

Lymphangioma of the Tongue - A Case Report and Review of Literature

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ABSTRACT

Lymphangiomas are benign tumours resulting from a congenital malformation of the lymphatic system. They are relatively uncommon and usually diagnosed in infancy and early childhood. Commonly located at head and neck, they rarely occur in the oral cavity. Intraoral lymphangiomas occur more frequently on the dorsum of tongue, followed by palate, buccal mucosa, gingiva, and lips. Lymphangioma of the tongue is a common cause of macroglossia in children associated with difficulty in swallowing and mastication, speech disturbances, airway obstruction, mandibular prognathism, openbite and other possible deformities of maxillofacial structures. We present the case of a 13-year-old female with lymphangioma of tongue. The clinical, radiological, and treatment modalities of this case are discussed.

Keywords: Congenital malformation, Lymphangioma, Macroglossia

CASE REPORT

A 13-year-old girl reported to my clinic with chief complaint of burning sensation of the tongue and inability to chew food due to lack of contact of teeth since four years. History reveals that she was apparently normal till 40 day after birth, after which a growth was observed in the tongue. On consultation with pediatric surgeon it was found to be a hyperplastic growth on the ventral surface of the tongue and was diagnosed as lymphangioma of the tongue and it was treated with surgical excision. Following the treatment patient was apparently normal for next 3 years after which the growth gradually increased in size and progressed to involve currently the entire dorsal surface, ventral surface and lateral border of the tongue. There was no relevant family history. On general examination patient was moderately built and nourished with vital signs within normal limit. On review of system there was slurring of speech with no evidence of dyspnea and dysphagia. On extra oral examination there was no gross facial asymmetry and the mouth opening was within normal limit.

On part of intraoral examination on inspection of soft tissues, dorsal aspect of the tongue appeared enlarged with pebbles like multiple red, blue, grey and clear vesicles [Table/Fig-1]. Tip of the tongue was also involved as a part of lesion [Table/Fig-2]. A prominent mass was seen on the base of the tongue, approximately 2x3 cm in size with numerous papillary and vesicle-like projections giving

an irregular and granular appearance [Table/Fig-3]. On palpation, it was soft, pebbly and slightly tender in nature. There was no restriction in functions of the tongue. On hard tissue examination anterior and posterior open bites were observed. Based on history and the clinical features the lesion was diagnosed as lymphangioma of tongue.

As part of investigation to rule out any vascular anomalies, a CT angiogram was done.

The following observations have been noted after analysing the CT angiography of tongue:

- It showed low velocity of blood supply
- It revealed no abnormalities of blood vessels in the tongue [Table/Fig-4].

The above observations are explained as follows:

As a consequence of low velocity of blood supply, there is no evidence of dilatation of the lingual artery bilaterally. Also, no dilated draining vein is seen extending from the tongue lesion to the internal jugular vein on either side. Apart from the above observations, it was noted that the common carotid, internal, external carotid and vertebral arteries are normal bilaterally.

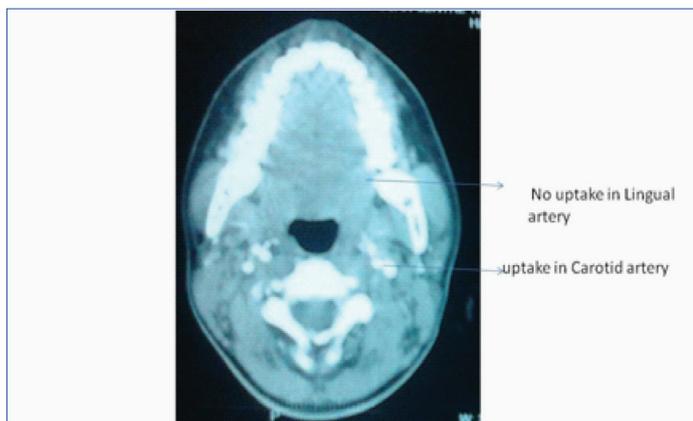
The patient was advised for surgical excision. However, the patient was not willing for treatment since there was no dysphagia or dyspnoea. Also, there was no cosmetic concern.



[Table/Fig-1]: Shows enlarged dorsal aspect of the tongue with pebbled surface

[Table/Fig-2]: Reveals multiple red, blue, white and clear vesicles on the tip of the tongue

[Table/Fig-3]: A prominent mass with clear pebbly surface at the base of the tongue



[Table/Fig-4]: Showed low velocity of blood supply and no abnormalities of the blood vessels in the tongue

DISCUSSION

Lymphangioma is a rare, benign, congenital malformation of unknown aetiology that originates from lymph vessels and this entity was first described by Virchow in 1854 [1]. Lymphangiomas are uncommon congenital hamartomas of the lymphatic system. Lymphangiomas have a marked predilection for the head and neck region, which accounts for about 75% of all cases and about 50% of these lesions are noted at birth and around 90% develop by two years of age. However, diagnosis of lymphangioma in adults is a rare occurrence [2]. The clinical appearance of lymphangioma depends on the extension of the lesion. Superficial lesions consist of elevated nodules with pink or yellowish color. Deeper lesions are described as soft, diffuse masses with normal color [3].

Two major theories have been proposed to explain the origin of lymphangiomas. The first theory is that the lymphatic system develops from five primitive sacs arising from the venous system. Concerning the head and neck, endothelial outpouching 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 towards the jugular sac. Finally, lymphangioma develop from congenital obstruction or sequestration of the primitive lymphatic enlargement [4].

Lymphangioma development is commonly observed in the neck aspect related to posterior triangle involving sites such as posterior border of sternocleidomastoid muscle, upper middle third of clavicle and anterior edge of trapezius [5]. However, at times anterior triangle also can become involved. Other associated areas are parotid and submandibular regions [6].

The most common locations are the head and neck, followed by the proximal extremities, buttocks and trunk. Sometimes they can be located at intestinal, pancreatic and mesenteric level.

Lymphangiomas rarely affect the oral cavity. Affected sites in the oral cavity may include the tongue, palate, gingiva, lips, and alveolar ridge of the mandible [7,8]. In the present case, tongue was involved wherein the dorsal aspect, tip and base of the tongue were involved.

In the oral cavity, this lesion is common in the first decade of life and mostly occurs on the dorsal surface and lateral border of the tongue. It rarely arises on palate, gingiva, buccal mucosa and lips which overlap with the present case. The anterior two-thirds on the dorsal surface of the tongue is the most common site for intraoral lymphangiomas leading to macroglossia. These patients tend to have speech disturbances, poor oral hygiene, and bleeding from the tongue associated with oral trauma. In this case the anterior two third was largely involved which was associated with disturbance in speech.

In this case though the patient has a satisfactory oral hygiene, white vesicles and macroglossia of the tongue is seen with patient complaining of burning sensation as well, suggestive of superimposed infection and slurring of speech due to enlarged tongue. When it occurs in the oral cavity it manifests as clear vesicles and the surface appears granular with translucent hue especially when it is superficial in nature. In certain situations due to disruption of the blood capillary into the lymphatic inner space it appears blue or red in color [9,10].

The clinical appearance of the lesion varies based on its whether it is superficial or deep. Superficial lesion present as papillary lesions with pebbly surface due to the occurrence of several translucent vesicles with same color as that of adjacent mucosa or occasionally with a mild reddish hue. Interestingly they give a tapioca pudding or frog eggs like appearance. The deeper lesions manifest as diffuse nodules which are soft in consistency and with negligible alterations in color or texture [11].

The consequences of deep seated lesions may result in swelling of tissues leading to obstruction in upper airway, extrusion of tongue, increased salivation, jaw deformity, pain and poor oral hygiene. Also marked problems with respect to chewing and speaking may result [12].

Whenever lymphangioma show a rapid growth an underlying factor such as an associated tumours or respiratory tract infection may be responsible [13].

The complications of lymphangioma may affect the patient broadly in four dimensions such as aesthetic, occlusal, functional and psychosocial aspects [14]. Complications related to infection can occasionally result in Ludwig's angina associated with an infected base of the tongue lymphangioma [15]. Seroma formation, Infections, minor bleeding, recurrent cellulitis, and lymph fluid leakage are some of the few postoperative complications of oral and cervical lymphangioma [16].

A classification of the lymphangioma of head and neck on the base of the anatomical involvement had been proposed by De Serres LM.

- 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;
- Stage/class IV – infrahyoid bilateral lesions [17].

According to their clinical presentation they are classified into macrocystic (cavities larger than about 2cm³), microcystic (cavities smaller than about 2 cm³), and mixed (combining these two types). The objectives of treatment of lymphangiomatous macroglossia are preservation of taste, restoration of tongue size for articulation, correction of mandibular and dental deformities, and cosmetics.

Histopathologically they are classified as capillary, cavernous and cystic lymphangioma. Marked dilatation of lymphatic vessels is appreciated histopathologically. Small capillary sized vessels, large dilated lymph channels and large macroscopic cystic spaces are respectively appreciated microscopically in capillary, cavernous and cystic lymphangioma [18]. Cavernous lymphangioma is the most common intra oral type [19]. Lymphangiomas are known to be associated with Turner's syndrome, Noonan's syndrome, trisomies, cardiac anomalies, fetal hydraps, fetal alcohol syndrome, and Familial pterygium colli [20].

The differential diagnosis for lymphangioma includes Hemangioma, Amyloidosis, Congenital hypothyroidism, Neurofibromatosis, Mongolism, Primary muscular hypertrophy [21].

The treatment of lymphangioma depends upon their type, size, involvement of anatomical structures and infiltration to the surrounding tissues. Microcystic lesions do not respect tissue planes, are diffuse and difficult to eradicate, whereas macrocystic lesions are localized and easily excised. The various treatment modalities for lymphangioma are surgical excision, radiation therapy, cryotherapy, electrocautery, sclerotherapy, steroid administration, embolization, and ligation, laser surgery with Nd-YAG, CO₂ and radiofrequency tissue ablation technique.

Laser photocoagulation has been reported useful in controlling the tongue size and removing superficial lymphangioma in some cases. OK-432, a lyphophilized incubation mixture of *Streptococcus pyogenes* and penicillin G potassium has been used to treat lymphangiomas. The sclerosing agent OK-432 is effective for macrocystic lymphatic malformations but showed less promise for microcystic lesions, mixed lesions, and lesions outside the head and neck region [22-24].

CONCLUSION

Although rarely encountered in the oral cavity, their early recognition allows proper initiation of treatment and prevents the occurrence of the complications. Also noteworthy is, follow up to prevent recurrence.

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