A Rare Case of Langerhans Cell Histiocytosis in the Hard Palate

Sert Damakta Nadir Bir Langerhans Hücre Histiyositoz Olgusu

Memiş Hilmi Atay[1], Seda Gün[2]

[1]Ondokuz Mayıs University Faculty of Medicine, Department of Internal Medicine, Division of Hematology, Samsun, Turkey
[2]Ondokuz Mayıs University Faculty of Medicine, Department of Pathology, Samsun, Turkey

To The Editor,

Langerhans cell histiocytosis (LCH) is an inflammatory neoplasm of myeloid origin characterized by infiltration of dendritic CD1a+/CD207+ cells [1]. LCH mostly affects pediatric patients with a median age of 3.8 years, but adult cases of LCH are also seen. Systemic involvement of LCH can be seen commonly for the bone, skin, hypothalamus, and pituitary stalk; however, localized involvement is also seen to a lesser extent [2]. Orbital lesions were reported to be more frequent in pediatric patients, but mandible and mucocutaneous lesions were more frequent in adult patients [3]. Here we report a rare case of adult LCH that presented with hard palate pain when eating.

A 32-year-old female patient was admitted to the dermatology clinic due to oral scars. Physical examination revealed an ulcerative lesion of 1x1 cm in size on the soft palate. The biopsy report showed an ulcerated lesion with granulation tissue. Treatment with 40 mg of oral prednisolone was initiated. After two weeks, prednisolone was tapered and discontinued because the patient's complaints had subsided. Six months later, a plaque-shaped lesion involving most of the soft palate was detected (Figure 1a). Complete blood count and biochemistry values were within reference ranges. No pathology was detected by positron emission tomography-computed tomography besides sclerotic lesions and heterogeneous 18F-fluorodeoxyglucose involvement in the bilateral distal femur (SUV_{max}: 3.47). Pathological cells in the re-biopsy sample were positive for S-100, CD1a, and cyclin D1 staining by immunohistochemistry. Although langerin (CD207) testing could not be performed, S-100 and CD1a positivity confirmed the diagnosis of LCH (Figures 1b and 1c). BRAF mutations were not detected by polymerase chain reaction. Administration of vinblastine at 6 mg/m^2 weekly and prednisolone at 40 mg/m^2 daily was begun. The chemotherapy protocol was discontinued due to the occurrence of severe

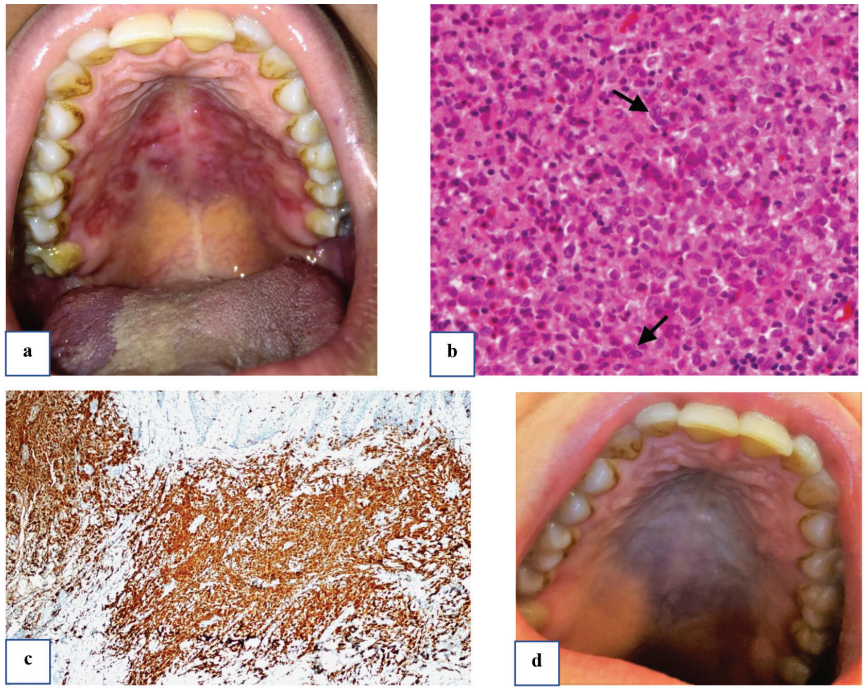

Figure 1. a) Erosive lesion on the hard palate before treatment. **b)** Infiltration containing eosinophilic leukocytes and histiocytes with coffee-bean-shaped appearance and kidney-shaped nuclei. Hematoxylin and eosin, 400^x. **c)** Langerhans cells exhibiting positivity for S-100. DAB, 100^x. **d)** Healed lesions on the hard palate after treatment.

neuropathy of the hands and feet in the second week of therapy. Cytarabine (100 mg/m^2 per dose at 5 days per month for 12 months) was started. The oral lesions had significantly improved after 3 months of treatment (Figure 1d).

Oral manifestations of LCH in cases of both isolated and multisystemic disease are rare. Clinically, intraoral LCH lesions may manifest as masses, gingivitis, ulcers, gingival bleeding, tooth loss, pain, cysts, leukoplakia, or non-healing wounds after surgery or tooth extraction. The differential diagnosis depends on the site of involvement. Oral involvement may histologically be confused with granulomas, allergic-like inflammation, stomatitis, cysts, foreign-body giant cell reaction, lymphoma, and metastasis [4]. Cyclin D1 is an efficient marker to differentiate neoplastic from reactive LC proliferation [5]. Treatment options for LCH vary according to the site of involvement, extent of the disease, and presence of loss of function of the involved organs. Various multimodality treatment approaches include systemic chemotherapy, steroids, purine analog antimetabolites, targeted therapy, radiotherapy, and surgery [6,7,8,9,10].

This case was presented here due to the rare clinical presentation. LCH should be considered in the differential diagnosis of intraoral lesions. Early diagnosis of LCH is important because curative treatment is available and symptoms can be relieved if it is treated early.

Keywords: Cytarabine, Oral lesion, Langerhans cell histiocytosis

Anahtar Sözcükler: Sitarabin, Oral lezyon, Langerhans hücreli histiyositoz

Ethics

Informed Consent: Received.

Authorship Contributions

Surgical and Medical Practices: M.H.A., S.G.; Concept: M.H.A.; Design: M.H.A.; Data Collection or Processing: M.H.A., S.G.; Analysis or Interpretation: M.H.A.; Literature Search: M.H.A., S.G.; Writing: M.H.A.

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Address for Correspondence/Yazışma Adresi: Memiş Hilmi Atay, M.D., Ondokuz Mayıs University Faculty of Medicine, Department of Internal Medicine, Division of Hematology, Samsun, Turkey
E-mail : draatay@gmail.com ORCID: orcid.org/0000-0001-9666-6955

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