Oral lymphangioma: A rare case report

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Abstract

Lymphangiomas are benign hamartomatous tumors of the lymphatic channels which present as developmental malformations arising from sequestration of lymphatic tissue that do not communicate with the rest of the lymphatic channels. Lymphatic vessels are filled with a clear protein-rich fluid containing few lymph cells. It can also occur in association with hemangioma. The onset of lymphangiomas are either at birth (60% to 70%) or up to two years of age (90%) and rare in adults. Lymphangiomas have marked predilection for the head and neck region (50-70%). The most common location in the mouth is the dorsum of tongue, followed by lips, buccal mucosa, soft palate, and floor of the mouth. On tongue, they may present as a localized or a diffused growth which may enlarge to cause macroglossia, impaired speech and difficulty in mastication. Herewith, we present a rare case of lymphangioma of tongue leading to macroglossia in a 8-year-old boy.

Keywords: Hemangioma, lymphangioma, lymphohemangioma, oral

Introduction

Lymphangiomas, just like other vascular malformations are lymphatic malformations which are characterized by abnormal proliferation of lymphatic vessels. They are benign hamartomatous tumors which are localized to head and neck region in about 75% of cases.[1] Lymphangioma was First described by Redenbacher in 1828[2] and lymphangiomas of the tongue was first described by Virchow in 1854.[3] A significant number of cases are present at birth, and 90% of cases are developed before 2 years of age.[4] When they occur in the oral cavity, the most common location is dorsum of the tongue, followed by lips, buccal mucosa, soft palate, and floor of the mouth.[5] Clinically, lymphangiomas are slow growing, painless soft tissue mass. The clinical appearance of lymphangioma depends on the extension of the lesion. Superficial lesions consist of elevated nodules with pink or yellowish color or seen as transparent grouped vesicles, which can be red or purple due to secondary hemorrhages.[6] Deeper lesions are described as soft, diffuse masses with normal color.[5] Various methods have been reported for the treatment of lymphangiomas. Surgical excision is mostly indicated when vital structures are not involved.[1,7] Procedures such as radiation therapy, cryotherapy, electrocautery, sclerotherapy, steroid administration, embolization, ligation, and laser surgery have also been proposed to treat lymphangiomas.[3,8,9] The following case report is of a patient with lymphangioma of the tongue and its management.

Case Report

An 8-year-old boy reported to the Department of Oral Medicine and Radiology with a chief complaint of growth on tongue since 6 years. Patient's mother noticed the growth when the patient was of 2-year-old, which was initially smaller in size and gradually increased to present size. Patient also complains of slurred speech and bleeding from the growth. Patient also reported difficulty in swallowing and chewing due to growth. On examination, multiple papular appearances was seen on the left dorsal half of the tongue [Figure 1] of size approximately 4 cm × 3 cm, which was exactly within the midline and not crossing the midline, extending anteriorly from the tip of the tongue to posteriorly 0.5 cm from circumvallate papillae. The surface appears to be irregular and granular. The high lingual frenal attachment was seen presenting as a tongue tie [Figure 2]. On palpation, growth

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was soft, nontender, and pebbly. Reduction in the tongue movements without any loss of sensory or motor functions. Diascopy test was carried out which was negative, and there were no palpable pulsations felt. Based on the history and clinical feature a provisional diagnosis of lymphangioma of the tongue was made, and excisional biopsy under general anesthesia was carried out [Figure 3]. The excised specimen was sent for histopathological examination and diagnosed as lymphangioma. Based on history, clinical and histological features a final diagnosis of lymphangioma was made.

Discussion

Lymphangioma is congenital vascular malformation pertaining to the lymphatic vessels. Lymphangiomas are rare, they account for 4% of all the vascular tumors and 25% of all benign vascular tumors in children. There is no sign of racial predominance and also equal sex incidence reported in most of the studies.

Various theories have been proposed for the pathogenesis of lymphangioma. Of these two major theories that explain the origin of lymphangiomas are as follows.[2] The first theory is that the lymphatic system develops from five primitive sacs arising from the venous system. Concerning the head and neck, endothelial outpouchings from the jugular sac spread centrifugally to form the lymphatic system. Another theory proposes that the lymphatic system develops from mesenchymal clefts in the venous plexus reticulum and spread centripetally toward the jugular sac.

In 1938, Goetsch noted that the sequestered lymphatic tissue forms the cyst, which enlarges from the accumulation of lymph caused by the projection of endothelial sprouts from the cystic walls. The sprouts further destroy tissue and force the lesion into areas of least resistance, between muscles and vessels, invading tissue planes and causing atrophy, fibrosis and hyalinization of the engulfed tissue.[10]

Whimster in 1976 stated that the basic pathological process is the collection of lymphatic cisterns in the deep subcutaneous plane. Whimster thought that the cisterns might come from a primitive lymph sac that does not contact with the rest of the lymphatic system during its embryonic development. A thick coat of muscle fibers that cause rhythmic contractions line the sequestered primitive sacs. Rhythmic contractions increase the intramural pressure, causing dilated channels to come from walls of cisterns toward the skin. Later he suggested that vesicles seen in lymphangioma are out pouching of these dilated projecting vessels. Finally, lymphangioma develops from congenital obstruction or sequestration of the primitive lymphatic enlargement.[11,12] De Serres *et al.* proposed a classification of the lymphangioma of head and neck based on of the spread the anatomical involvement.[13]

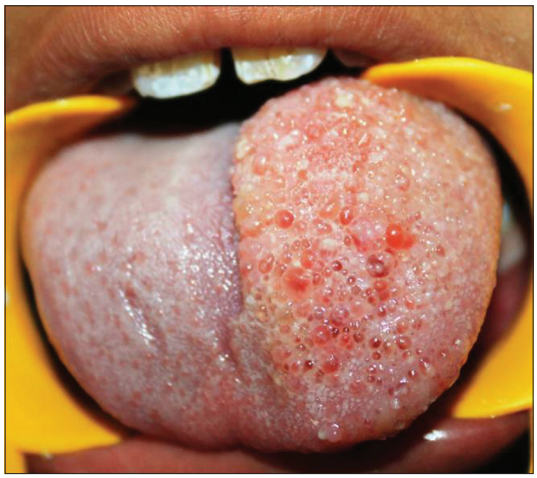

Figure 1: Growth seen on the left dorsal surface of tongue extending from midline to lateral surface

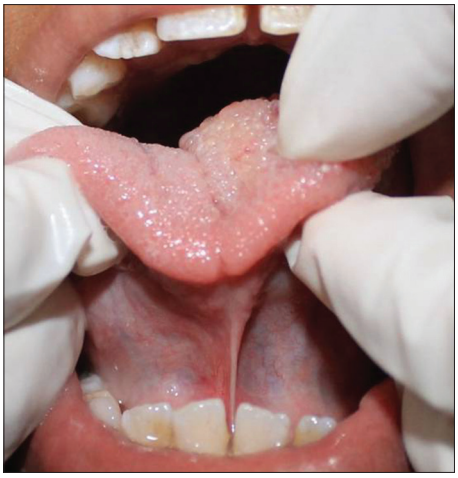

Figure 2: High frenal attachment at the ventral surface of tongue

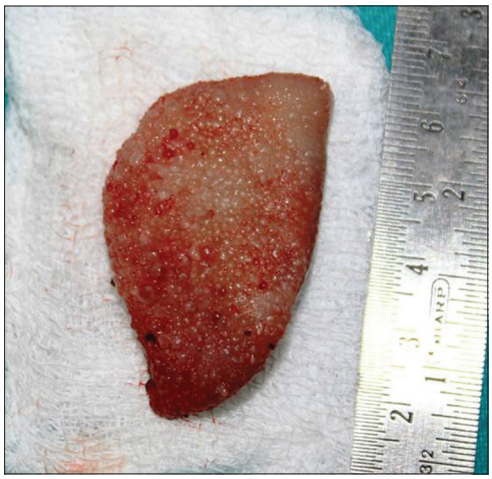

Figure 3: Excised surgical specimen

- Stage/Class I: Infrahyoid unilateral lesions
- Stage/Class II: Suprahyoid unilateral lesions

- Stage/Class III: Suprahyoid and infrahyoid unilateral lesions
- Stage/Class IV: Suprahyoid bilateral lesions
- Stage/Class V: Suprahyoid and infrahyoid bilateral lesions

Lymphangiomas may be present anywhere on the skin and mucosa, commonly seen in head and neck region, followed by the proximal extremities, buttocks, and trunk. Rarely they can be located at intestinal, pancreatic and mesenteric level. Lymphangioma rarely affects the oral cavity. When oral cavity is affected, it may include the tongue, palate, gingival and oral mucosa, lips, and alveolar ridge of the mandible. Brennan *et al*.[14] revised 49 cases of oral lymphangioma and found that 17 were located in tongue while only one case was described at retromolar region and one at soft palate. In the present case, growth was seen involving left dorsal surface of the tongue.

Clinically, oral lymphangiomas manifest as a plaque constituted from small vesicles with thin walls, translucide like frog eggs. Part of the vesicles are full with clear content (lymph), part has a blood content suggesting co-existence of the involvement of the lymphatic anomalies with abnormalities of the blood vessels, which can be appreciated in the present case.

The deep lesions generally are shown as masses of diffuse growth, which depending on the anatomical location may determine damages such as tissue swelling, obstruction of upper airways, pain, tongue's extrusion, sialorrhea, deformity of jaws, as well as difficulties in mastication, speech, and during oral hygiene.[7,15]

If the tongue is affected, anterior dorsal part of tongue is a common site. Pathognomonic features of lymphangioma of the tongue are irregular nodularity of the surface of the tongue with gray and pink projections and macroglossia.[3] These patients will have speech disturbances, poor oral hygiene, and bleeding from tongue associated with oral trauma. In the present case, macroglossia occurred due to a lesion on the left dorsal surface of the tongue and patient presented with the slurring of speech.

Differential diagnosis of lymphangioma includes number of oral lesions such as hemangioma, teratoma, dermoid cyst, thyroglossal duct cyst, amyloidosis, neurofibromatosis, heterotopy of gastric mucosal cyst, and granular cell tumor, all of which cause macroglossia.

Histopathological features of lymphangioma consist of lymphatic vessels with marked dilatations. Vessels are often infiltrated into the adjacent soft tissue and demonstrated as lymphoid aggregate in their wall. Endothelial lining is thin, and the spaces consist of proteinaceous fluid and lymphocytes. Sometimes secondary hemorrhage may be noticed in the lymphatic vessels. The lymphatic space contains lymphatic fluid, red blood cells, lymphocytes, macrophages, and neutrophils.

Histopathologically lymphangioma are classified as:[16,17]

- Lymphangioma simplex (composed of small thin-walled lymphatics)
- Cavernous lymphangioma (comprised of dilated lymphatics vessels with surrounding adventitia)
- Cystic lymphangioma (consisting of huge, macroscopic lymphatic spaces surrounded by fibrovascular tissue, and smooth muscle)
- Benign lymphangioendothelioma (lymphatic channels appears to be dissecting through dense collagenic bundles).

Various treatment modalities have been employed for the treatment of lymphangioma of the tongue. The main objective of the treatment of tongue lymphangiomas is the preservation of the taste sensation, restoration of the tongue size for articulation, and cosmesis.

The various treatment modalities for lymphangioma are surgical excision, radiation therapy, cryotherapy, electrocautery, sclerotherapy, steroid administration, embolization, and ligation, laser surgery with Nd-YAG, CO_2 12 and radiofrequency tissue ablation technique. Surgical excision is the treatment of choice as lymphangiomas are encapsulated or partially circumscribed.[18] However, encapsulation is not always complete, and cellular infiltration of adjacent tissues becomes inevitable. Successful treatment requires the inclusion of a surrounding border of normal tissue, provided that vital structures are not damaged.[19,20] Nd-YAG laser surgery has become widely preferred because of its advantages as less bleeding and edema versus standard methods of surgical resection.[21] Sclerosing agents are ineffective, probably as a result of the discontinuous basement membrane of the lymphatic vessels.[14] In choosing the ablative therapy of lymphangiomas, the exact knowledge of the anatomy, and spread of lesions are very important.

Recurrence is common because of its infiltrative nature. According to the study conducted by Orvidas and Kasperbauer, the recurrence rate for lymphangiomas was 39%.[22] The high percentage of oral involvement correlated with a high degree of persistent disease. The most common sites for recurrence are the tongue and hypopharynx and/or larynx. Because the lesions described in this case report exhibited a superficial location, we opted for their surgical removal. After 1 year of follow-up, no sign of lesion relapse was identified.

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Conflicts of interest
There are no conflicts of interest.

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