Lymphangioma of the tongue - a case report and review of literature

Shetty SR

Department of Oral Medicine and Radiology, AB Shetty Memorial Institute of Dental Sciences, Mangalore, Karnataka

E-mail: drshishirshettyomr@yahoo.com

Abstract

Lymphangiomas are uncommon congenital hamartomas of the lymphatic system, usually diagnosed in infancy and early childhood. They are commonly located in head and neck region and rarely present in the oral cavity. Most common site of occurrence in oral cavity is the tongue and it appears as cluster of translucent vesicles resembling the appearance of frog's egg or tapioca pudding. Early recognition is of utmost importance to initiation of proper treatment, and avoiding serious complications. Herewith, we present a case of lymphangioma of tongue with follow up of 3 years.

Key words: Lymphangioma, Tongue, Macroglossia.

Introduction

Described for the first time by Redenbacher in 1828, currently the lymphangiomas are classified as malformations and not as neoplasms. Lymphangioma is a benign hamartomatous tumor of lymphatic vessels with a marked predilection for the head and the neck region. Lymphangiomas represent about 6% of the total number of benign tumors of the soft tissue in patients aged less than 20 years[1].

Three theories have been proposed to explain the origin of this abnormality. The first suggests that a blockage or arrest of normal growth of the primitive lymph channels occurs during embryogenesis, the second that the primitive lymphatic sac does not reach the venous system, while the third advances the hypothesis that, during embryogenesis, lymphatic tissue lays in the wrong area[2].

Differential diagnosis of lymphangiomas of the tongue include hemangioma, amyloidosis, congenital hypothyroidism, mongolism, neurofibromatosis, various storage diseases and primary muscular hypertrophy of the tongue, all of which may cause macroglossia[3].

Case report

A nine year old boy **(Figure 1)** reported with a complaint of large tongue for last 6 years. The patient was accompanied by his mother who first noticed his abnormally large tongue, when he was three years old. The tongue had gradually enlarged over the years. The patient had difficulty in speaking and chewing food since past six years. Occasional history of burning sensation and halitosis was also given by the patient.
The patient's medical history was non contributory but the patient's mother suffered from pulmonary tuberculosis during pregnancy. The disease was cured with medication

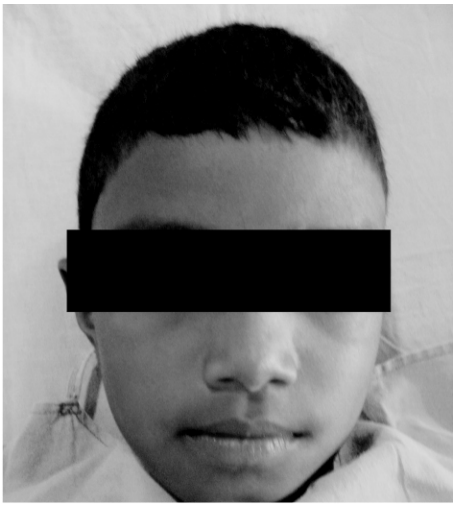

Figure 1 Extraoral view of the patient at 9 years of age

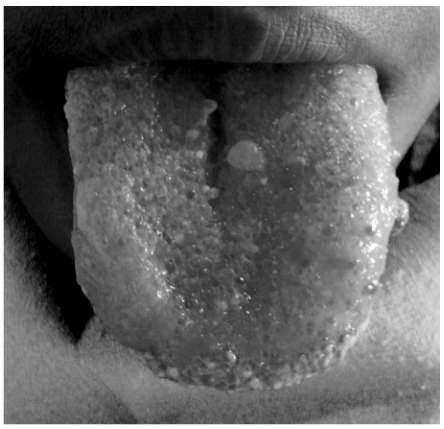

Figure 2 - Dorsal aspect of the tongue - showing enlarged, reddish tongue with multiple granular lesions resembling frog's egg or tapioca pudding and deep fissuring

six months after the delivery. The patient had two elder brothers with no relevant medical problems.
The patient had difficulty in maintaining oral hygiene due to his abnormally large tongue. Intraoral examination revealed abnormally large tongue with granular pink & reddish areas **(Figure 2).** The ventral surface of the tongue on the left side showed reddish areas **(Figure 3)** and the lateral borders showed indentations of dentition. A diascopy test on the reddish areas proved negative. On probing, the surface showed presence of deep fissures on the dorsal surface in between the pebbled areas **(Figure 4)**.
Based on the typical "tapioca pudding" or "frog's egg" appearance presented by our patient, a provisional diagnosis of lymphangioma of tongue was made. Ultrasound scanning could not be performed because the tongue couldn't be stabilized. Patient was unwilling for biopsy or surgical excision.
Immediate treatment rendered to patient was oral prophylaxis and restoration of decayed tooth. The patient was kept on follow up of six months interval for a period of three years during which there was a considerable decrease in size of lesion and fissuring, reduced reddish areas and marked decrease in pain and burning sensation **(Figures 5,6,7,8)**. After three years we were able to convince the patient for biopsy. Incisional biopsy was then performed and histopathology revealed stratified squamous epithelium with areas of hyperkeratosis, irregular elongated rete- ridges, and acanthosis. The sub epithelial tissue shows dilated lymphatic spaces of irregular size and shape. The lymphatic spaces are lined by a thin endothelial lining and contain a proteinaceous fluid **(Figure 9, 10)**.

Discussion

Lymphangiomas are benign hamartomatous tumors of the lymphatic vessels. They most likely present as developmental malformations arising from sequestration of lymphatic tissue that do not communicate with the rest of the lymphatic channels.
Lymphangiomas have been classified into:-[4]
1. Lymphangioma simplex (capillary lymphangioma), which consists of small, capillary sized vessels.

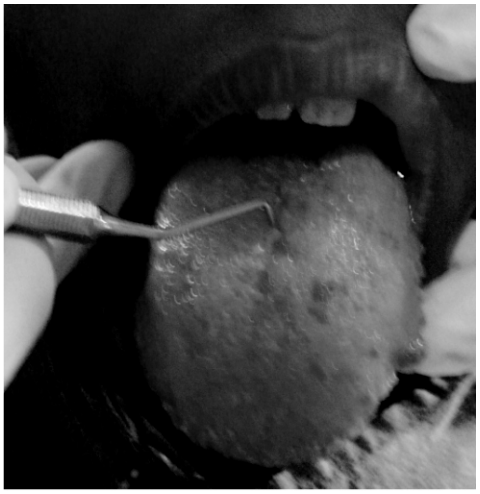

Figure 4 - deep fissuring present on the dorsal surface of tongue

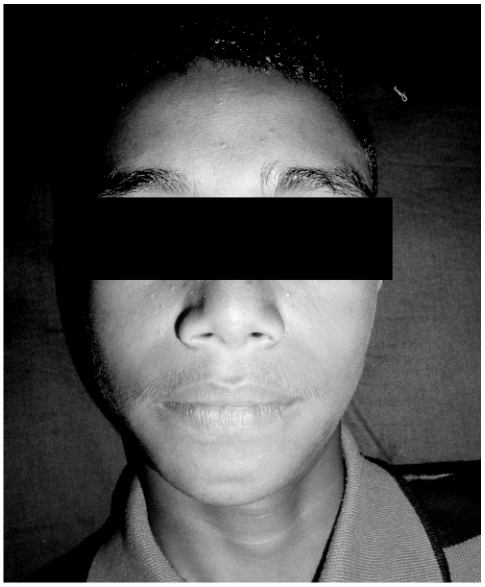

Figure 5 - Extraoral view of the patient at 12 years of age

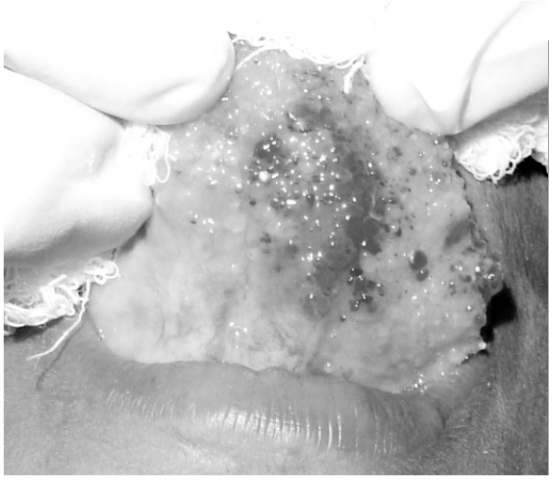

Figure 3 – Ventral surface of the tongue showing erythematous vesicle-like lesions.

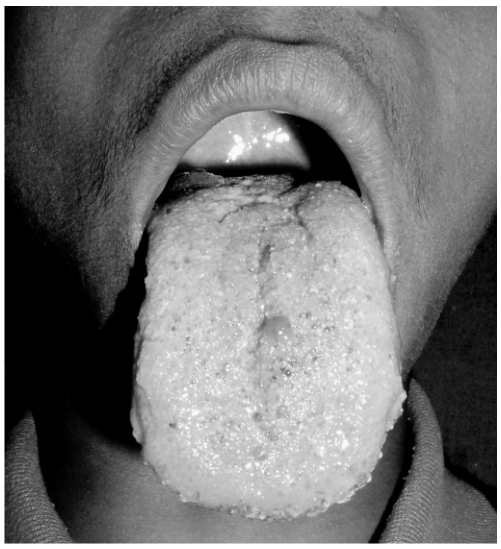

Fig 6 - Marked reduction in the size of erythematous granular areas on dorsal surface of tongue after 3 years

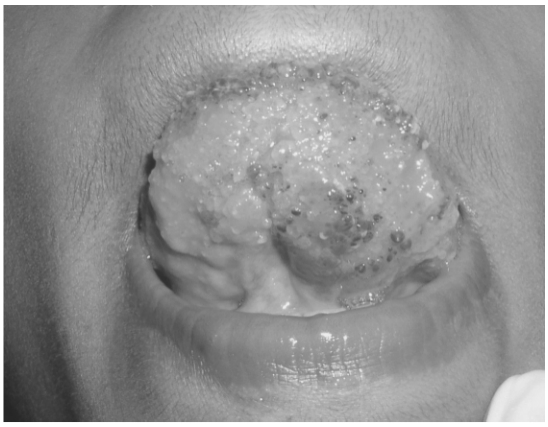

Figure 7 - Ventral surface of the tongue showing reduction in erythematous vesicle - like lesions at 12 years of age

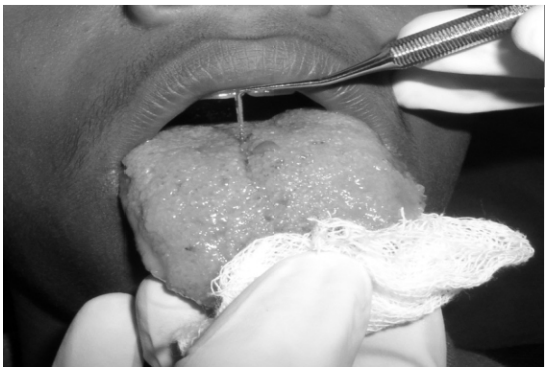

Figure 8 - Marked reduction in fissuring present on the dorsal surface of tongue

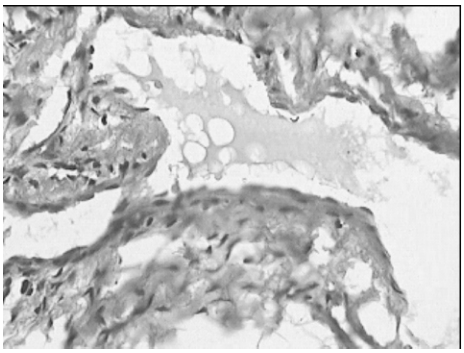

Figure 9- Photomicrograph (low power - 10X) showing stratified squamous epithelium with areas of hyperkeratosis, irregular elongated rete - ridges, and acanthosis. The sub epithelial tissue shows dilated lymphatic spaces of irregular size and shape.

2. Cavernous lymphangioma, which is composed of large, dilated lymphatic vessels.
3. Cystic lymphangioma (cystic hygroma), which exhibits large macroscopic cystic spaces.

Lymphangiomas have a marked predilection for the head and neck region, which accounts for about 75% of all cases. About 50% of the lesions are noted at birth and around 90% develop by 2 years of age. Oral lymphangiomas may occur at various sites but they are most frequently seen on the anterior two-thirds of the tongue, which often results in macroglossia.

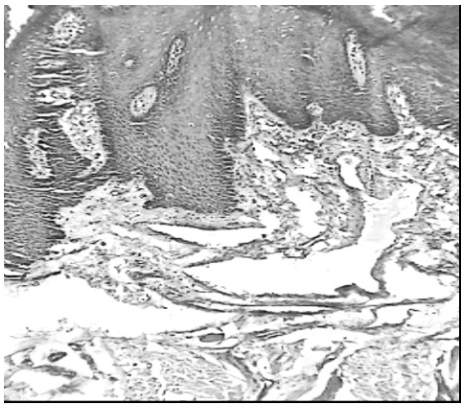

Figure 10- Photomicrograph (high power - 40X) showing lymphatic spaces lined by a thin endothelial lining and containing proteinaceous fluid.

These patients tend to have speech disturbances, poor oral hygiene, and bleeding from tongue associated with oral trauma. Our patient also reported with similar complaints. It can also present in the palate, buccal mucosa, gingiva, and lip. The tumor is superficial in location and demonstrates a pebbly surface that resembles a cluster of translucent vesicles. The surface resembles the appearance of frog's egg or tapioca pudding. Secondary hemorrhage into the lymphatic spaces may cause these vesicles to become purple. Similar clinical presentation was seen in our case. The deeper lesions appear as a nodule or masses without significant change in surface texture or color[4].

Histologically, lymphangiomas are composed of lymphatic vessels that may show marked dilatation (cavernous hemangioma) or macroscopic cyst like structure (cystic hygroma). The lining endothelium is thin and the spaces contain proteinaceous fluid and occasional lymphocytes. In intraoral tumors, the lymphatic vessels are characteristically located just beneath the epithelial surface and often replace the connective tissue papillae. This superficial location results in translucent, vesicle like clinical appearance[4].

For lesions that are asymptomatic and does not interfere much with function, the management is through watchful observation as some lesions have tendency to regress with time. As in our case, the patient did not complain much of interference in function and was kept for observation for 3 years during which the lesion seemed to regress considerably.

For larger lesions, complete surgical excision is treatment of choice but this is not always possible like in cases of extensions into base of tongue, floor of mouth, larynx, neurovascular structures of neck and mediastinal extension.

Earlier, steroids, electro coagulation, cryotherapy or radiation therapy has been used with variable results. Recurrent, residual, unresectable or surgically challenging tumors are treated with intralesional injection of sclerosing agents like 25% dextrose, hypertonic saline, bleomycin, OK-432 before surgery. A change in consistency of the tumor, manifests by softening, is followed by marked shrinkage. In recent years Carbon dioxide[6] and Neodymium Yitrium Aluminum Garnet (Nd-YAG) laser photocoagulation [9]surgery has become popular.

Sclerosants used previously diffuse through discontinuous basement membrane, which allowed the sclerosing agent

to penetrate freely in to the connective tissues, diluting the concentration and rendering less effective. Some agents could affect the skin resulting in scar formation[5,7]. Bleomycin is not used now-a-days as it results in pulmonary fibrosis leading to death. Presently, OK-432 is the preferred intralesional sclerosant which is a lyophilized incubation mixture of the low-virulent streptococcus pyogenes strain and penicillin G potassium[5,7,8]. When administered intralesionally, it evokes inflammation and infiltration of neutrophils and macrophages into the cystic spaces. There is also increase in activity of natural killer cells, helper cells, and T-cells. In addition cytokines (Interlukin-6 and Tumor Necrosis Factor) are also produced. This results in interaction between these activated cells and cytokines directly on the endothelial cells to increase its permeability causing accelerated lymphatic drainage and increased lymph flow leading to shrinkage of the cystic spaces without skin damage and scar formation[5, 7, 8].

Conclusions

Although rarely encountered in the oral cavity, lymphangiomas represent a condition that must be recognized early. A pediatrician may be the first person to come across such lesion and may aid in early recognition and proper treatment of the patient.

References

1. Ligia S . Lymphangioma of the oral cavity. Romanian J Morphol Embryol 2006; 47:373–377.
2. Grasso DL. Lymphangiomas of the head and neck in children. Acta Otorhinolaryngol Ital 2008; 28:17-20.
3. Eversole LR. Benign tumors of the oral cavity. In: Greenberg Martin S, Glick Michael. Burket's Oral Medicine: Diagnosis and treatment. 10th ed. Spain, BC Decker Inc, 2003; 137-193.
4. Neville BW, Damn DD, Allen CM, Bouquet JE. Oral and Maxillofacial Pathology. 3rd ed. India, Saunders, 2009: 507-570.
5 Jeeva Rathan J, Harsha Vardhan BG, Muthu MS, V, Saraswathy K, Sivakumar N. Oral lymphangioma: A case report. J Indian Soc Pedod Prev Dent 2005; 23:185-189.
6 Barak S, Katz J, Kaplan I. The CO2 laser in surgery of vascular tumors of the oral cavity in children. J Dent Child 1991; 58:293-296.
7 Ogita S, Tsuto T, Deguchi E, Tokiwa K, Nagashima M, Iwai N. OK- 432 Therapy for unresectable lymphangiomas in children. J Pediatr Surg 1991; 26:263-270.
8 Ogita S, Tsuto T, Nakamura K, Deguchi E, Iwai N. OK-432 therapy in 64 patients with lymphangioma. J Pediatr Surg 1994; 29:784.
9 Tasar F, Turner TF, Sener BC, Sencift K. Lymphangioma treatment with NdYAG laser. Turk J Pediatr 1995; 37:253-256.