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Case Report:

Lymphangioma Neck Presenting as a Secondary Lesion of the Tongue

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Abstract: Lymphangiomas are uncommon, benign, congenital tumors of the lymphatic system, usually occurring in the head and neck but rarely involve the tongue. They cause both aesthetic anomalies and obstructive symptoms. Early recognition and surgical excision of these tumors is essential. In recent years, sclerosing therapy with OK-432 has become the preferred treatment. This case presents a very unusual presentation of a lymphangioma neck presenting as a secondary lesion on the tongue post sclerotherapy.

Key Words: Lymphangioma neck; Tongue; Sclerotherapy; Excision.

Introduction:

Lymphangiomas are congenital malformations of lymphatic vessels filled with a clear protein-rich fluid, containing few lymph cells. They occur due to sequestration of lymphatic tissue that does not communicate with the rest of the lymphatic channels.[1] When lymphangioma becomes large and extends into the neck, it is referred to as cystic hygroma. Most (75%) of all lymphangiomas occur in the head and neck region of which half are noted at birth and 90% of these lesions develop by 2 years of age. Cystic hygromas may suddenly increase in size due to infection or hemorrhage but can also shrink spontaneously. Three theories have been proposed to explain the origin of this abnormality. The first suggests that during embryogenesis there is arrest of normal growth of the primitive lymph channels, the second that the primitive lymphatic sac does not reach the venous system, while the third advances that, during embryogenesis, lymphatic tissue lies in the wrong area.[2]

Oral lymphangiomas involving the tongue often results in macroglossia. The palate, buccal mucosa, gingiva, and lip are other common sites of invovement.[3,4] The tumor is superficial in location and demonstrates a pebbly surface that resembles a cluster of translucent vesicles. The deeper lesions appear as a nodule or masses without significant change in surface texture or color.

Herewith, we present a very unusual presentation of a lymphangioma neck presenting as a secondary lesion on the tongue, 2 years post sclerotherapy in a 3 years old girl.

Case Report:

A 3 years old female child presented to the otolaryngology out patient department with swelling over the dorsum of the tongue for the last three months. It was insidious in onset, not associated with any difficulty in swallowing, respiratory discomfort, pain, bleeding or history of trauma. On examination, 1.5x2 cm lesion with irregular nodularity and presence of numerous papillary pink projections was noted on the dorsal surface at the posterior one-third of the tongue (Fig 1). However the tongue was normal in size. According to the parents, when the child was one year of age, she had history of swelling in the left submandibular region, 2.4x1.6 cm in size which was diagnosed as cystic hygroma in the Department of Paediatric surgery of our institute. CT scan revealed a heterogenously enhancing cystic lesion with a thick enhancing wall and multiple septae involving the left submandibular, retropharyngeal and parapharyngeal spaces, pushing the trachea towards the opposite side causing compromise of the airway (Fig 2). The patient had received 3 injections of intralesional bleomycin at the dose of 0.3 units/ kg of body weight, following which the swelling had completely subsided. The patient remained asymptomatic for 2 years after which the patient developed this lesion on the tongue and presented in the department of Otolaryngology. The lesion looked liked a papilloma. An excisional biopsy of the tongue lesion was done and the histopathological was suggestive of lymphangioma circumscriptum revealing dialated lymphatic channels and collection of lymphocytes and vascular proliferation (Fig 3). Postoperative period was uneventful. On retrospective analysis of CT scan findings, we found that the neck lesion was reaching up to the posterior border of the tongue.

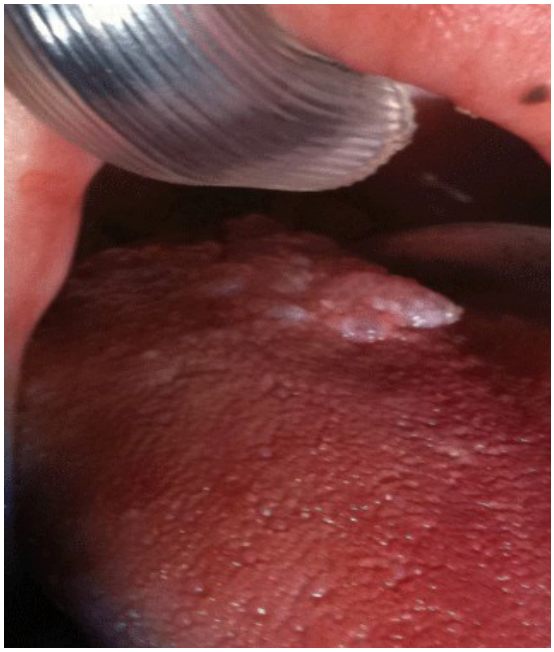

Figure 1: Papillary pink projections noted on the dorsal surface at the posterior one-third of the tongue.

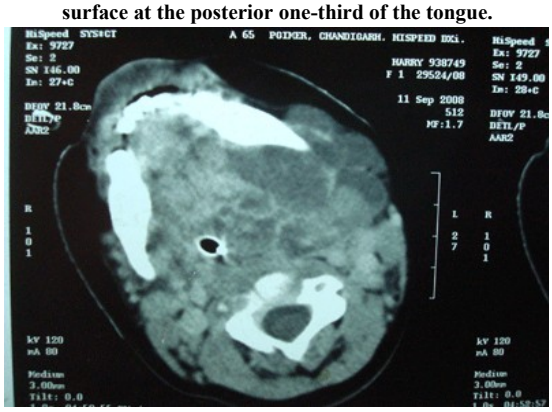

Figure 2: Contrast Enhanced Computed Tomography scan showing heterogenously enhancing cystic lesion with multiple septae involving the left submandibular and parapharyngeal spaces up to the base of tongue.

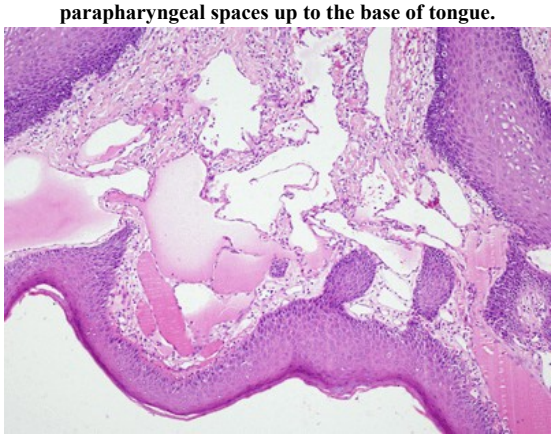

Figure 3: Photomicrograph of the localized lesion showing dilated thin walled lymphatic channel filled with eosinophilic fluid (H&E, x 200)

Discussion:

Lymphangioma was first described by Virchow in 1854. Almost three fourths of these lymphangiomas are localized in the head and neck region and about 80% of these cases are children less than 2 years old.[5] They are hamartomatous, congenital malformations of the lymphatic system occurring as a result of sequestration of lymphatic tissue that has retained its potential for growth and do not communicate with other lymphatic tissue.[6] Embryologically it is derived from five primitive buds developing from the venous system which include paired jugular sacs, paired posterior sacs and a single retroperitoneal sac.[6] Lymphangiomas are relatively rare in the oral cavity, occur mostly on the dorsal surface and lateral border of the tongue and rarely arise on the palate, gingiva, buccal mucosa and lips. These tumors rarely regress and they keep on growing slowly with secondary infection and trauma. A rapid increase in size is seen in infection or bleeding, when they can cause speech difficulty, respiratory distress, dysphagia and sleep apnea.[7] In the present case, the swelling was noticed from one year of age and there was no evidence of any dysphagia and dyspnea. Lymphangiomas of the tongue usually present with macroglossia, speech disturbances, poor oral hygiene, and bleeding from tongue associated with oral trauma.[3,4,8] In our case, however, the tongue was normal in size with no clinical symptoms, noticed by parents of the patient.

Complete surgical excision is the treatment of choice. In case of inaccessible sites like base tongue, floor of mouth, larynx, neurovascular structures of neck, mediastinum and recurrent and residual tumors, intralesional injection of sclerosing agents have been tried before surgery. Commonly used agents include 25% dextrose, hypertonic saline, bleomycin, aethoxysklerol, OK-432 (picibanil).[9] These not only change the consistency of the tumour but also cause a marked reduction in size and improvement in cosmetic appearance. In recent year's Carbon dioxide and Neodymium Yitrium Aluminum Garnet (Nd-YAG) laser photocoagulation surgery has become popular. Earlier steroids, electrocoagulation, cryotherapy or radiation therapy has been used with variable results but most effective treatment is surgery.[10] Multimodality therapy should be available because neither surgery nor sclerotherapy in isolation can provide optimal treatment for every lymphatic malformation. A proportion of these children may require multiple treatments.

On retrospective analysis of CT scan findings done at the time of presentation of the neck swelling, there was an ill defined fat plane with the posterior border of the tongue, suggesting the presence of the lesion near the tongue base which did not subside with sclerotherapy and reappeared as a tongue lesion after a span of 2 years.

We therefore wish to highlight that a cystic hygroma presenting in the neck, may have extensions into the tongue base, that remain unnoticed and untreated and may present a few years later as a separate lesion on the tongue, even after the neck swelling has been treated either medically or surgically.

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