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Case report

Rare case of unifocal Langerhans cell histiocytosis in four-month-old child

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ABSTRACT

Langerhans cell histiocytosis (LCH) comprises a group of disorders, the common feature of which is Langerhans cell proliferation. The clinical presentation is highly varied. The severity and prognosis of the disease are dependent on the type and extent of organ involvement. This paper reports a rare case of a four-month-old white male with unifocal LCH limited exclusively to the mandible, discussing the diagnosis, radiographic and immunohistochemical aspects, treatment and monitoring multidisciplinary of the case.

1. Introduction

Langerhans cell histiocytosis (LCH), formerly known as histiocytosis X, is characterized by an abnormal proliferation of histiocytes of unknown cause [1]. It is a rare disease that affects five children per million [2]. The disease primarily occurs in bone, but can be associated with a clinical spectrum ranging from a solitary lytic bone lesion to a multi-systemic disease, including extra-osseous manifestations, with a significant mortality rate [3,4].

There are three clinical types of LCH [5,6]. The unifocal subtype (single system, single site), previously referred to as eosinophilic granuloma, commonly affects the bones (up to 80%), lymph nodes or lungs as the primary target [6]. Bone lesion is common in older children, whereas adults tend to have lung involvement [7]. The multifocal subtype (single system, multiple sites) affects several sites in any particular organ system [6]. This subtype has been referred to in the past as Hand–Schuller–Christian disease. The oral cavity and cranial lesions are commonly involved and are often present with skull lesions, diabetes insipidus and exophthalmos [6]. This subtype mainly affects children from two to six years of age and is fatal in 15% of patients [7]. The multiple-organ-system subtype, previously referred to as disseminated histiocytosis or Letterer–Siwe disease, affects multiple sites in different organ systems, can be seen in the first year of life and has the worst outcome [6]. Manifestations may include persistent fever, irritability, anorexia, super-infection, diarrhea, pancytopenia, sepsis, failure to thrive and purpuric rash [3]. The oral cavity, skin lymph nodes, brain, lung and liver are other organ systems that may be affected [6].

The treatment of LCH is either by surgical excision, chemotherapy, radiation therapy or a combination of these modalities [8]. Prognosis depends mainly upon the involvement of multiple organ systems. Young patients presenting with disseminated disease and organ dysfunction have the highest mortality rate [9].

This article describes a rare case of unifocal LCH (eosinophilic granuloma) in the mandible of four-month-old white male, discussing the diagnosis, radiographic and immunohistochemical aspects and treatment.

2. Case report

A four-month-old white male was referred to our department with the complaint of gingival lesion lasting 45 days. The patient was in good general health and his past medical and dental history did not reveal any significant events. The patient had been medicated by a pediatrician with a topical anti-inflammatory and antibiotic (Gingilone®), topical anti-fungal (Nystatin®) and systemic non-steroidal anti-inflammatory drug (ibuprofen®), with no response. The extra-oral examination revealed that the patient was ruddy, hydrated, eupneic, anicteric and without noteworthy facial asymmetry. The intra-oral examination revealed a lesion in lower gingival mucosa measuring approximately 1.5 cm × 1.0 cm

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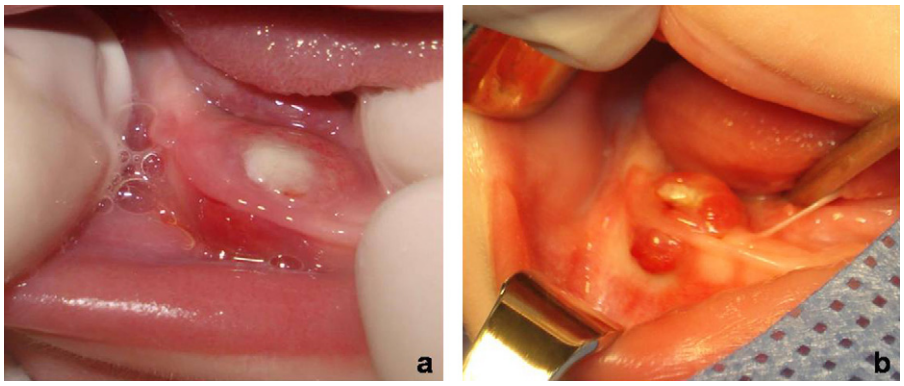

Fig. 1. (a) Clinical aspect of lesion in lower gingival mucosa with erythematous color and a central area of ulceration. (b) Two weeks after the first exam the lesion exhibited a proliferative growth.

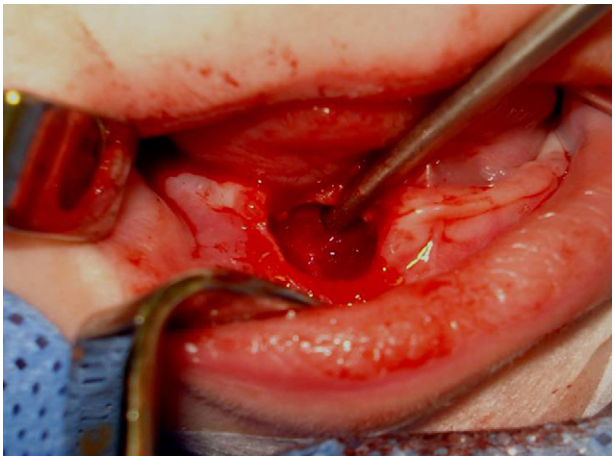

Fig. 2. Surgical view revealing a single bone cavity.

in its greatest diameter, covered by mucosa, with an erythematous central area of ulceration, fibrous consistency upon palpation and no bleeding (Fig. 1a). The clinical diagnostic hypotheses were pyogenic granuloma, giant cell tumor of the newborn and Langerhans cell histiocytosis.

Periapical radiography resulted in a very dark and therefore non-conclusive image. The parents were advised of the need for surgical exploration of the lesion under general anesthesia due to the difficulty of handling a four-month-old child. Preoperative exams were requested, which were within normal limits. The surgical procedure was performed in a hospital two weeks after the first consultation, at which point, the lesion exhibited a proliferative growth, as it was more vegetative, lobulated and enlarged (Fig. 1b).

During the surgical procedure, intraosseous involvement of the lesion was observed, with the formation of a single bone cavity (Fig. 2) occupied by a mass of fibrous tissue with a good cleavage plane. The tooth germs of the deciduous right lateral incisor and canine were floating and enveloped in the tumor mass. Considering the bone destruction pattern and tooth involvement, a frozen biopsy was performed in order to rule out malignant lesion. The biopsy revealed an inflammatory lesion. Total curettage of the lesion was performed. The histological examination of the surgical specimens revealed diffuse proliferation of large mononuclear histiocytic cells (Langerhans cells) of a round or oval shape, pale vesicular nucleus and eosinophilic cytoplasm (Fig. 3). Abundant numbers of eosinophils and lymphocytes were observed. The histiocytic cells exhibited strong nuclear and cytoplasmic immunohistochemical positivity to S-100 (Fig. 4a) and CD1a showing a membrane reaction (Fig. 4b). Thus, the diagnosis of Langerhans cell histiocytosis was established.

The patient was referred for evaluation at the Department of Oncology (Support Group for Children and Adolescents with Cancer). Laboratory and image analyses were performed – conventional X-ray examinations [skull (Fig. 5a), spine (Fig. 5b) and hook bone (Fig. 5c)], computed tomography, ultrasound examination of the abdomen and nuclear medicine (Fig. 6). The results revealed no involvement of any other organ and that the lesion was located exclusively in the jaw. Thus, this was a case of the unifocal form of LCH. Clinical (Fig. 7) and radiographic follow up after six months revealed that the operated area was completely repaired (Fig. 8). Computed tomography of the

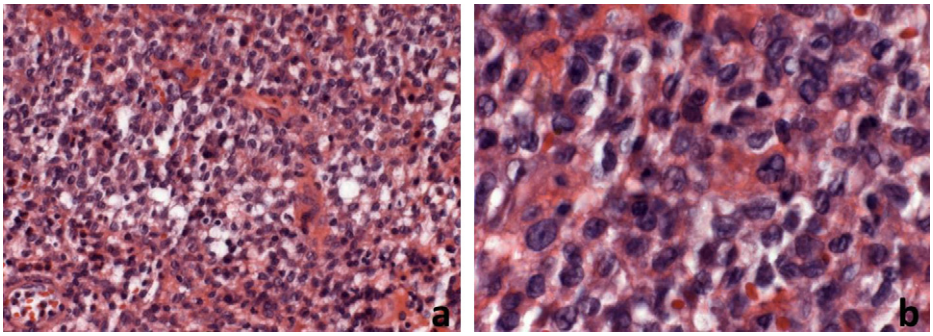

Fig. 3. (a) Dense infiltration of large mononuclear histiocytic cells associated to eosinophil, lymphocyte and macrophage infiltration (H&E, original magnification, 200×). (b) High power view revealing histiocytic cells with round shape, pale vesicular nucleus and eosinophilic cytoplasm (H&E staining, original magnification, 400×).