Lymphangioma Circumscriptum of the Buccal Mucosa: Case Report

Yanak Mukozasına Yerleşmiş Lenfanjiyoma Sirkumskriptum

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ABSTRACT Lymphangiomas are congenital and hamartomatous structures which affect the lymph vessels of skin and subcutaneous tissue. Lymphangiomas are divided into two major groups, superficial and deep, based on the depth and size of the abnormal lymph vessels. The superficial group includes lymphangioma circumscriptum (LC), whereas the deeper group includes cavernous lymphangiomas and cystic hygromas. The most frequent locations of LC are head, neck, proximal extremities, buttocks and trunk. Mucosal involvement is rare but do occur. A 29-year-old man presented with raised lesion his buccal mucosa since childhood. Clinical examination revealed translucent mucosal vesicles on the left buccal mucosa. Incisional biopsy revealed features of lymphangioma circumscriptum after histopathological examination. To our knowledge current case is the first report of LC which is localized to the buccal mucosa.

Key Words: Lymphangioma; mouth mucosa

ÖZET Lenfanjiyomalar deri ve subkutan dokudaki lenf damarlarını etkileyen konjenital ve hamartamatöz yapılardır. Anormal lenf damarlarının derinliği ve yapısına bağlı olarak lenfanjiyomalar iki majör gruba ayrılır. Yüzeyel grupta lenfanjiyoma sirkumskriptum yer alırken derin grupta, kavernöz lenfanjiyom ve kistik higroma yer alır. En sık etkilenen alanlar, baş, boyun, proksimal ekstremiteler, kalçalar ve gövdedir. Mukozal tutulum nadir olmakla birlikte görülebilir. Yirmidokuz yaşında erkek hasta çocukluğundan beri sol yanak mukozasında olan kabarıklık şikayeti ile polikliniğimize başvurdu. Fizik muayenesinde, sol bukkal mukozada translüsen mukozal veziküller saptandı. Yapılan insizyonel biyopsinin histopatolojik incelemesi sonucu lenfanjiyoma sirkumskriptum ile uyumlu olarak belirlendi. Bildiğimiz kadarıyla bu olgu, bildirilmiş bukkal mukoza yerleşimli ilk lenfanjiyoma sirkumskriptum olgusudur.

Anahtar Kelimeler: Lenfanjiyoma; ağız mukozası

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Lymphangiomas are hamartomatous, congenital malformations of the lymphatic system that may involve the skin and subcutaneous tissues. They can occur on any cutaneous surface or mucous membrane. The most common skin sites are the head and neck, proximal extremities, buttocks, and trunk, while the intestines, pancreas, and mesentery are other potentially affected organs.[1] Currently, however, lymphangiomas are divided into two major groups, superficial and deep, based on the depth and size of the abnormal lymph vessels. The superficial group includes "lymphangioma circumscriptum" (LC) that was named in 1989 by Morris, whereas the

deeper group includes cavernous lymphangiomas and cystic hygromas.[1,2] In a study by Flanagan et al. it was found that there was no correlation among cutaneous LC, mucous membrane lymphangioma, or internal organ lymphangioma.[3]

CASE REPORT

A 29-year-old man presented with raised lesion on his buccal mucosa since childhood. He did not have any other cutaneous and systemic symptoms. Clinical examination revealed three, each of one cm translucent grouped mucosal vesicles on the left buccal mucosa (Figure 1). There was no pathologic finding among the physical examination. Buccal mucosa biopsy revealed features of lymphangioma, such as collection of subcutaneous lymphatic cisterns communicating through dilated channels with superficial vesicles (Figure 2).

DISCUSSION

LC is a congenital malformation of superficial lymphatics present at birth or soon after. It presents as a group of vesicles containing clear fluid (resembling frog's spawn), the size of which can be up to 5 mm in diameter. Sometimes the vesicles can be hemorrhagic. Hemorrhage can occur spontaneously or secondary to insignificant trauma.[4] The etiology is unclear; however, lymphatic obstruction has been suggested as a possible cause.[5] In

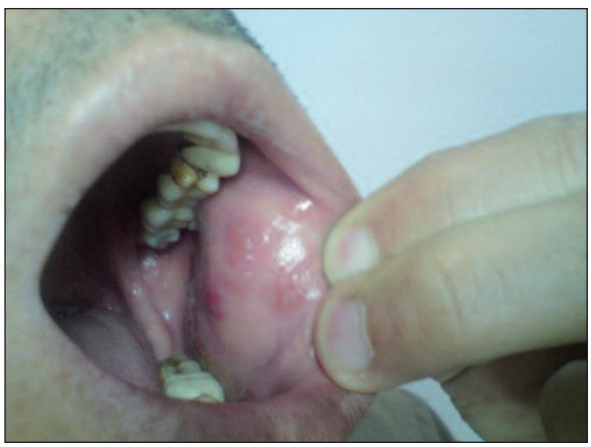

FIGURE 1: Translucent mucosal vesicles on left buccal mucosa.
(See color figure at http://www.turkiyeklinikleri.com/journal/turkiye-klinikleri-journal-of-case-reports/1300-0284/tr-index.html)

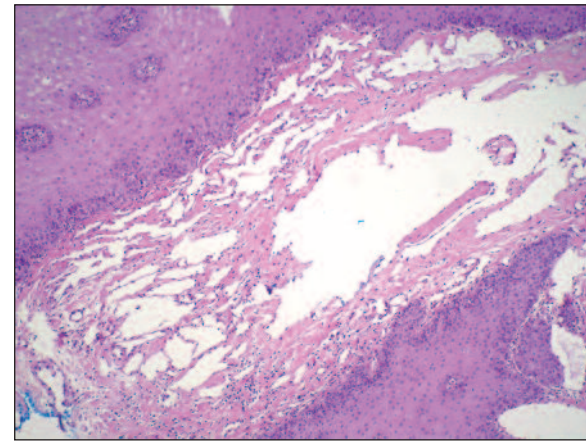

FIGURE 2: Collection of subcutaneous lymphatic cisterns communicating through dilated channels with superficial vesicles (hematoxylin and eosin stain X 20)
(See color figure at http://www.turkiyeklinikleri.com/journal/turkiye-klinikleri-journal-of-case-reports/1300-0284/tr-index.html)

1970, Peachey et al. divided LC into two main groups: classic and localized. The classic form of LC is typically seen at or soon after birth, is often larger than 1 cm², and usually covers the proximal limbs. It is unusual for the classic form to progress from its original vesicular appearance to warty plaques. Conversely, the localized form is seen at any age, is often less than 1 cm², and may appear anywhere on the body. In neither form has communication between the abnormal lymphatics and the normal lymph system been shown.[5-7] Lymphangiomas usually occurs as lobulated masses or cystic lesions arising in the head and neck, axilla, and abdomen.[6] Mucosal involvement is rare but do occur. LC located in vulva have been reported previously.[8] Oral lymphangiomas may occur at various sites but are more frequent in the anterior half of the tongue, where they often result in macroglossia. The superficial lesions are manifested as papillary lesions, which may be the same colour as the surrounding mucosa or of a slightly reddish color.[9]

To our knowledge, LC localized to the buccal mucosa has been never reported in published work, current case is the first report. The lesion was considered for the localized form of LC because of beginning childhood and occupying a small area. Treatment of LC is generally undertaken on the basis of cosmetic considerations, persistent lymphatic leakage, and recurrent infections particu-

larly with *Staphylococcus aureus*, may significantly impair quality of life. Other risks associated with LC include lymphangiosarcoma, which has been reported at the site of previously irradiated lesions, and squamous cell carcinoma, which can develop in longstanding cases.[7] Discoloration of the vesicles can lead to confusion with hemangiomas even with malignant melanoma.[5,7] In additionally to surgical excision recent therapies for LC have been focused on sclerotherapy, radiofrequency ablation, cryotherapy, laser therapy and topical imiquimod.[10-12] Our patient does have not have any symptom while eating or chewing, so he refused excision. Regular follow-up was advised to him for complications.

In conclusion we suggest that LC should be considered in the differential diagnosis of vesicular lesions on the buccal mucosa and a biopsy is essential for making an accurate diagnosis.

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