients with multi organ involvement. With treatment, the overall survival rate for patients with the multi-organ disease is about 80%. Death is more likely among at-risk patients who do not respond to initial therapy. Disease recurrence is common. A chronic remitting and exacerbating course may occur, particularly among adults.

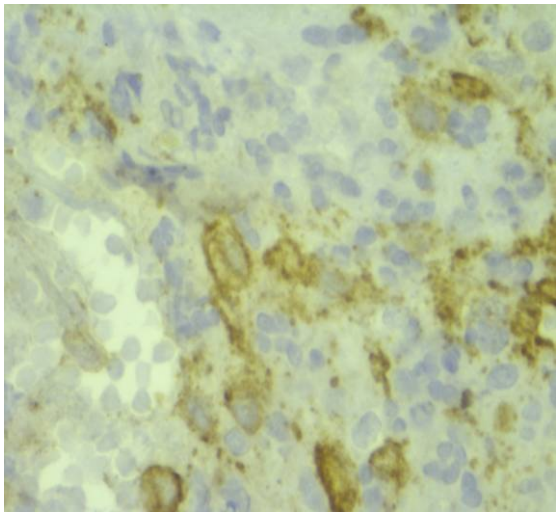

Figure 8: Membranous immunoreactivity of the Langerhans cells for CD1a (x100).

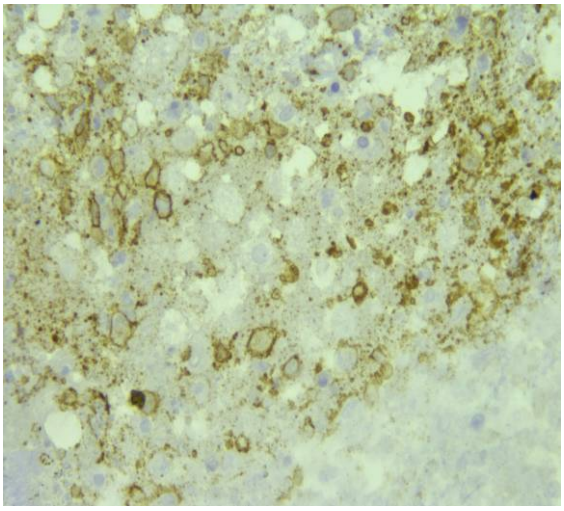

Figure 9: Membranous immunoreactivity of the Langerhans cells as well as reactive histiocytes in the background for CD68 (x40).

Treatment

1. Supportive care
2. Receiving chemotherapy by consult
3. Corticosteroid therapy
4. Methotrexate
5. Follow up

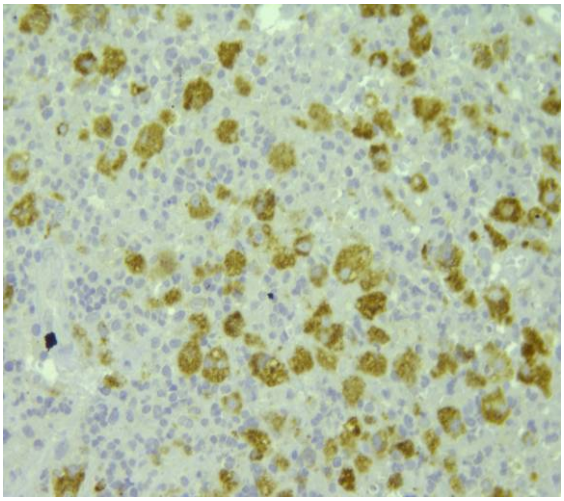

Figure 10: cytoplasmic immunoreactivity of the Langerhans cells for S100 protein (x40).

Discussion

Langerhans' cell histiocytosis is a rare disease, the etiology and pathogenesis of which remain unknown. A variety of etiological factors have been proposed including immunologic reactions, viruses, bacteria and genetic influence [1,2]. Langerhans' cells are dendritic bone marrow-derived cells situated suprabasally in most stratified squamous epithelia. They are thought to act as antigen-presenting cells during induction of immune responses. Besides having functions which are similar to other dendritic cells and macrophages, Langerhans' cells are specialized and able to migrate, playing an important role in antigen presentation to the T-lymphocytes. It has been suggested that they play a key role in the induction of immune responses and also in immunopathological reactions taking place at cutaneous and/or mucosal levels. Langerhans' cells may represent a "first line" of sensitization of the immune system, leading to clearance of the antigen or to pathological phenomena. It is not known, however, what leads to the proliferation of these cells in the histiocytosis lesions [2].

Because of the microscopic features which are presented in Langerhans' cell histiocytosis, an inflammatory etiology has been proposed. A bacteriologic origin has also been suggested although no specific causative microorganism has been identified in histiocytosis lesions [3].

The oral manifestations of Langerhans' cell histiocytosis may be the first and/or the single sign of the disease. These manifestations present distinct characteristics when compared to those found in other locations. This is probably due to the unique anatomy of the tooth-supporting structures, and to the bacterial milieu of the mouth which predisposes to secondary infections. That is why lesions of histiocytosis have been erroneously diagnosed as periodontal disease. A number of signs can be clinically observed such as severe periodontal loss, erosion and ulceration of the mucosa, bleeding gingiva, purulent exudate, mobility and premature exfoliation of the teeth, precocious eruption of complete dentition and ectopic eruption of permanent molars [3]. The radiographic features comprise solitary or multiple areas of well-defined radiolucency in the alveolar bone mimicking severe periodontal disease. Destruction of lamina Dura provides the radiographic appearance of "floating teeth" which has been considered, by some authors, as the most representative radiographic feature of the disease [4].

Eosinophilic granuloma is considered to be the mildest form of the disease and appears to be less aggressive than the other varieties. The destruction of the alveolar bone is thought to be one of the characteristic signs of eosinophilic granuloma and, in this location, the disease may simulate severe localized periodontitis or periapical infection [5,6]. This may be diagnosed as periodontal disease because it shares similar clinical and radiographic characteristics with severe periodontal disease and the microbiologic composition is also similar to the micro biota found in periodontal disease. Osseous lesions are an important feature of eosinophilic granuloma. When these lesions occur in children, they may be mistaken for prepubertal periodontitis, and when they occur in young adults, they may resemble rapidly progressive periodontitis, which is why a differential diagnosis between eosinophilic granuloma and early-onset periodontitis must be established [7].

Conclusion

In summary we describe a case of eosinophilic granuloma (Langerhans cell histiocytosis) has been rarely and the association between the periodontal disease and eosinophilic granuloma (Langerhans cell histiocytosis) has been rarely reported in the literature.

References

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