

Lymphangioma of tongue: A case report and review of literature

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Abstract

Lymphangioma is a benign hamartomatous hyperplasia of lymphatic vessels, with three-fourths of all cases occurring in the head and neck region. The complications of lymphangioma may affect the patient broadly in four dimensions such as aesthetic, occlusal, functional and psychosocial aspects. Complications related to infection can occasionally result in Ludwig's angina associated with an infected base of the tongue lymphangioma. Therefore it is important to diagnose and treat the lesion at the earliest. Here we present a case of lymphangioma of tongue in a 4-year-old female patient.

Keywords: lymphangioma, macroglossia, tongue.

1. Introduction

Lymphangioma is a benign hamartomatous hyperplasia of lymphatic vessels, with three-fourths of all cases occurring in the head and neck region.[1-4] Lymphangioma was first described by Redenbacher in 1828 and lymphangiomas of the tongue was first described by Virchow in 1854.[2] The onset of lymphangiomas are either at birth (60 to 70%) or up to two years of age (90%). It is rare in adults.[3-6]

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.[2,6] 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.[2,4] 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.[2]

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.[6] The most common head and neck location is the lateral neck, where this lesion typically contains large cystic spaces and is commonly called cystic lymphangioma or cystic hygroma.[1] The intraoral lymphangioma most commonly occurs on the tongue, but is seen also on the palate, buccal mucosa, gingiva and lips. The superficial lesions are manifested as papillary lesions which may be of the same color as the surrounding mucosa or of a slightly redder hue. The deeper lesions appear as diffuse nodules or masses without any significant change in surface texture or color.[1,5] 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.[6] If the tongue is affected, considerable enlargement may occur, and

to this clinical feature the term 'macroglossia' may be applied.[1,5] Macroglossia may lead to difficulty in swallowing and mastication, speech disturbances, exclusive nasal breathing, airway obstruction, mandibular prognathism and other possible deformities of maxillofacial structures.[2,5] The complications of lymphangioma may affect the patient broadly in four dimensions such as aesthetic, occlusal, functional and psychosocial aspects. Complications related to infection can occasionally result in Ludwig's angina associated with an infected base of the tongue lymphangioma.[6] Hence it is inevitable to treat lymphangioma as early as possible.

2. Case report

A 4- year- old female patient reported to our surgery department with a growth on the left lateral border of tongue, associated with biting of the tongue on mastication. Her parents noticed it increasing in size along with the growth of the child. The patient also complained of bleeding from the lesion site on incurring trauma. On inspection, there was a bluish colored marked soft tissue growth with numerous papillary and vesicle- like projections which made it appear irregular and granular [Figure 1]. On palpation, the swelling was soft, non-tender and pebbly. The mouth opening was normal and there was no restriction of functions of the tongue however slurring of speech was noted. On the basis of history and clinical features a provisional diagnosis of lymphangioma was made. Under general anesthesia, a deep suture was placed on the tip of the tongue for retraction. Single linear incision was outlined on the base of the elevated mass on the lateral surface of the tongue. Initially, the incision was mucosal all around the elevated mass and subsequently it was deepened to excise it completely.

Following excision, the tongue was approximated in two layers. The excised mass measured 2.5×2×1.5 cm. On histopathological examination it was confirmed to be a lymphangiomatous lesion. The patient was discharged after 4 days with a normal looking tongue. There was no evidence of recurrence of the lesion on follow-up of 2 years.

Figure 1: Lymphangioma of tongue



3. Discussion

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. The anterior two-thirds on the dorsal surface of the tongue is the most common site for intraoral lymphangiomas leading to macroglossia which was consistent finding in presented case. These patients tend to have speech disturbances, poor oral hygiene, and bleeding from the tongue associated with oral trauma.⁶ In this case the anterior two third was largely involved which was associated with disturbance in speech. 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[6] which explains the blue color of lesion in the presented case.

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).[4,6] A classification of the lymphangiomas has been suggested by Watson and McCarthy based upon their study of 41 cases. In this classification the following divisions are proposed: (1) simple lymphangioma, (2) cavernous lymphangioma, (3) cellular or hypertrophic lymphangioma, (4) diffuse systemic lymphangioma, and (5) cystic lymphangioma or hygroma.[1]

However a simplified histopathological classification includes 4 categories:[1-4]

- **Lymphangioma simplex** (capillary lymphangioma, lymphangioma circumscriptum), composed of small, thin-walled lymphatics.
- **Cavernous lymphangioma** comprised of dilated lymphatic vessels with surrounding adventitia.
- **Cystic lymphangioma** (cystic hygroma), consisting of huge, macroscopic lymphatic spaces with surrounding fibrovascular tissues and smooth muscle.
- **Benign lymphangioendothelioma** (acquired progressive lymphangioma), lymphatic channels appear to be dissecting through dense collagenic bundles.

These categories are somewhat artificial and many lesions are combinations of categories. The differential diagnosis for lymphangioma includes Hemangioma, Amyloidosis, Congenital hypothyroidism, Neurofibromatosis, Mongolism, Primary muscular hypertrophy.[2,6] Lymphangiomas are known to be associated with Turner's syndrome, Noonan's syndrome, trisomies, cardiac anomalies, fetal hydrops, fetal alcohol syndrome, and Familial pterygium colli.[6]

Histopathologic features consist of lymphatic vessels with marked dilatations. The vessels often diffusely infiltrate the adjacent soft tissues and demonstrate lymphoid aggregates in their walls. The lining endothelium is thin and the spaces contain proteinaceous fluid and lymphocytes. Secondary hemorrhage may be noticed in the lymphatic vessels. The

lymphatic spaces contain lymphatic fluid, red blood cells, lymphocytes, macrophages, and neutrophils. Surrounding connective tissue stroma consists of loose fibrotic tissue with a number of inflammatory cells.[1,2,7]

The treatment of lymphangioma depends upon their type, size, involvement of anatomical structures and infiltration to the surrounding tissues.[2-4,6] 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.[6] Microcystic lesions do not respect tissue planes, are diffuse and difficult to eradicate, whereas macrocystic lesions are localized and easily excised. Treatment is aimed at complete surgical excision. Partial surgical excision, injection of sclerosing solutions (OK432), electrocoagulation, cryotherapy, embolization, steroid administration, radiation and laser surgery may be the other modalities of treatment of diffuse lymphangioma of the tongue. Since they do not respond to sclerosing agents, pressure therapy, radiotherapy or any known chemotherapy, they are either tolerated by the patient or treated surgically. [2-4,6] Seroma formation, infections, minor bleeding, recurrent cellulitis, and lymph fluid leakage are some of the few postoperative complications of oral and cervical lymphangioma.[6]

Dental preventive programs must be performed especially for children with macroglossia until surgery is possible. The inability to perform normal dental hygiene activities increases the risk of caries and gingivitis. Adequate surgery helps patients to keep the tongue inside as a good cosmetic result and they also have less orthognathic deformities.[5]

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