Case Report

Langerhans cell histiocytosis – a rare tumor at rare site: A case report in adult

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Langerhans cell histiocytosis (LCH) is a rare monoclonal proliferative disease originating from Langerhans cell. It is seen more commonly in children than in adults. Here, we present a case of LCH of hard palate in a middle aged female. This patient presented with a history of ulceroproliferative growth over the right side of hard palate for the past 6–7 months. Clinically, a provisional diagnosis of suspected malignant neoplasm was made. Trucut biopsy from the site was done and the lesion was histopathologically diagnosed as LCH. Furthermore, IHC was also done to confirm the diagnosis.

KEY WORDS: Hard palate, Langerhans cell histiocytosis, non-healing ulcer

INTRODUCTION

Langerhans cell histiocytosis (LCH) is an abnormal clonal proliferation of cells that morphologically and immunophenotypically resemble Langerhans cells. These cells originate from bone marrow myeloid precursors.[1] It expresses HLA-DR and CD 1a. The cytoplasm of cells has HX bodies also known as Birbeck granules. The disease has several older names that include eosinophilic granuloma, Hand-Schüller-Christian disease, Letterer-Siwe disease, and histiocytosis X. The name LCH was introduced later. According to recent classification, LCH is divided into single-system and multisystem.[2,3] LCH can appear at any age but peak incidence is seen between 1 and 4 years of age with male predilection.[2,4] The age of onset varies depending on the variety of LCH and is rarely seen in adults with an annual incidence of 1–2 cases/million persons.[5] A condition known as LCH causes an overabundance of the immune system's Langerhans cells to accumulate in the body. The body typically contains Langerhans cells, which aid in the control of the immune system. These cells are particularly prevalent in the skin, lymph nodes, spleen, lungs, liver, and bone marrow. Excess immature Langerhans cells in LCH typically develop into granulomas, which are benign tumors. Nowadays, a lot of scientists think that LCH is a type of cancer.

The aim of this case report is to raise awareness about this disease among clinicians in differential diagnosis of other common cancer in this site. Furthermore, to reduce long term/permanent consequences of this disease and to improve quality of life among these patients.

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CASE REPORT

A 55-year-old female visited the outpatient department of surgery RMCH with chief complaints of swelling and pain in the region of hard palate. The patient first noticed the swelling 6–7 months back. There was no history of any regression in size of swelling.

On examination, an ulceroproliferative growth was present over right side of hard palate, 2 × 1 cm in size with ill-defined margins. Based on the history, provisional clinical diagnosis of suspected malignant neoplasm was made and patient was subjected to radiographic and hematological investigations. Complete hemogram showed values that were within the normal range. Excisional biopsy was done in the same department and sample was sent to pathology department for histopathological examination.

Pathological Findings

Gross examination showed multiple soft-tissue pieces. Grey-brown to grey-white in color, measuring 2 × 1 × 0.5 cm. Representative sections from the tissue showed oral mucosa lined by stratified squamous epithelium, sub epithelial area predominantly showed sheets of large, polyhedral cells having linear nuclear groove (coffee bean appearance), foamy eosinophilic cytoplasm, and inconspicuous nucleoli. Intervening stroma also showed moderate infiltration of inflammatory cells comprising of eosinophils, neutrophils, and lymphocytes [Figure 1a-d]. Hence, a diagnosis of LCH was suspected. Immunohistochemical staining for S-100 and CD68 was positive and weak-to-moderate diffuse positive for CD45. Ki-67 show 40–45% activity [Figure 2].

DISCUSSION

LCH is part of a group of syndromes called histiocytosis X, which are characterized by an abnormal proliferation of histiocytes. It is first described by Lichenstein in 1953.[6]

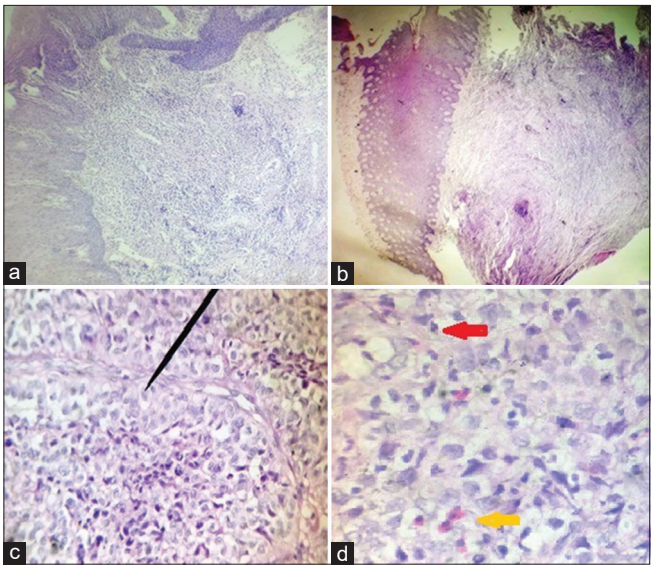

Figure 1: (a and b) H&E stained section shows oral mucosa lined by stratified squamous epithelium; sub epithelial area predominantly shows sheets of large, polyhedral cells. (×100 magnification; low power), (c) H&E stained section shows sheets of tumor cells having vesicular nuclei with linear nuclear groove (coffee bean appearance), foamy eosinophilic cytoplasm, and inconspicuous nucleoli. (×400 magnification; high power), (d) H&E stained section shows infiltration of inflammatory cells comprising of eosinophils (yellow color ←) and neutrophils (red color ←) in intervening stroma. (×400 magnification; high power)

Incidence of this disease is reported as 8.9 cases per million among children and 1–2 cases per million in adult populations.[2,7]

LCH most commonly involves skin and bone, followed by hematopoietic system, lymphnodes, liver, spleen, soft tissue, lung, thymus, and pituitary gland. In oral cavity soft tissues, gingiva is commonly involved followed by maxillary and hard palate.[8]

The pathogenesis of LCH is a matter of debate to determine whether LCH is a reactive (non-cancerous) or neoplastic (cancerous) process. Reactive nature of LCH is supported by occurrence of spontaneous remissions and the extensive secretion of multiple cytokines by dendritic cells (cytokine storm). Whereas somatic mutations in the BRAF gene and clonal proliferation help to support the neoplastic nature of LCH. The somatic gene mutations are acquired during a person's lifetime and are present only in certain cells. These changes are not inherited.

Making a protein that is often turned on and off in response to signals that regulate cell growth and development is made possible by the BRAF gene. The BRAF protein is continuously active and sends messages to the nucleus in afflicted cells as a result of somatic mutations, even when these chemical signals are not present. By permitting the Langerhans cells to proliferate and divide uncontrolled, the hyperactive protein may aid in the emergence of LCH.

LCH has long-term permanent consequences such as diabetes insipidus, orthopedic disabilities, neuropsychological defects, stunted growth, skin scarring, hearing defects, chronic pulmonary dysfunctions, liver cirrhosis, and secondary malignancies. Early diagnosis and effective treatment prevent the progression of disease.

The observation in our study was correlated and compared with studies of different authors. In our knowledge, only few cases

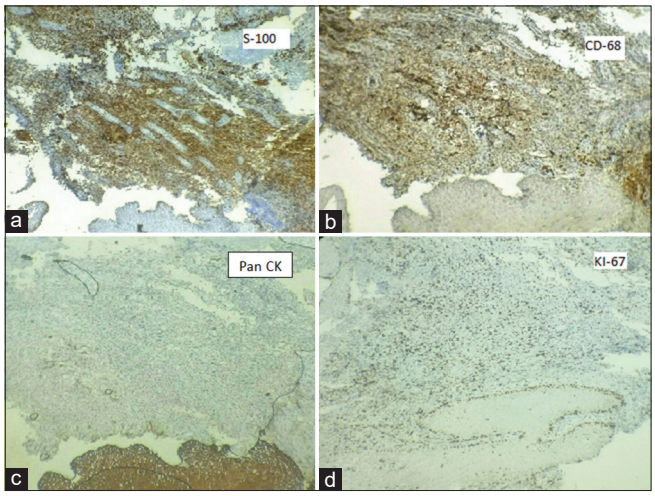

Figure 2: Immunohistochemical staining shows (a) Langerhans cell highlighted with S-100. (×100 magnification); (b) Langerhans cells highlighted with CD-68. (×100 magnification); (c) Negative Pan-CK staining in Langerhans cells. (×100 magnification); (d) Ki-67 showing 40–45% activity. (×100 magnification)

Table 1: Comparison of studies by different authors

Author	Year	Age of patient	Sex	Symptoms
Tenório *et al*.[10]	2020	37 years	Female	Ulcerative lesion on alveolar ridge that extended to the hard palate
Varsha *et al*.[9]	2016	45 years	Male	an ulcer on the hard palate
Our study	2021	55 years	Female	Ulcerative lesion over hard palate

of LCH of hard palate have been reported. In Varsha *et al*.'s study, a male was affected by LCH whereas in Tenório *et al*.'s study, a female was affected.[9,10] Similarly in our case report, the patient was a middle-aged female. In two studies, patient presented with ulcer over hard palate and diagnosis was made on histopathological examination and was confirmed by immunohistochemistry. Similarly in our study, histopathological examination and IHC were also performed [Table 1].

CONCLUSION

LCH of hard palate must be considered in the differential diagnosis of other malignancies in oral cavity especially of hard palate because if the disease is diagnosed early, it helps the clinicians to treat the patients early and reduce its complications and mortality. This improves the quality of life of patients. Furthermore, the case report highlights the valuable role of histopathology and immunohistochemistry in diagnosing this lesion.

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