



When not all swellings are mucoceles: A case of lip lymphangioma in the maxillofacial region

Abstract

Lymphangiomas are tumour-like benign, developmental hamartomatous malformations of the lymphatic system which is distinguished by abnormal proliferation of lymphatic vessels. It was first reported by Virchow in 1854. In this article we report a unique case of lymphangioma of the lower lip of a 11-year-old patient who is under orthodontic treatment. A detailed history and clinical evaluation, and histopathology confirmed the diagnosis of lymphangioma.

Keywords: lymphatic malformation, lip, mucocele, oral lymphangiomas

**Geetha Sridharan*,
Aditya Agrawal, Pradeep
Devadoss, Srinivasa Prasad
Tangutur, Nissi Evelyn,
Kavitha Bottu**

*Department of Oral and Maxillofacial
surgery, Meenakshi Ammal Dental College
and Hospital, Chennai, Tamil Nadu, India*

**Author for correspondence:
drgeethaomfs@gmail.com*

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Introduction

Vascular malformations were first described by Redenbacher in the year 1828. Few refer lymphangiomas as hamartomas and other lesions as congenital malformations. Intemperate secretion from the cell lining causes obstruction of lymphatics leading to slow-growing, asymptomatic, pain-free cystic mass causing acute facial abnormality. These conditions are more often predominant during the time of birth or at infancy (50%) and other vascular tumours in children constitute about (25%), which has more female predilection compared to male [1,2].

These are commonly evident in head and neck region about (50%-70%) but they are sparse entity in oral cavity region [3,4]. Frequent sites of occurrence in the oral region are the tongue (dorsum and lateral border), palate, cheeks, floor of the mouth, gingiva and the lips. Other differential hamartomatous lesions are haemangiomas, odontomas, and cherubism etc [3,5].

Case Report

A 11-year, female patient accompanied by parents, reported to our institution with a chief complaint of small swelling on lower lip for 1 year. Upon history of present illness, she was apparently alright a year back when they noticed a small pea size soft and compressible swelling on the lower lip which regresses upon compression with no associated pain. Small pea size swelling

resolved on its own 6 months back. Recently 2 months back they again noticed same pea size swelling in the same region which is not associated with pain and not regressing upon compression. Patient was under orthodontic treatment since a year. On local clinical examination, small pea size vermilion colour nodule, non-tender and non-compressible is present on lower lip. Provisionally diagnosed as mucocele of lower lip of size approximately 3 mm × 3 mm (FIGURE 1). Treatment done was excision under local anaesthesia and histopathological examination done upon which final diagnosis came up as lymphangioma of lower lip (FIGURE 2).

Upon histopathological examination of the excised specimen, H&E-stained soft tissue section showed dilated lymphatic channels submucosally, lined by widely spaced flattened endothelial cell nuclei and lumen with foci of granulation tissue formation with inflammatory component. The adjacent area showed a lobule of mucous salivary acini (FIGURE 3.A-3.C). Patient follow up done up to 3 months which showed remarkable healing and no recurrence (FIGURE 4).

Discussion

Lymphangiomas emerge from the segregate of primordial lymphatic cell channels which insistently store lymph but stall to join the larger vessels, leading to lymphatic blockage, thus showcasing a cystic appearance which exhibits a miscellaneous pattern in defining lymphangiomas. In paediatric population



FIGURE 1. Pre-Operative image of the lesion measuring 3 mm × 3 mm in the lower lip region.



FIGURE 2. Specimen excised with underlying muscle layer.

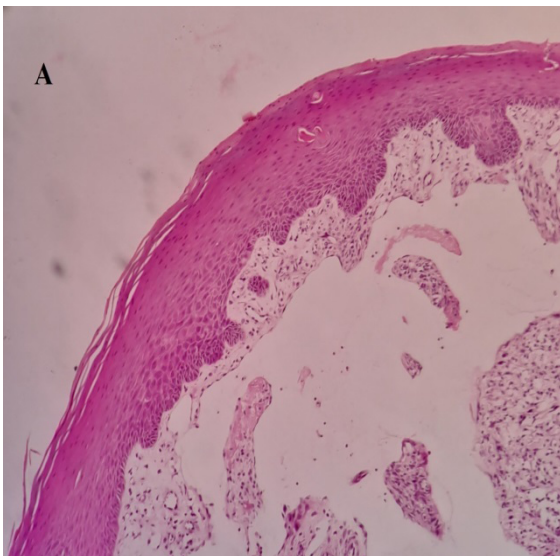


FIGURE 3A : 4X magnification, lymphatic channels beneath the surface epithelium with a foci of granulation tissue.

lymphangiomas prevails with an incidence of about 1 in 16000 live births and are ascribed to about 6-11% of benign maxillofacial tumours [6]. There are two major entities: Acquired lymphangiomas and Congenital lymphangiomas, whereas the other factors like prolonged history of trauma, blockage of lymph vessels due to infection, inflammation, radiotherapy, or surgery are inclined towards acquired lymphangiomas and lymphangiomas associated with haemangiomas,

definite chromosomal aberrations like Noonan syndrome, Turner's syndrome, Down syndrome, Foetal alcohol syndrome, Cardiac anomalies and foetal hydrops are the predisposing factors for congenital lymphangioma [7]. Various rationales have been proposed for the pathogenesis of lymphangiomas. Etiopathogenesis of lymphangioma is depicted in **TABLE 1**.

Histopathologically, lymphangiomas are

FIGURE 3B: 10X magnification, lumen beneath the surface epithelium lined by Endothelium.

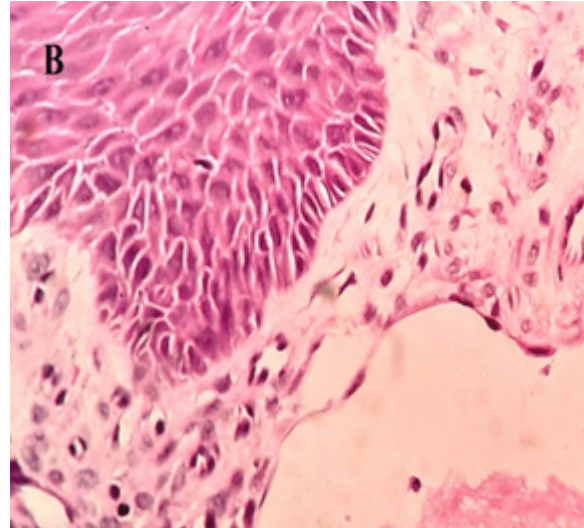


FIGURE 3C: 40X magnification, widely spaced flattened endothelial cell nuclei lining the epithelium.

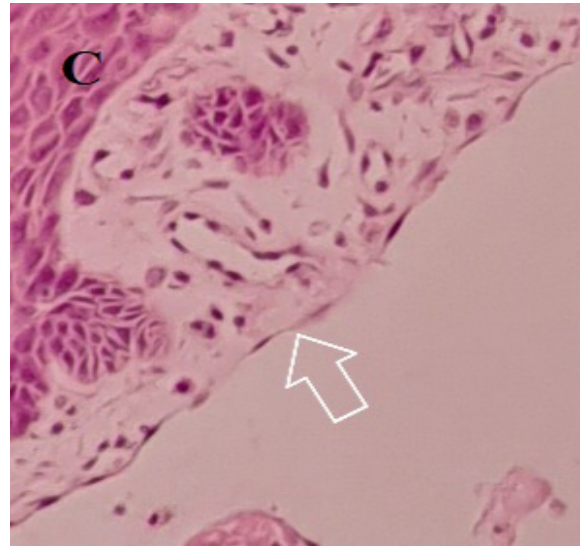


FIGURE 4. Post-Operative.



characterized by the presence of dilated lymphatic vessels and cysts, lined by flattened endothelial cells, and separated by fibrous septae containing lymphoid aggregates and lymphoid follicles. The cysts are usually filled with a clear lymphatic fluid, which is rich in lymphocytes, macrophages, and plasma cells [5,6]. In the case of lymphangioma of the lip, the histopathological examination would reveal similar findings. The excised specimen would show dilated lymphatic channels submucosally, lined by widely spaced flattened endothelial cell nuclei and lumen with foci of granulation tissue formation with inflammatory component. The adjacent area may show a lobule of mucous salivary acini, but these would be separated from the lymphatic cysts by fibrous septae.

It is important to differentiate lymphangiomas from other cystic lesions, such as mucoceles, as the treatment approach differs [8,9]. Lymphangiomas lack a universally standardized classification; typically, they are categorized into superficial and deep lymphangiomas based on the size and depth of the abnormal lymphatic channels (TABLE 2). Even tumours that appear well-defined clinically do not exhibit encapsulation. In deeper lesions, vessels are observed scattered amidst adipocytes and striated muscle bundles. Lymphangiomas can be subclassified into four types (TABLE 3).

These categories are relatively arbitrary, and numerous lesions exhibit a combination of characteristics from multiple categories [10].

Lesions in the oral cavity may include haemangioma, lingual thyroid, thyroglossal duct cyst, granular cell tumour and cheilitis. Granulomatosis, mucocele/ranula, and cellulitis

are all possibilities for differential diagnosis [6,11]. Mucoceles, on the other hand, are common benign lesions that arise from minor salivary glands and are filled with mucin. They typically present as soft, compressible, painless swellings that fluctuate in size and can appear bluish or translucent. Mucoceles are often associated with trauma or injury to the affected area [12,13].

Lymphangiomas are usually diagnosed according to their clinical presentation and correlated with the histopathological report and diagnostic radiographic study which expedite by affirming the diagnosis. The shape, size, site and margins of the lesion are assessed with Ultrasonography (USG) which is a requisite to check whether the lesion is multicystic or unicystic with internal septae. Ultrasound with colour-flow Doppler aids in identifying the component of the lesion, whereas angiography helps not only to avert the vascular lesions but also to differentiate the entities [14,15]. Nevertheless, Computed tomography and magnetic resonance imaging is indisputable to ultrasonography in identifying the adjacent structures and a delineation of the extent of the lesion [14].

Restoring the functions and preventing any potential problems should be the main goals of the therapeutic techniques. Though there are plethora of treatment options for management of lymphangiomas like laser therapy (carbon-dioxide laser and Nd:YAG, cryotherapy, sclerotherapy (25% Dextrose, hypertonic saline, bleomycin, picibanil (OK-432) [15, 16], Sodium Tetradecyl Sulphate (STS), ethanol, and corticosteroid [15], electrocautery, intralesional steroids, interferon alpha, therapeutic embolization and ligation, Flashlamp Pulsed Dye Laser (FPDL),

TABLE 1. Rationales for pathogenesis of the lymphangioma

Rationale I	Congenital Lymphangiomas result from sequestration of primordial lymphatics
Rationale II	The failure of the lymphatic vessels to drain into the veins causes lymph accumulation and leads to the formation of large cyst-like dilated lymphatics
Rationale III	Lymphangiomas may develop through aberrant budding of the lymphatic vessels, thus establishing new anomalous divisions.
Rationale IV	Long-standing inflammation may evoke lesion development by triggering the proliferation of lymphatic channels
Rationale V	Increased expression of vascular endothelial growth factor-C (VEGF-C) and vascular endothelial growth factor R3

TABLE 2. Clinical classification of lymphangioma

Type I	Superficial Multicystic type
Type II	Deep cavernous type
Type III	Cystic Hygroma

TABLE 3. Histopathological classification of Lymphangioma

Lymphangioma Simplex	Composed of small, thin-walled lymphatics
Cavernous lymphangioma	Comprised of dilated lymphatic vessels with surrounding adventitia
Cystic lymphangioma	Consisting of huge macroscopic lymphatic spaces with surrounding fibrovascular tissues and smooth muscle
Benign lymphangioendothelioma	Lymphatic channels appear to be dissecting through dense collagenic bundles

radiotherapy but the substantiated treatment is surgical excision of the lesion [16-18].

One such advancement is the use of laser therapy in the treatment of lymphangiomas. Lasers have been shown to be effective in the treatment of vascular malformations, including lymphangiomas, by selectively destroying the abnormal blood vessels while sparing the surrounding normal tissue. Several studies have reported successful outcomes in the treatment of lip lymphangiomas using various types of lasers, including carbon dioxide (CO₂) laser, neodymium-doped yttrium aluminium garnet (Nd: YAG) laser, and Pulsed Dye Laser (PDL) [18,19].

Based on our case, it is crucial to emphasize that even for lesions displaying typical clinical characteristics, it is essential to conduct additional clinical investigations and complementary examinations, such as biopsy and histopathological analysis, to confirm the diagnosis accurately and determine the appropriate treatment approach.

Conclusion

In conclusion, lymphangiomas should be considered in the differential diagnosis of

maxillofacial lesions presenting with clinical features similar to mucoceles. A thorough clinical and histopathological evaluation is essential for accurate diagnosis and appropriate management. Dental professionals should be aware of the clinical and histopathological features of lymphangiomas to avoid misdiagnosis and ensure optimal patient care.

Ethical Approval

Not applicable

Conflict of Interest Statement

The authors declare that they have no conflict of interest.

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Consent to Publish

Written consent to publish was obtained for publication of the patient's identifiable information.

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