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Letterer siwe disease (LSD): A review

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Abstract
Langerhans cell histiocytosis is defined as a clonal proliferation of Langerhans phenotypic-like cells. Letterer-Siwe disease is the most common and serious of these entities, affecting mainly infants up to two years of age. We present two cases of this rare disease, diagnosed after dermatological examination, highlighting its typical aspects. The most frequent LCH occurs in children between 1 and 3 years of age, although it may occur at any age. Its annual incidence seems to be between 2 to 5 cases per million children. It is twice more frequent among males. Its pathogenesis remains unknown. Its reactional or neoplastic nature is still debated, although there are more arguments favoring the first option.

Keywords: LCH (Langerhans cell histiocytosis), clonal proliferation, Letterer-Siwe disease

Introduction
The histiocytic disorders cover a wide range of primary and secondary, solitary and multiple, benign and malignant disorders. Langerhans cell histiocytosis (LCH) is a reactive disorder in which cells having the phenotypic markers of epidermal Langerhans cells are found in skin and other organs where they cause damage by excessive production of cytokines and prostaglandins. The Letterer-Siwe syndrome is considered to be the acute disseminated form of the disease, characterized by cutaneous lesions, hepatomegalies, splenomegalies, and ganglionic hypertrophies, usually occurring in infants and newborns. Bone lesions occur in the skull, long bones, and mandible. Lesions in the mandible show a definite radiolucent image which may mimic both juvenile and severe periodontal disease. Eosinophilic granuloma is the most frequently reported and mildest form of the disease. This variety is considered to be a chronic localized form, characterized by single or multiple osseous lesions, usually affecting children and young adults. Any bone in the skeletal system, including the mandible, may be affected. The prognosis is excellent and the lesions may spontaneously recede within one or two years.

Epidemiology
LCH is an infrequent disease. The relative incidence of LCH is not well known, due principally to the heterogeneous clinical expression, but is estimated at approximately 2-5 cases per million inhabitants per year, being more frequent between the first and third decades of life, although it may affect any age group.

Etiopathogenesis
It may be caused by a dysfunction of the immune system, representing a hypersensitive reaction to an unknown antigen, with stimulation of the histiocytes-macrophage system. Deficiency of suppressor lymphocytes (T8), altered immunoglobins, autoantibodies, anomalous lymphocytic response to various mitogens and structural changes in the thymus in all the advanced forms have been found in LCH patients.

Oral manifestations of LCH
Bone lesions, alongside the cranium, the maxilla and mandible are the most affected bones, usually infilltrat-ing together. Mandibular lesions are clearly more frequent in all three forms of LCH.
Dagenais *et al*. in a review of 29 cases of LCH found the majority of bone lesions presented in the posterior section of the mandible (distal and canine region) and in the ramus of mandible. When osteolysis is found in the anterior area of the mandible it is as an extension of the posterior.

Solitary intra-bony lesions: localized outside the alveolar process, these are the most frequent in the initial phases. The images are circular or elliptical, solitary or univocal, found principally in the body and ramus of mandible.
Alveolar lesions with bone neo formation: formation of new bone in lesions classified as intra-bony is observed in a high number of cases. This is a relevant characteristic when differentiating LCH lesions from those of periodontal disease.

Diagnostic and complementary examination
Biopsy by conventional microscopy shows areas of conjunctive tissue related with a mixed inflammatory infiltrate. Nonmalignant histiocytic proliferation is seen together with the Langerhans cells (with Birbeck granules).
The biopsy is similar in all LCH except in the acute disseminated form, as this may demonstrate microscopic findings of other diseases, such as acute forms of lymphoma. There are no specific laboratory tests for LCH, however, blood and urine tests exist that reveal the extent and seriousness of the disease. Routine laboratory analyses, liver function tests and coagulation tests are made. Imaging studies include X-ray of thorax, computed tomography (CT) and magnetic resonance imaging (MRN) of the affected areas, with the aim of delimiting the bone and soft tissue lesions. CT is useful to evaluate the cranium and facial bones, which are difficult to visualize in conventional radiographies.

Prognosis and treatment
The most important factors that may worsen the prognosis are firstly, visceral involvement (liver, lung, bone marrow), as this has a negative effect on survival. Secondly, where age at first presentation is less than two years since mortality rises to 50%. Thirdly, when the disease spreads to various bones or soft tissues. In general, it is considered that the younger the patient, the worse the prognosis.
According to Zhang *et al*, these patients should be treated surgically and only complemented with a low-dose radiotherapy and/or chemotherapy in serious cases, especially in disseminated forms. Localized and isolated mandibular lesions may be efficiently treated by surgical curettage. When surgery leaves large bony defects, autologous bone grafts can be made in an attempt to reduce the risk of pathological fracture and to facilitate bone regeneration.
Some authors Suggest treatment with low-dose radiotherapy in large or multifocal lesions that recur or progress after surgery, in lesions carrying risk of fracture, lesions inaccessible to surgery, painful or disseminated lesions, or in those occurring in the centers of the mandible during infancy.

Conclusion
The clinicians' view should not be limited to the periodontal tissue conditions, but include the patient as a whole. The presence of previously mentioned systemic symptoms associated with periodontal lesions may be a guide for correct LCH diagnosis. The suspicion of the LCH should be considered in case of recurrence of periodontal lesions and rapid severe and localized loss of periodontal bone.

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