Lymphangioma of Tongue a Rare Entity: A Case Report

Sagar Gupta¹, Kaushal Vegad²

¹Assistant Professor, Department of Surgery, SBKSMI & RC, Vadodara, Gujarat, India; ²Resident, Department of Surgery, SBKSMI & RC, Vadodara, Gujarat, India

Abstract

Lymphangioma is relatively rare congenital malformations of lymphatic vessels that are filled with a clear protein-rich fluid with a few lymph cells. They result due to abnormal development of the lymphatic system with an obstruction to lymph drainage from the affected area. This is an interesting case of lymphangioma of the tongue in a 2-year-old female child. Oral lymphangioma is uncommon lesions that tend to involve the dorsal surface of the tongue. Conservative surgical excision is preferred in superficial, localized lesions and hence an insight of this condition of prime importance in its diagnosis and management. Oral lymphangioma is relatively uncommon lesions occurring at the dorsal region of the tongue. Superficial and localized lesions should be treated by conservative surgical excision with low relapse rates.

Keywords: Congenital, Lymph, Vascular malformations

INTRODUCTION

Lymphangioma was first described by Virchow as a rare, benign, congenital disease of unknown etiology that originates from lymph vessels.¹ It is an embryonic tumor also called as malformation or hamartoma. Incidence of lymphangioma is about 6% of all the benign soft tissue tumors, in the population younger than 20 years.² It arises due to a congenital anomaly of lymphatic system resulting in clusters of small lymphatic vesicles in wrong area, that fails to anastomose with the main channel and results in blockage of the lymph drainage resulting in inflation of the small vesicles.³ Few studies consider it as a true neoplasm; derived from the transformed lymphatic endothelial cells or/and stromal cells, instead of being a congenital malformation.⁴

Most common site of lymphangioma is head, neck and oral cavity.³ Tongue lymphangioma shows multiple blisters like nodules or a pebbly surface resembling a group of translucent vesicles on the dorsal surface of tongue.⁴ Lymphangiomas are a very common cause of macroglossia in infancy. They are often associated with a group of congenital malformations like Turner’s and Noonan’s syndrome or could present in association as trisomy, cardiac anomalies, hydrops fetalis, Fetal alcohol syndrome, and familial pterygium coli. In children, Cystic hygroma is characterized by a sudden growth predisposing to an infection or hemorrhage.⁵

Traditionally, lymphangioma are classified into four categories based on their histologic appearance as lymphangioma simplex (capillary lymphangioma, lymphangioma circumscriptum), which is composed of small, thin-walled, endothelium that is lined by capillary-sized lymphatic vessels; while cavernous lymphangioma comprises of dilated lymphatic vessels with surrounding adventitia; cystic lymphangioma (cystic hygroma), that comprises of huge, macroscopic lymphatic spaces with surrounding fibro-vascular tissues and smooth muscle; Benign lymphangioendothelioma (acquired progressive lymphangioma) that is characterized by dissecting lymphatics through the dense collagen.⁶

However, histologic categories do not correlate with their clinical presentation or response to any therapy. Hence, lymphangioma can present as either
Lymphangioma can be defined as congenital malformations in the lymphatic system. There are two different theories for the origin of the lymphatic system in humans. The first theory states that the 6th week of embryogenesis, the lymphatic system develops from five primitive sacs that are developed from the venous system. In the head and neck region, endothelial out-pouching from the jugular sac spreads out to form the peripheral lymphatics, which was proposed by Sabin. While, Godart proposed mesenchymal clefts develop into lymphatic system.

During 6th week of embryogenesis, lymphangioma arises from incomplete development and obstruction of the lymphatic system. The failure of primordial lymphatic sacs to connect with the venous system leads to incomplete canalization and obstruction to the lymph flow and development of the cyst. The second theory states that the deposition of lymph tissues in the wrong area during embryogenesis leads to failure of canalization with the lymphatic system.

In the first decade of life, in the oral cavity, this lesion is common and involves the dorsal and lateral area of the tongue. Lymphangioma rarely arises on palate, gingival, buccal mucosa and lips. The most common site for intraoral lymphangiomas leading to macroglossia is the anterior two-thirds of the dorsal surface of the tongue. These patients present with speech disturbances, poor oral hygiene, speech disturbances and bleeding from the tongue associated with oral trauma.

In our case, the patient had poor oral hygiene and presented as spontaneous regression of lymphangioma that is rare and presents as slow, progressive enlargement in contrast to haemangioma that usually regresses. Progressive swelling of the tongue occurs due to gradual dilation of the lymphatic channels with each episode of infection. The objectives of treatment of lymphangiomatous macroglossia are restoration of tongue size for articulation, preservation of taste, correction of dental and mandibular deformities. Management for lymphangioma includes surgical excision, radiation therapy, cryo-therapy, electro-cautery, sclerosing with sclerosing agent, laser surgery with Nd-YAG, CO₂ and radiofrequency tissue ablation technique. The usual treatment of lymphangioma is surgical excision.

Simple excision was performed in our patient and on follow-up after 1 year, patient was asymptomatic and there was no recurrence.

**CONCLUSION**

Oral lymphangiomas are relatively uncommon lesions occurring at the dorsal region of the tongue. Superficial and localized lesions should be treated by conservative surgical excision with low relapse rates. Thus for the correct diagnosis and proper management, the knowledge of this condition is of prime importance.
REFERENCES