Lymphangioma of the Tongue

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ABSTRACT
Lymphangiomas are benign, hemartomatous tumors of lymphatic vessels. They most likely represent developmental malformations that arise from sequestrations of lymphatic tissues that do not communicate normally with rest of lymphatic system. Our present knowledge of lymphangioma indicates that it has a predilection for the head and neck, which accounts for 50 to 75% of all cases. About half of all lesions are noticed at birth and around 90% develop by 2 years of age. Lymphangioma is a rare medical condition and not many cases are reported in scientific literature. A case of lymphangioma of tongue in a 17-year-old male is reported.

Keywords: Amyloidosis, Hemangioma, Lymphangioma, Macroglossia, Neurofibromatosis.

INTRODUCTION
Lymphangioma is considered to be a lymphatic malformation, similar to other vascular malformations, yet characterized by an abnormal proliferation of lymphatic vessels. Clinically, lymphangiomas are a slow growing and painless soft tissue mass. This frequently presents without a clear anatomic outline, dissects tissue planes, can be very extensive.1

Three subtypes of lymphangioma have been reported. The subtypes are probably variants of the same pathologic process, and size of vessels may depend on nature of surrounding tissues. Cystic lymphangiomas most often occur in neck and axilla, where loose adjacent connective tissues allow for more expansion of vessels. Cavernous lymphangiomas are more frequent in mouth, where the denser surrounding connective tissue and skeletal muscle limit vessel expansion. Cervical lymphangiomas are more common in posterior triangle of neck and are typically soft, fluctuant masses. They occur less frequently in anterior triangle. Occasionally, they may extend into mediastinum or oral cavity. Such tumors can be massive and measure equal or more than 15 cm in size.2

Oral lymphangiomas may occur at various sites but are most common on anterior two-third of tongue, where they often result in macroglossia.

CASE REPORT
A 17-year-old male reported to the Department of Oral Medicine and Radiology at Seema Dental College and Hospital, Rishikesh, with a chief complaint of abnormal projections on tongue since 10 years. Patient had undergone extraction of 38 under LA one month back with prolonged postextraction bleeding.

Detailed examination of the dorsum of tongue showed distribution of multiple granule-like projections (Fig. 1). These projections were of various sizes and shapes and of different colors—white, purple and red. Similar appearance was seen on ventral surface also, though the white projections were fewer in number. Deep fissure was present on both sides of midline on dorsum of tongue. Slight enlargement of the tongue was also noted. The mucosa appeared normal in other areas of the oral cavity. The clinical appearance of tongue was highly suggestive of lymphangioma.

On microscopic examination, lymphatic vessels diffusely infiltrate adjacent soft tissues and demonstrate lymphoid aggregates in their walls. Some channels contain RBC, which creates uncertainty as to whether they are lymphatics or blood vessels.

DISCUSSION
Lymphangioma is a lymphatic malformation, a benign proliferation of lymph vessels, forming a yellowish tumor on skin, composed of a mass of dilated lymph vessels. Lymphangiomas are rare, they account for 4% of all vascular tumors and approximately 25% of all benign vascular tumors in children. There is no sign of racial predominance. Also equal sex incidences are reported in most studies. Lymphangiomas can become evident at any age, but often occur at birth or in early stages of life. Small lymphangiomas less than 1 cm in size occur on alveolar ridge in around 4% of black neonates.3,4

In 1976, Whimster5 studied the pathogenesis of lymphangioma. He said that the basic pathologic process is the collection of lymphatic cisterns (a receptacle for holding water or other liquid) in deep subcutaneous plane. These cisterns are separated from normal network of lymph vessels, but they communicate with superficial lymph vessels through vertical,
dilated lymph channels. Whimster thought that the cisterns might come from a primitive lymph sac that does not connect with the rest of lymphatic system during its embryonic development. A thick coat of muscle fibers that cause rhythmic contractions line the sequestered primitive sacs. Rhythmic contractions increase intramural pressure, causing dilated channels to come from walls of cisterns toward the skin. He suggested that vesicles seen in lymphangioma are outpouchings of these dilated projecting vessels. Lymphatic and radiographic studies support Whimsters’s observations.

Differential diagnosis of lymphangiomas of tongue includes hemangioma, congenital hypothyroidism, mongolism, amyloidosis, neurofibromatosis, primary muscular hypertrophy of the tongue, all of which may cause macroglossia.1

Treatment includes surgical excision (most common) and sclerotherapy. Recurrence is common because of their infiltrative nature. Prognosis is good for most of the patients, although large tumors of neck or tongue may result in airway obstruction or death.5,7 Some success with sclerotherapy for unresectable lymphangiomas has been reported using OK-432, a lyophilized incubation mixture of a low-virulent strain of streptococcus pyogenes with penicillin G potassium, which has lost its streptolysin S-producing ability.8

In the present case, the patient was not willing for any treatment for his tongue lesion.

REFERENCES