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CAVERNOUS LYMPHANGIOMA OF THE TONGUE IN AN ADULT: A CASE REPORT

Yetişkin Bir Bireyde Dilde Kavernöz Lenfanjiom: Bir Olgu Sunumu

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ABSTRACT

Lymphangioma is a benign hamartomatous lesion caused by congenital malformation of the lymphatic system. This benign tumor is detected most commonly at birth or in early childhood but rarely in adults. On clinical examination, most lymphangiomas contain clear lymph fluid, but some may present as transparent vesicles containing red blood cells due to hemorrhage. In addition, lymphangioma may occur in association with hemangioma. This tumor occurs most commonly in the head and neck area, but rarely in the oral cavity. The dorsum of the tongue is the most common location in the mouth, followed by the lips, buccal mucosa, soft palate, and floor of the mouth. There are various treatment approaches for lymphangioma, but surgical excision is the preferred method. We present a case of a 26-year-old man with lymphangioma on the anterior dorsal part of the tongue, not associated with any dysfunction in mastication or speech disorders.

Keywords: *Lymphangioma; oral cavity; oral; tongue; surgery*

ÖZ

Hamartamatöz lezyon olarak da bilinen lenfanjiomlar, lenfatik sistemin konjenital malformasyonu sonucu ortaya çıkan iyi huylu tümörlerdir. Genellikle doğumda ve çocukluğun erken dönemlerinde fark edilmekle beraber, yetişkinlerde nadiren ortaya çıkarlar. Çoğunlukla içeriğinde temiz lenf sıvısı barındıran bu lezyonlar, bazen kanamadan dolayı kırmızı kan hücreleri ihtiva eden transparan vesiküller de içerebilir. En sık baş-boyun bölgesinde lokalize olurlar, nadiren de oral kavitede gözlemlenirler. Ağız içerisinde dil dorsumunu takiben en sık görüldüğü alanlar sırasıyla dudaklar, bukkal mukoza, yumuşak damak ve ağız tabanıdır. Lenfanjiomların tedavisine yönelik değişik tedavi yaklaşımları benimsenmiştir. Cerrahi eksizyon bunlardan en sık tercih edilen yöntemdir. Bu makalede 26 yaşında erkek hastada, dilin dorsal yüzeyinde bulunan ve çiğneme sisteminde herhangi bir disfonksiyon ile konuşma bozukluğu yaratmayan lenfanjiom olgusu sunulmuştur.

Anahtar kelimeler: *Lenfanjiyom; ağız boşluğu; ağız; dil; cerrahi*

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Introduction

Lymphangioma is a benign hamartomatous tumour that occurs because of congenital malformation of the lymphatic system and is characterized by benign proliferation of lymphatic vessels. It is also known as lymphatic hamartoma (1). Lymphangioma is usually detected at birth or in early childhood, and 90% of cases develop before 2 years of age (2-4). This tumor may arise anywhere in the body, but the shoulder, armpit, abdomen, neck, pharynx, eyelids, and conjunctiva are most frequently affected. Although the head and neck are common locations for lymphangioma, the oral cavity is rarely involved. Intraoral lymphangioma occurs mostly on the dorsal surface and lateral border of the tongue, and rarely on the palate, gingiva, buccal mucosa, and lips (5). In children, intraoral lymphangioma, especially tongue lymphangioma leading to macroglossia, may cause speech disturbances, poor oral hygiene, mandibular prognathism, openbite, yawning, chewing difficulties, and maxillofacial deformities (1). Therefore, appropriate and timely treatment is essential to avoid undesirable consequences, which in most cases are caused by macroglossia due to tongue tumors (4). Histopathologic classification of lymphangioma is as follows: (A) lymphangioma simplex, small thin-walled lymphatic vessels; (B) cavernous lymphangioma, dilated lymphatic vessels with surrounding adventitia; (C) cystic lymphangioma, large lymphatic spaces surrounded by fibrovascular tissue; and (D) benign lymphangioendothelioma, lymphatic channels separated by collagen bundles (2).

Various modalities have been applied for treatment of lymphangioma of the tongue. Treatment is aimed at symptomatic relief of pain, edema, lymph and blood leakage, and superinfections, as well as addressing cosmetic concerns. The preferred treatment is surgical excision; however, this cannot be applied in all cases and leaves a scar (6). Other treatment modalities include cryotherapy, radiation therapy, steroid administration, sclerotherapy, electrocautery, embolization, ligation, laser surgery, and radiofrequency tissue ablation. Successful treatment necessitates inclusion of a surrounding border of normal healthy tissue without damage of vital structures.

Case Report

A 26-year-old male was referred to the Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, Istanbul University. He was complaining about an abnormal lesion on his tongue which is present since childhood. The patient's history was taken, and physical examination was performed together with the submission of samples for blood tests. There was no obvious medical history to explain the lesion, such as respiratory infection. Intraoral examination revealed a swelling on the dorsal part of the tongue, approximately 1.5 × 1.5 cm in size, with numerous papillary and vesicle-like projections resulting in a granular and transparent appearance (Figure 1). The lesion was soft and slightly tender on palpation, and probing caused no bleeding and showed that the overlying mucosa was firmly adhered to the lesion. There was no restriction in the functions of the tongue, but the patient reported that the tongue changed color from time to time. The results of the blood tests were normal. The clinical differential diagnoses included papillary hemangioma, pyogenic granuloma, hemangioma, and papillary hyperplasia.

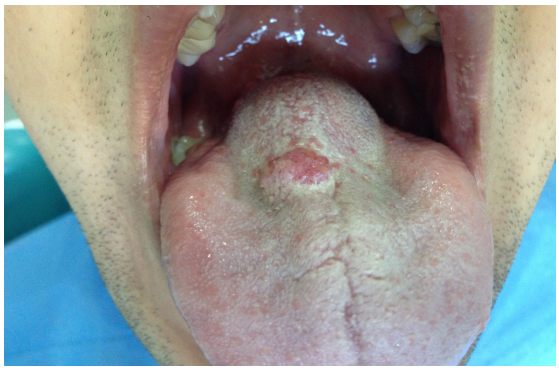

Figure 1. *Clinical photograph of the lesion on the dorsal surface of the tongue.*

The patient was advised to undergo surgical excision of the lesion, and excisional biopsy of the lesion including 2 mm margins of healthy tissue was performed using local anesthesia (Figure 2, Figure 3). One week after the postoperative examination revealed that wound healing was normal (Figure 4). The patient was followed up regularly, and no signs of recurrence were present 1 year after the surgery (Figure 5). Histopathological examination revealed that the lesion was covered by stratified squamous epithelium with parakeratosis, spongiosis, and papillomatosis (Figure 6). Many enlarged lumens of various diameters and lined by thin-walled endothelial cells were observed in the lamina propria, just below the epithelium. The lumens contained proteinaceous material and small numbers of lymphocytes or

erythrocytes or both. Just below the lesion, there was infiltration of plasmocytes and focal lymphocytes in the superficial part of the connective tissue. Normal connective tissue and skeletal muscle tissue were observed in the deeper tissue layers. PAS staining revealed colonies of microorganisms containing radial extensions on the surface of the lesion. Based on these histopathologic features, the diagnosis was lymphangioma.

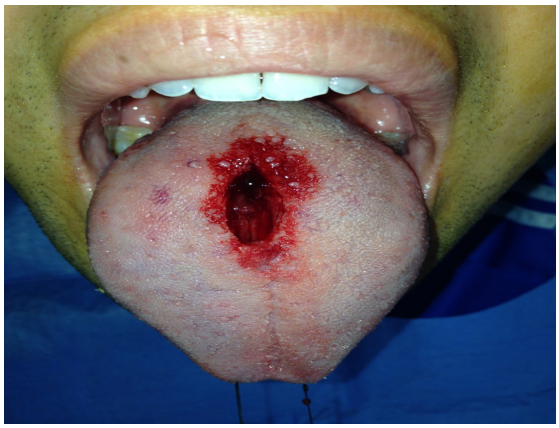

Figure 2. *The tongue after excision of the lesion.*

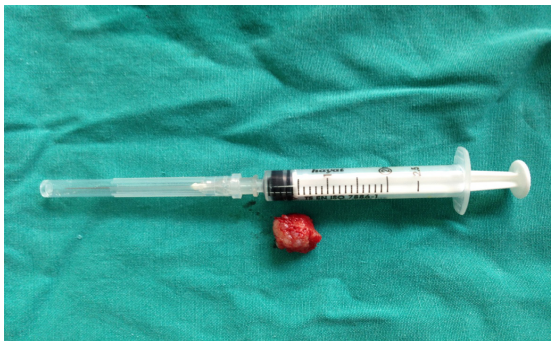

Figure 3*. The appearance of the excised lesion.*

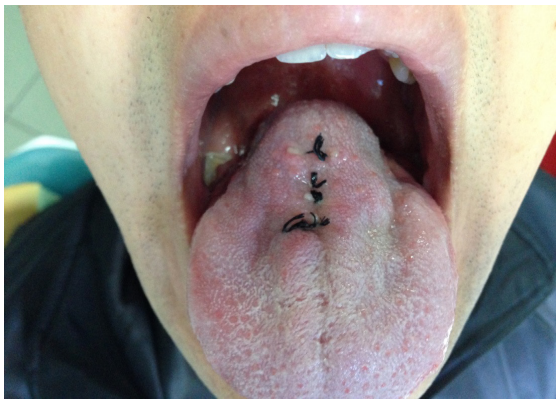

Figure 4. *One week after the operation and closure of the wound.*

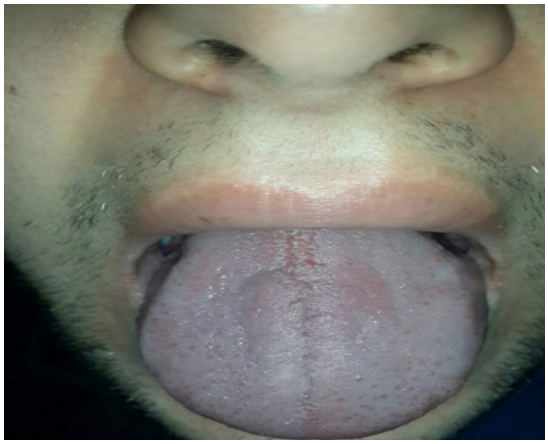

Figure 5. *The appearance of the tongue 1 year after the operation.*

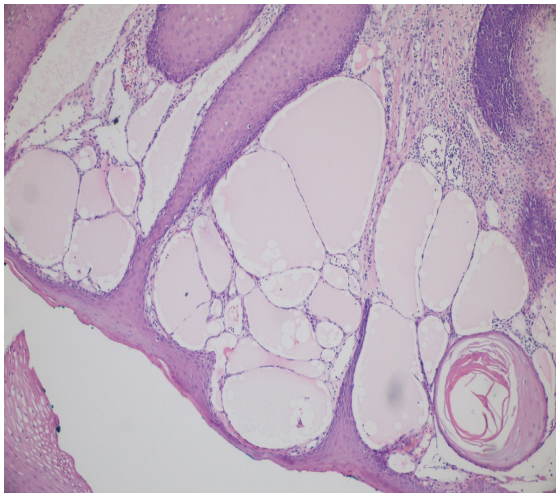

Figure 6. *Histological appearance of the enlarged lymphatic vessels under the surface epithelium (H&E, ×100).*

Discussion

Lymphangioma is a benign congenital malformation that originates from lymph vessels with unknown etiology. Redenbacher first described lymphangioma in 1828, and lymphangioma of the tongue was reported later in 1854 by Virchow (2, 7). Lymphangioma may be congenital or acquired, but is usually detected at birth or in early childhood. Sixty per cent of lymphangiomas diagnosed at birth and 80–90% in the second year of life (8). However, lymphangioma occurs rarely in adults (9). There are 2 theories about the origin of congenital lymphangioma. The first proposes that the lymphatic system develops from 5 primitive sacs arising from the venous system and endothelial outpouching from the jugular sac

expands to form the lymphatic system in the head and neck area. The second theory proposes that the lymphatic system is formed by mesenchymal clefts in the venous plexus reticulum that spread toward the center of the jugular sac (10). Differently than the infant lymphangioma; the etiology of lymphangioma acquired in adulthood include; trauma, inflammation, and lymphatic obstruction as possible pathogenic mechanism. Lymphangioma may present anywhere in the skin or mucosa, but is commonly located in the head and neck region (2). In the oral cavity, this lesion usually appears in the first decade of life and mostly occurs on the dorsal part and lateral border of the tongue. It rarely arises on the palate, gingival or buccal mucosa, and lips (7).The most common site for intraoral lymphangioma leading to macroglossia is the anterior two-thirds of the dorsal surface of the tongue. A classification of lymphangioma of the head and neck based on the anatomical involvement has been proposed by De Serres *et al*.(11) as follows: stage/class I, infrahyoid unilateral lesions; stage/class II, suprahyoid bilateral lesions; stage/class III, suprahyoid or infrahyoid unilateral lesions; stage/class IV, suprahyoid bilateral lesions; stage/class V, suprahyoid or infrahyoid bilateral lesions; and stage/class IV, infrahyoid bilateral lesions. Brennan *et al*.(12) reviewed a series of 49 cases of lymphangioma that were localized in the oral region, and found that 17 involved the tongue while only 1was located in the retromolar region and 1on the soft palate. The diagnosis of lymphangioma is not difficult, being mainly clinical. The clinical appearance varies with the size of the lesion. Superficial lesions have pink or yellowish nodules whereas deeper lesions are described as soft and smooth with natural color (13).

When lymphangioma occurs in the oral cavity, it manifests as distinct vesicles, which, when superficial in nature, result in a granular and translucent appearance. Because of damage of blood capillaries within the lymphatic structure, these lesions may have a blue or red appearance (1, 14). Lymphangiomas may be divided into 3 types on the basis of histological characteristics: capillary, cavernous, and cystic (1, 3). The patients with lymphangioma patients may be adversely affected in four ways: esthetic, occlusal, functional, and psychological (15). Lymphangiomas that are localized in the head and neck area can cause airway obstruction, speech disturbances, and swallowing and mastication difficulties (1, 10). In addition, rapid growth can be associated with hemorrhage or obstruction of the upper respiratory tract (16) and 50% of children with these lesions may require tracheotomy (17). The patients with lymphangioma in the oral cavity tend to have abnormalities such as speech disturbances, poor oral hygiene, and bleeding from the tongue associated with oral trauma. In this case, the lesion was present on the dorsal part of the tongue and did not affect speech or breathing (1). Differential diagnoses for lymphangioma include hemangioma, amyloidosis, congenital hypothyroidism, neurofibromatosis, and primary muscular hypertrophy (18). Lymphangioma may occur in association with hemangioma in Maffucci's syndrome (7) or, as in the present case, be confused with hemangioma because of the presence of blood-filled spaces. Unlike hemangioma, juvenile lymphangioma, which accounts for about 6% of all tumors, is benign and is accepted as a true vascular malformation (19). In addition, spontaneous regression of lymphangioma is rare and slow progressive enlargement of tongue lesions may lead to swelling of the tongue. Lymphangioma may also be associated with Turner's syndrome, Noonan's syndrome, trisomies, cardiac anomalies, fetal hydrops, and familial pterygium colli (1). Treatment of lymphangioma is difficult and depends on the size and type of lesion, association with anatomic structures, and infiltration to the surrounding tissues. Although microcystic lesions are diffuse and hard to remove, macrocystic lesions can be localized and excised easily (7). Even though no complications occurred in the present case, the wide area of excision including the full lesion with 2 mm margins of healthy tissue might have resulted in postoperative complications such as wound dehiscence and scarring. In addition, the patient continues to be monitored for signs of recurrence.

Conclusion

Lymphangioma occurs rarely in the tongue. Early recognition allows proper initiation of treatment and prevents the occurrence of complications. Conventional treatment with surgical excision with sufficient depth and width was recommended in the present case, because recurrence is common with other treatment modalities.

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Conflict of interest
None declared.

References

1. Usha V, Sıvasankari T, Jeelani S, Asokan GS, Parthiban J. Lymphangioma of the tongue - a case report and review of the literature. J Clin Diagn Res 2014;8(9):ZD12-ZD14.
2. Bhayya H, Pavani D, Avinash Tejasvi ML, Geetha P. Oral lymphangioma: A rare case report. Contemp Clin Dent 2015;6(4):584-587.
3. Ikeda H, Fujita S, Nonaka M, Uehara M, Tobita T, Inokuchi T. Cystic lymphangioma arising in the tip of the tongue in an adult. Int J Oral Maxillofac Surg 2006;35(3):274-276.
4. Kayhan KB, Keskin Y, Kesimli MC, Ulusan M, Unur M. Lymphangioma of the tongue: Report of four cases with dental aspects. Kulak Burun Bogaz Ihtis Derg 2014;24(3):172-176.
5. Pinto A. Pediatric soft tissue lesions. Dent Clin North Am 2005;49(1):241-258.
6. İlknur T AS, Bıçak MÜ, Özer E, Fetil E. A case of lymphangioma treated with pulsed dye laser. Türkderm 2012;46(2):107-109.
7. Kheur SM SR, Yashwant I, Desai RS. Lymphangioma of tongue: A rare entity. Indian Journal of Dental Advancements 2011;3(3):635-637.
8. Patel NJ, Sciubba J. Oral lesions in young children. Pediatr Clin North Am 2003;50(2):469-486.
9. Sunil S, Gopakumar D, Sreenivasan BS. Oral lymphangioma - case reports and review of literature. Contemp Clin Dent 2012;3(1):116-118.
10. Stanescu L, Georgescu EF, Simionescu C, Georgescu I. Lymphangioma of the oral cavity. Rom J Morphol Embryol 2006;47(4):373-377.
11. de Serres LM, Sie KC, Richardson MA. Lymphatic malformations of the head and neck. A proposal for staging. Arch Otolaryngol Head Neck Surg 1995;121(5):577-582.
12. Brennan TD, Miller AS, Chen SY. Lymphangiomas of the oral cavity: A clinicopathologic, immunohistochemical, and electron-microscopic study. J Oral Maxillofac Surg 1997;55(9):932-935.
13. Goswami M, Singh S, Gokkulakrishnan S, Singh A. Lymphangioma of the tongue. Natl J Maxillofac Surg 2011;2(1):86-88.
14. Uguru C, Edafioghor F, Uguru N. Lymphangioma of the tongue with macroglossia: A case report. Niger J Med 2011;20(1):166-168.
15. Mosca RC, Pereira GA, Mantesso A. Cystic hygroma: Characterization by computerized tomography. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2008;105(5):e65-69.
16. Hartl DM, Roger G, Denoyelle F, Nicollas R, Triglia JM, Garabedian EN. Extensive lymphangioma presenting with upper airway obstruction. Arch Otolaryngol Head Neck Surg 2000;126(11):1378-1382.
17. Alqahtani A, Nguyen LT, Flageole H, Shaw K, Laberge JM. 25 years' experience with lymphangiomas in children. J Pediatr Surg 1999;34(7):1164-1168.
18. Greinwald JH, Jr., Burke DK, Sato Y, Poust RI, Kimura K, Bauman NM, Smith RJ. Treatment of lymphangiomas in children: An update of picibanil (ok-432) sclerotherapy. Otolaryngol Head Neck Surg 1999;121(4):381-387.
19. Orvidas LJ, Kasperbauer JL. Pediatric lymphangiomas of the head and neck. Ann Otol Rhinol Laryngol 2000;109(4):411-421.

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