

Langerhans cell histiocytosis (Histiocytosis X) – in the mandible

Histiocitose das células de Langerhans (Histiocitose X): na mandíbula

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ABSTRACT

This article reports a case of 65 year-old man consulted in a private radiology dental clinic for a panoramic radiography, where was indicated a radiolucent area, extending into the periapical region of the teeth 35 to 43 to the base of the mandible. Radiography diagnosis was difficult. The immunohistochemical examination showed immunoreactivity for the proteins CD 68, S-100 and CD 1a. Histopathological diagnosis was obtained for Langerhans cell granuloma (histiocytosis X). Langerhans cell histiocytosis is caused by the abnormal proliferation of histiocytes, previously called Histiocytosis X. It is a rare disease that preferentially affects children and young adults with a predilection for males and can affect different parts of the body.

KEYWORDS

Adult; Histiocytosis; Langerhans; Mandible.

RESUMO

Este artigo relata o caso de um homem de 65 anos que procurou uma clínica de radiologia dental particular para a realização de uma radiografia panorâmica. Foi detectada uma área radiolúcida na região apical dos dentes 35 a 43 que se estendia até a base da mandíbula. O exame imunoistoquímico da lesão mostrou proteínas reativas como a CD 68, S-100 e CD 1a. O diagnóstico do histopatológico foi de Granuloma de Células de Langerhans (Histiocitose X). A Histiocitose das células de Langerhans é causada pela proliferação anormal de histiócitos. É uma doença rara que preferencialmente acomete crianças e adultos jovens e tem predileção pelo sexo masculino, podendo afetar diferentes partes do corpo.

PALAVRAS-CHAVE

Histiocitose de células de Langerhans; Mandíbula.

INTRODUCTION

Langerhans cell histiocytosis (LCH), previously called histiocytosis X, refers to a spectrum of disease characterized by idiopathic proliferation of histiocytes producing focal or systemic manifestations [1]. First studies suggested some causes as a result of the proliferation and dissemination of abnormal histiocytic cells of the Langerhans cell system. [2]. In fact, the etiology of LCH is unknown.

Other authors report that LCH cells are clonal and a cancer associated mutation (BRAFV600E) was found in more than half of investigated specimens, indicating that LCH may be more a neoplastic (but not a malignant) disease than a reactive disorder. Although, these cases represent a minority of all LCH patients [3,4,5].

Affects more children and young adults (1,2,6,7) with a predilection to the males gender (52%), but can develop in women [8]. According to Histiocytosis Association is estimated that LCH occurs in 1-2 adults per million people (9). Several regions of the body can be affected as bones, skin, liver, spleen, lymph nodes, lung and brain [6,10].

LCH is used to describe three reticuloendotheliosis disorder [11]. Eosinophilic granuloma (EG) localized form and can be monostotic or multiple lesions. Hand-Schüller-Christian syndrome (HSC) is chronic and a widespread lesion formed by foam reticuloendothelial cells and has a triad of signs: cranial bone lesions, exophthalmos and diabetes insipidus. And finally Letterer-Siwe disease (LSD) is a disseminated acute beginning and is less than 3 years old. LSD rarely affect the jaws, its development is rapid and fatal. [12,13,14].

Oral lesions may be the earliest manifestation. Panoramic radiograph of the jaws is an excellent for detection of jaw lesions. The lesions often develop quickly and may appear as a painful palpable mass [14]. Lesions of the oral mucosa may precede evidence of disease

elsewhere. The mandible is affected twice as often as the maxilla [16].

In this paper, we report a case of a 65-years-old man patient who presented to a radiolucent lesion in anterior region of the mandible in panoramic routine.

CASE REPORTED

Male patient, leucodermic, 65 years old, farmer and smoker, presented in Panoramic radiography (Figure 1) multiloculated radiolucent area, extending into the periapical region of the teeth 35 to 43 to the base of the mandible. Clinically patient presented increased volume of fibrous consistency, asymptomatic, localized buccal surface of the alveolar ridge in the mental region. CT scan performed on Multi-Slice (Figure 2) revealed presence of multiloculated hypodense area, extending from the region of tooth 35 to 43, to the base of the mandible, measuring 40 x 24 x 21 mm., with destruction of the cortical bone buccal and lingual side in the region involved (Figure 3). Hyperdense subcutaneous points in the region of the buccinator muscle, anterior to the right masseter. And the structure of hyperdensity irregularly located at the base of the skull, compatible with projectile fragments of a firearm. Presence of depression surface irregular, located in the alveolar crest in the region of tooth 46 compatible bone defects. Calcification tonsils side left and right. Incisional biopsy was performed in multiple fragments of soft tissue (products of curettage) of varying shapes and sizes, color brown / blackish, flaccid consistency and measuring entire 2.0 x 2.0 x 2.0 cm. Biopsy revealed tissue rich in mononuclear cells of large histiocytic aspect of nuclei ranging from round the grooved, interspersed with intense eosinophilic inflammatory infiltrate. It was also observed chronic inflammatory cells lymph myeloma and hemorrhagic areas. The immunohistochemical examination shows immunoreactivity for the proteins CD 68, S-100 and CD 1a (Figure 4). Histopathological diagnosis was obtained for Langerhans cell granuloma (histiocytosis X).

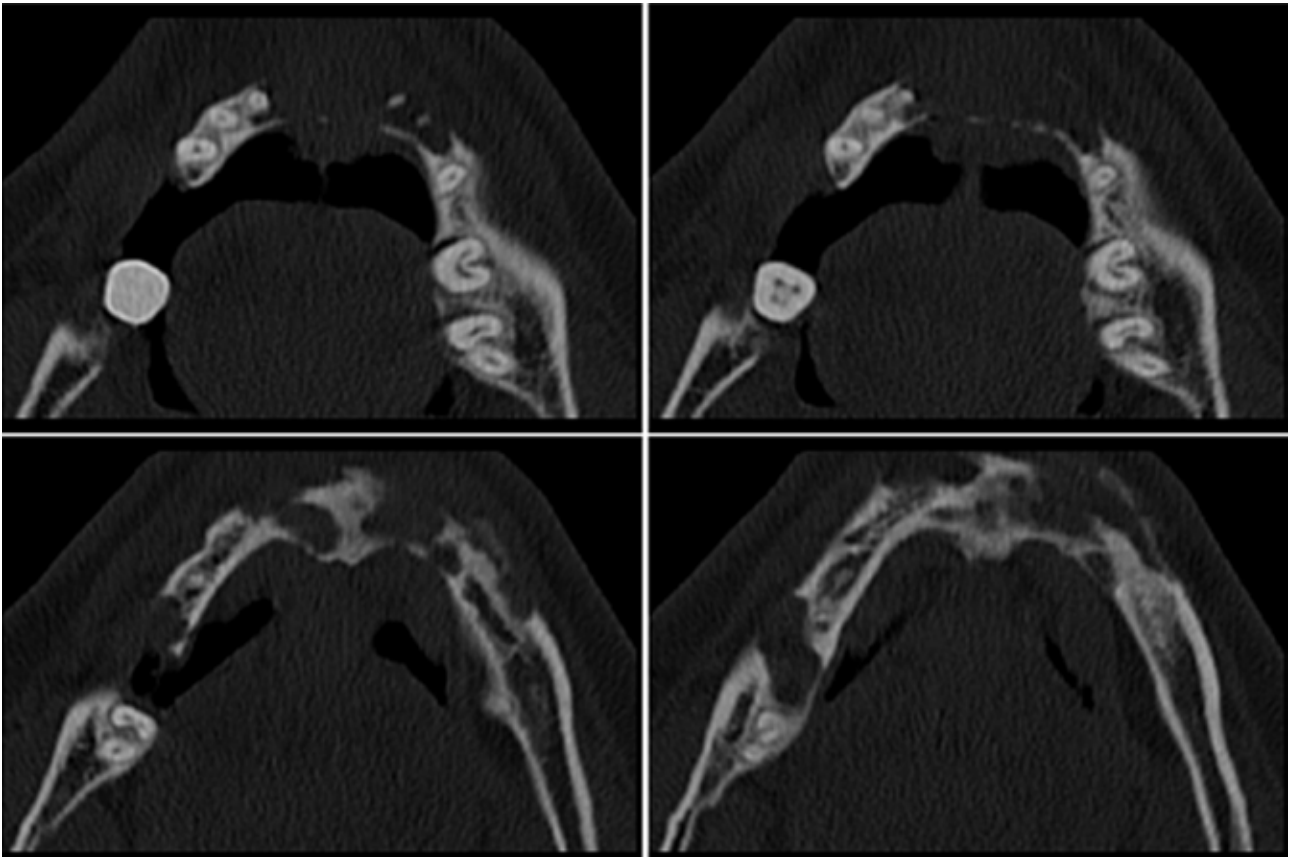


Figure 1 – Norm Axial CT of the mandible.

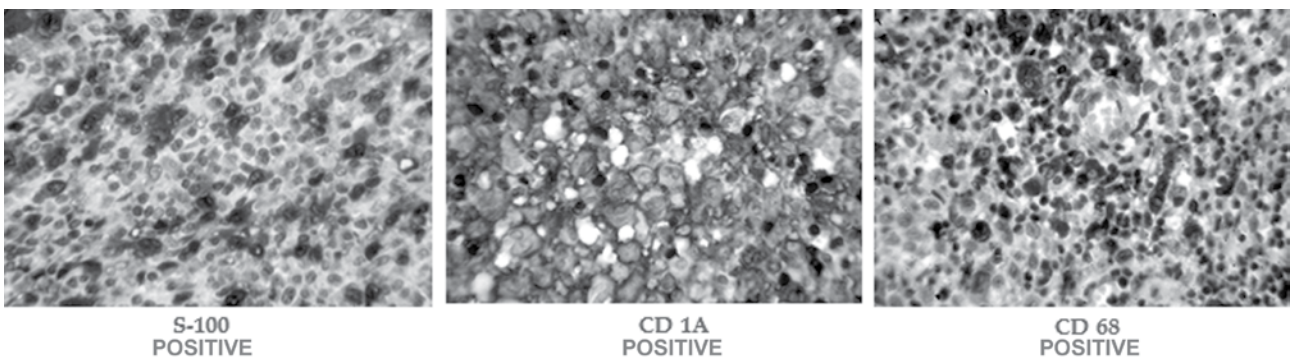


Figure 2 – Sections were incubated with the antibodies listed above and the reactions were verified by the detection technique of polymer-conjugated secondary antibodies and peroxidase.

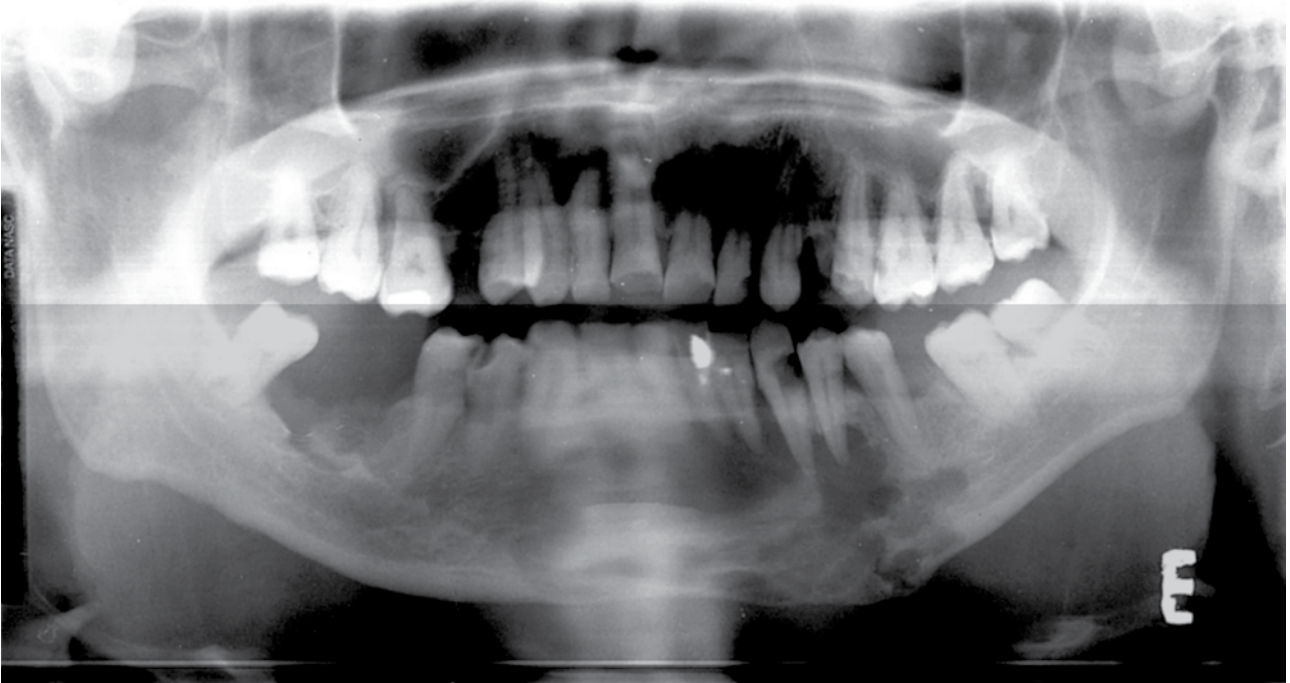


Figure 3 – Panoramic radiograph.

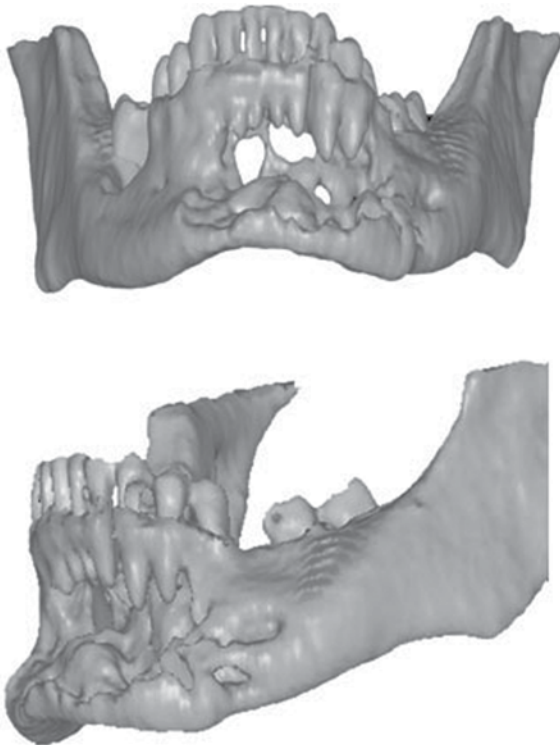


Figure 3 – Reconstruction of the mandible in 3D CT.

DISCUSSION

LCH is a rare disease known to affect children and young adults, their diagnosis is difficult clinically and radiographically by the similarity of the lesions with benign and malignant tumors [1].

In the case reported the patient was elderly man and according to the Histiocytosis Association of America estimates that LCH occurs in 1-2 adults per million people. According to another registry based in Germany, it is estimated that 46% of adult patients had bone lesions, 17% skin, 7% pituitary, 4% liver/spleen, 2% brain, and 2% gastro intestinal tract. In children occurs in 1:200.000 (9).

The lesion on the panoramic radiograph of this case shows radiolucent, multilocular, moth eaten appearance, alternating areas with more or less radiolucency, leading to suspicion of rupture of bone plate (confirmed on CT). Lesion causing cortical expansion in the base of the mandible, cortical destruction in the lower

jaw and cuts in the same region of the mental foramen. It affects the region of 35 to 45 teeth. The region of tooth 46, in principle, has no connection with the previous injury (Figure 3).

The typical radiographic appearance of the EG jaw lesions was primarily that of a destructive radiolucent process, usually with well-defined margins. Alveolar bone loss was variable, but frequently the destruction of the supporting alveolar bone was severe teeth appeared to be “floating in air” (appearance on anterior region of mandible (Figure 1) [15,17]. The mandible (10%-20%) is the most common site of all EG cases and posterior region will more involve

than the anterior regions [10,13,18]. The jaw lesions of the acute and chronic disseminated diseases were generally smaller in size and had less distinct borders than did the eosinophilic granuloma lesions [15].

The differential diagnosis of eosinophilic granuloma is difficult for its variable appearance [7]. The radiographic differential diagnosis included neoplastic odontogenic, infiltrative nonodontogenic, and primary malignant processes [13]. Bone lesions of LCH can be multifocal and unifocal [19]. The differential diagnosis includes various pathologies as shown in table 1.

Table 1 – Differential diagnosis includes various pathologies

Oral Lesions	<ul style="list-style-type: none"> - Advanced periodontal disease - Periapical abscess 	
Bone Lesions	<ul style="list-style-type: none"> - Osteomyelitis - Ewing’s sarcoma - Brown tumor hyperparathyroidism - Multiple odontogenic keratocyst - Multilocular cyst - Leukemia - Lymphoma 	
Multifocal LCH	Children	Adults
Unifocal LCH	<ul style="list-style-type: none"> - Metastatic neuroblastoma - Intrabony hemangioma - Fibrous dysplasia - Hemophilic pseudotumor - Epidermoid cyst - Giant cell granuloma 	<ul style="list-style-type: none"> - Osteolytic metastasis - Multiple myeloma - Myxoma - Ameloblastoma - Osteogenic sarcoma - Fibrosarcoma

The treatment depends on the size and structures that have been involved. Consist of conservative management (observation), surgical intervention (such as incisional biopsy, curettage, or total excision), radiation therapy, chemotherapy, administration of intraosseous steroids, on some combination of these techniques. Surgical curettage and topical injection of steroid are usually the preferred treatments for solitary jaw lesions. Chemotherapy is rarely used in patients with solitary bone lesions [1,3,10].

The prognosis for patients with LCH is generally favorable 92% of the patients in whom results of clinical follow-up were available had good outcomes. However, patients with multifocal disease more often had waxing and waning of symptoms and required longer courses of chemotherapy than those with localized disease [20].

CONCLUSION

LCH is a rare disease and its cause and pathogenesis is not clear yet. Often affects children, so appearance at older ages is relatively rare. When involved in the jaw, studies have shown that has a predilection for the posterior region, makes this case more uncommon. Radiograph and the biopsy findings with immunohistochemical examination positive for the proteins CD 68, S-100 and CD 1a are diagnostic. Radiological distinction from malignant is sometimes difficult. Treatment depends on the type of lesion and the affected region. Prognosis for Eosinophilic granuloma is the most favorable.

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