Oral lymphangioma of the buccal mucosa a rare case report

Saligrama Seema Yoganna, Rame Gowda Rajendra Prasad¹, B. Sekar²

ABSTRACT

The lymphangioma are benign hamartomatous tumors of lymphatic vessels that arises from the sequestration of lymphatic that fails to communicate with the lymphatic system. Most common intra oral site being the anterior two-thirds of tongue, usually superficial in location and demonstrates a pebbly surface that resembles a cluster of translucent vesicles, they are typically soft and fluctuant masses. Secondary hemorrhage into the lymphatic spaces may cause some of these vesicles to become purple. They have been known to grow to large size causing difficulties in mastication and speech. A variant of lymphangioma is cystic hygroma grows as lymphatic anomaly found in the neck commonly present with significant airway obstruction. We present a rare case of lymphangioma affecting the buccal mucosa of a 14-year-old male.

KEY WORDS: Buccal mucosa, hamartoma, lymphangioma

Lymphangiom was benign hamartomatous malformations that arise from sequestration of lymphatics tissue that fails to communicate normally with the lymphatic system. These remnants may have some capacity to proliferate, but more importantly they accumulate vast amount of fluid which accounts for their cystic appearance, the entity first described by Virchow in 1854.[1]

Lymphatic system is a diffuse network of endothelial channels, makes its appearance during the 6th week of embryonic development. Vasculoendothelial growth factor receptor-3 a tyrosine kinase receptor is important for the lymphatic development. Lymphatics channels are extensive unidirectional systems with the blunt endings, which retrieves excessive fluid from interstitium and transport it to regional lymph node and return it to venous system by the way of thoracic duct, they also helps in absorption of protein and lipid from liver and small intestine, respectively.[2]

Watson and McCarthy classified them into five types: (1) Simple lymphangioma, (2) cavernous lymphangioma, (3) cellular or hypertrophic lymphangioma, (4) diffuse systemic lymphangioma and (5) Cystic lymphangioma or hygroma.

Oral manifestations

Three fourths of lymphangiomas occurs in head and neck region. Majority of the cases of lymphangiomas are present at birth, 95% of the tumors arise before the age of 10 years (Watson and McCarthy)[3]. 88% of the lymphangioma appears by the end of the 2nd year of life (Hill and Briggs).

The most common intra oral site being tongue, while it also seen in the palate, buccal mucosa, gingival, and lips. Anterior two-thirds is most commonly affected with enlargement causing “macroglossia”[4]. Lip involvement and its attendant deformity are referred as microcheilia. Lymphangioma of the alveolar ridge in neonates has been reported by Levin et al.[5]

A cystic hygroma is a congenital multiloculated lymphatic lesion that can arise anywhere, but is classically found in the left posterior triangle of the neck. This is the most common form of lymphangioma. It contains large cyst like cavities containing watery fluid. Microscopically cystic hygroma consists of multiple locules filled with lymph. In the depth, the locules are quite big, but they decrease in size toward the surface.

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Syndromes associated with oral lymphangiomas are: (1) Turner’s syndrome, (2) Noonan’s syndrome, (3) trisomies, (4) fetal hydrops, and (5) fetal alcoholic syndrome.

**Case Report**

A 14-year-old boy reported to the department of oral maxillofacial pathology, (Vinayaka Mission Dental College, Salem) with the chief complaint of swelling on left cheek region for the past 5 years. Previous history revealed that patient had swelling since birth and it was operated at age of 3 years through extra oral approach and a scar was present in the left cheek region [Figure 1]. After which the patient was apparently normal, then at the age of 9 years again the swelling appeared in the same region, which was growing slowly and attained the present size of 4 × 3 cm [Figure 2], and was not associated with pain. Patient complained of difficulty in mastication and speech, unaesthetic appearance. No history of bleeding and pus discharge associated with the swelling, no history of swelling in other parts of the body.

**Intraoral examination**

Intraoral examination showed a diffuse swelling measuring around 4 cm × 3 cm in size in relation to occlusal line of left buccal mucosa extending anteriorly from the corner of the lip to the retromolar region posteriorly, surface of the swelling had pebble like appearance. On palpation swelling was soft and nontender and nonpulsatile and had no dilated veins. Extraoral examination: A diffused extra oral swelling present in the left cheek region of about 5 × 6 cm in size; anteroposteriorly extending - from the left corner of the mouth to the anterior preauricular region, superoinferiorly- from the left ala of the nose to lower border of the body of mandible, no dilated veins over the swelling no pulsation felt [Figure 1] skin is pinchable with no sinus and fistula formation. There is scar over the swelling due to previous surgery and measuring about 3 × 0.5 cm [Figure 3].

A provisional diagnosis of intra oral lymphangioma of the buccal mucosa was established. A differential diagnosis of arteriovenous malformation. Routine blood investigations were found to be normal. An incisional biopsy was performed under local anesthesia and specimen was subjected to histopathological examination. Microscopically, it showed numerous dilated lymphatic vessels lined by thin endothelial cells (H and E, ×10).
cells, characteristically located just beneath the epithelial surface and spaces within the lymphatic vessels contain proteinaceous fluid [Figures 4 and 5]. This superficial location results in the translucent, vesicle-like clinical appearance [Figure 2]. The entire lesion was excised, no recurrence has been observed and the patient is under follow-up [Figure 6].

**Discussion**

Lymphangiomas are hamartomatous, congenital malformations of the lymphatic system. There are two theories for the origin of the lymphatic system in humans. One theory is that the lymphatic system develops during the 6th week of embryogenesis from five primitive sacs that arise from the venous system. In the early 1900s, Sabin proposed that endothelial out pouchings from the jugular sac spread to form the peripheral lymphatic system. Godart, on the other hand, proposed that the lymphatic system developed from mesenchymal clefts in the venous plexus and spread centripetally toward the jugular sac. [1]

The second theory is that lymph tissues are deposited in the wrong area during embryogenesis and subsequently fail to join the normal lymph system. [2] They are sometimes termed lymphangiectasia because they are cystic dilations of malformed lymphatic channels and therefore will collect lymph. [3]

The most common presentation is that of painless soft mass that gradually enlarges and then remain static over a long period, occasionally enlargements and shrinkages occur, a residual mass remains. Due to their soft quality the clinical differential diagnosis include lipomas, salivary retention phenomena and hemangiomas. [4]

Many lymphangiomas have some blood in their lymphatic channels, they are most often confused with hemangiomas and are termed as hemangiolymphangioma, and these basically represent lymphangiomas with communication to normal blood vessels. [5]

Levin et al. have described lymphangiomas of the alveolar ridges of neonates in 3.7% of 1470 black neonates, these lymphangiomas forms blue domed, fluid filled swellings on the posterior crest of the alveolar ridge and posterior lingual surface of the mandibular ridge. [6]

The superficial multicystic type is the most common type, it develop slowly as soft enlargements with pebbly surface. The deep cavernous type tends to expand outward. The cystic hygroma type tends to expand outward, creating a generalized enlargement of the area. This type is more often reported to undergo episodes of expansion and shrinkage than the other types. Histopathologically lymphangiomas with secondary hemorrhage are sometimes confused with cavernous hemangiomas, histologic features that favors the diagnosis of lymphangioma over hemangioma are more irregular lumens lined by spaced nuclei and proteinaceous material filling the lumen. Lymphatic endothelium express factor VIII-associated antigen and CD31 and hence immunohistochemical procedures are not reliable means of distinguishing hemangiomas from lymphangiomas. [7]

Diagnostic workup includes biopsy for histopathologic examinations, which will confirm the diagnosis, ultrasonography will detect the cystic nature and fluid component of a lymphangioma and angiography will rule out vascular lesions. [8]

**Conclusion**

Although the lymphangioma is a benign lesion, it may cause significant morbidity due to its large size, critical location or proclivity to become secondarily infected. The management of lymphangiomas includes early histopathologic diagnosis followed by surgical management and use of sclerosing agents. The use of radiotherapy has declined because of the malignant transformation of previously irradiated lymphangiomas. [9]
References

1. Virchow. J. Ueber Makroglossic and Pathologische Neubildung Vonquergestreiften Muskelfasern, Virchows Arch. F. Path. Anot, 7:127;1854.

2. Enzinger FW, Weiss SW. Soft Tissue Tumors. 4th ed. St. Louis: Mosby; 2001.

3. Livesey JR, Soames JV. View From Beneath. Pathology In Focus Cystic Lymphangioma In The Adult Parotid. J Laryngol Otol. 1992; 106:566-8.

4. Shafer’s WG, Hine MK, Levy BM. Text book of oral pathology 5th ed, Delhi: Elsevier India Pvt Ltd; 2006:207-8.

5. Levin LS, Jorgenson RJ, Jarvey BA. Lymphangioma Of The Alveolar Ridges In Neonates. Pediatrics 1976;58:881-84.

6. Neville BW, Damz DD, Allen CM, Bouquot JE. Soft Tissue Tumours (2004) In Oral and Maxillo Facial Pathology 2nd ed By: Neville BW, Philadelphia: WB Saunders, 475-7.

7. Robert E. Marx, Diane Stern. Textbook of Oral and Maxillofacial Pathology: A Rationale For Diagnosis and Treatment 2003. Quintessence Publishing Co, Inc, 1st ed. (November 1, 2002), 435-37.

8. Fonkalsrud EW. Surgical management of congenital malformation of the lymphatic system. Am J Surg 1974;128:152-9.

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