Abstract: Langerhans cell histiocytosis is a relatively rare unique disease process characterized by an abnormal proliferation of immature dendritic cells usually affecting children and young adults. The purpose of this report is to describe a unique and very rare case of Langerhans cell histiocytosis in the left Zygomatic bone.

Keyword: Langerhans Cell Histiocytosis, Zygomatic Bone, Histiocytosis X, ENT

INTRODUCTION:
Langerhans cell histiocytosis (LCH) is a rare unique disorder of the reticuloendothelial system characterized by an abnormal proliferation of histiocytes and eosinophilic leukocytes. Langerhans cell histiocytosis formerly referred to as histiocytosis X, can be divided into three categories: eosinophilic granuloma, Hand-Schüller-Christian disease and Abt-Letterer-Siwe disease. Clinically LCH is rarely possible.

The disease is most common in infants and younger children, but cases where histiocytosis occurs in adults, even the elderly, are not infrequent. Sometimes this is due to diagnostic difficulties, so that the appropriate diagnosis is made in an adult patient. Most scientific reports concern patients aged below 21. The etiology of the disease is unclear. In pathogenesis a key role is played by Langerhans cells, which are structurally and functionally different from those cells in healthy subjects. An important phenomenon in the pathogenesis of the disease is uncontrolled, monoclonal proliferation of Langerhans Cells, which leads to eosinophil infiltration in tissues and organs. Cytokines and prostaglandins secreted by these abnormal histiocytes can damage organs involved in the disease process, or even to systemic consequences such as anaphylactic shock. Abnormal immunoregulation due to abnormal release of cytokines as well as unidentified infectious agents are considered among the causes of the disease. Due to the fact that the inflammatory infiltrate of histiocytes leads to destruction of the structure of the affected tissue and the disease can progress from a limited to generalized form, some researchers categorize Langerhans cell histiocytosis as a borderline disease between inflammatory changes and cancer. The method of treatment of histiocytosis confirms this thesis, as in addition to the commonly used steroids, methods reserved for cancer patients such as surgical excision, radiotherapy or chemotherapy are successfully applied. Symptoms of the disease vary greatly and depend on which organs were infiltrated. The diagnosis of histiocytosis is determined by the characteristic image of histopathological material obtained by biopsy. The earlier the stages of the disease are, the easier it is to identify Langerhans cells, due to fibrosis granulomas which occur in the course of the disease. Then Immunohistochemical markers such as S-100 protein, ATPase, alpha-mannosidase, lecithin, and vimentin can be useful, although they are not very specific and serve only as a complement to histopathology. Definitive diagnosis in doubtful cases may be obtained by observing the presence of Langerhans granules (X-body, Birbeck granules) in the electron microscope or the presence of CD1 proteins on the cell surface. In case of local lesions most often is limited to surgery and/or irradiation. Prognosis depends on the extent of the lesion. In the case of limited form the pathological process involves a single organ, most often bones (40-78%). The effectiveness of treatment reaches 97% and spontaneous remissions occur in patients who are not treated. Other common locations of localized form are lymph nodes and skin. For the generalized form the prognosis is worse. It mainly occurs in infants or children under 3 years old and shows multiple organ involvement, leading to their insufficiency. Lesions are mainly localized in the lungs, liver, spleen and central nervous system (usually manifesting as pituitary insufficiency).

CASE REPORT:
A twelve year old boy reported to the OPD with chief complaints of pain and swelling left upper cheek just below the left lower eyelid over a period of approximately forty five days. The child appeared generally healthy and moderately built. Past medical and family history were unremarkable. Intra oral examination was normal. A swelling, of variable consistency was noticed on the left Zygomatic region.
LCH is still a very rare disease in the head & neck region, the etiology, and pathogenesis of which remain unclear. Varieties of etiological factors have been proposed including immunologic reactions, viruses, bacteria, and genetic influences. Possible development of LCH under the influence of colony stimulating factor (GM-CSF), interleukin-3, and tumor necrosis factor-alpha have also been suggested and recently, cytogenetic studies have proposed the role of tumor suppressor genes (p53), oncopogenes (c-my, h-raps), growth factors, cell surface immunologic markers and apoptotic factors in LCH as well. Unifocal disease involves a single site of involvement and has a good prognosis.

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