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Case Report
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A Single-centre Case Series of Oral Lymphangiomas

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ABSTRACT

Introduction: lymphangiomas are developmental and congenital malformations that represent gradual distention of the lymph vessels, which is a benign and rare occurrence in the oral cavity. It is a dilated lymphatic channel containing lymph material that does not connect with other channels.

The various imaging modalities are Orthopantomography (OPG), Ultrasonography (USG), Computed tomography (CT), Magnetic resonance imaging (MRI), and Color Doppler, which are helpful in explaining the spread of the lesion. Histopathologically, lymphangiomas appear as numerous small or larger lymphatic vessels lined by plump endothelial cells with empty or lymphatic fluid. Surgery along with a sclerosing agent is the most effective treatment, which shows a good prognosis depending on the size, type, and anatomical site of the lesion.

We present a case series of oral lymphangiomas of the cheek, floor of the mouth, and tongue with varying clinical and imaging presentations.

Keywords: Hamartoma; Lymphangioma; Benign neoplasm; Vascular malformations; Endothelial cells.

Introduction

Lymphangiomas are benign hamartomatous tumors that emerge from lymphatic tissue sequestrations that do not communicate with other lymphatics. Lymphatic channels are filled with a clear, protein-rich lymph fluid that contains only a few lymphocytes[1]. The majority of lymphangiomas appear at birth (60–70% of cases) or develop in the first two years of life (90% of cases), with a 2:1 to 3:1 male to female distribution[1,2,3,4]. The most common site for lymphangioma is the tongue, followed by the lips, buccal mucosa, soft palate, and floor of the mouth. This article provides a case series of four case studies of oral lymphangiomas with varying clinical, imaging, and histopathologic presentations. [Table-1]

Table 1. Clinical and radiologic presentation of our case series

Case Nº	Age	Sex	Site	Symptom	Radiographic presentation	Histopathology feature
1	14 years	Male	Right side of face	Swelling	Focal nodular thickening of the soft tissue at right cheek anterior to the right masseter	Simple type
2	28 years	Male	Left cheek	Swelling	Cyst with clear fluid at left cheek with irregularly septate and situated below the subcutaneous fat	Simple type
3	40 years	Female	Tongue	Swelling Tapioca-pudding appearance	Extensive thin-walled cystic mass noted in the submental region	Cystic type
4	28 years	Female	right lateral dorsum of the tongue	Swelling	-Not used-	Cystic type
5	40-years	female	lower jaw	Swelling	extensive thin-walled cystic mass lesions at cervicomandibular area.	Cystic type

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Case Reports

Case 1:

A 14-year-old male reported a solitary, diffuse swelling on the right side of the face for one month, which was roughly ovoid and measured about 3x3 cm in size. The swelling was non-erythematous, had a smooth surface, and was not tender on palpation [Figure-1]. Ultrasonography (USG) revealed a focal nodular thickening of the soft tissue at the right cheek anterior to the right masseter [Figure-2]. The fine-needle aspiration cytology smear revealed yellowish fluid with lymphocytes and proteinaceous material, indicating lymphangioma.

Case 2:

A 28-year-old man reported diffuse swelling in his left cheek for 8 months. The swelling was 3x2.5 cm in size, ovoid in shape, non-tender with a soft to firm consistency, and smooth and non-erythematous on the surface. [Figure-3] A 2.3 x 1.4 cm cyst with clear fluid at the left cheek with an irregular septum and located below the subcutaneous fat was revealed by USG, indicating a lymphangioma. [Figure-4]

Case 3:

A 40-year-old female reported a diffuse swelling in the submandibular and submental region for 2 months that was insidious in onset and had a gradual and constant increase in size, measuring about 8 x 4 cm. The swelling was non-tender, soft in consistency, and compressible. Intraorally, multiple, pinpoint, pebbled eruptions with a frog spawn or tapioca pudding appearance were seen over the dorsal surface and lateral border of the tongue. [Figure-5] An ultrasound revealed an extensive, thin-walled cystic mass in the submental region, indicating a lymphangiomatous lesion. [Figure-6]

Case 4:

A 28-year-old female presented with a nodular mass on the right lateral dorsum of the tongue for 5 months with a papillary appearance, soft consistency, and bleeding noticed. [Figures-7] Endothelium-lined lymphatic channels filled with lymph in connective tissue were diagnosed as lymphangiomas on histopathologic examination. [Figure-8]

Case 5:

A 40-year-old female patient presents with swelling beneath the lower jaw of 7 years' duration and pain that aggravates with chewing. The swelling was non-tender, non-lobulated, soft to firm in consistency, and there was no pus, blood, or watery discharge. [Figure-9] On ultrasound examination, extensive thin-walled cystic mass lesions were discovered in the cervico-mandibular region. [Figure-10] A lymphangioma was confirmed by FNAC and biopsy. [Figure-11]

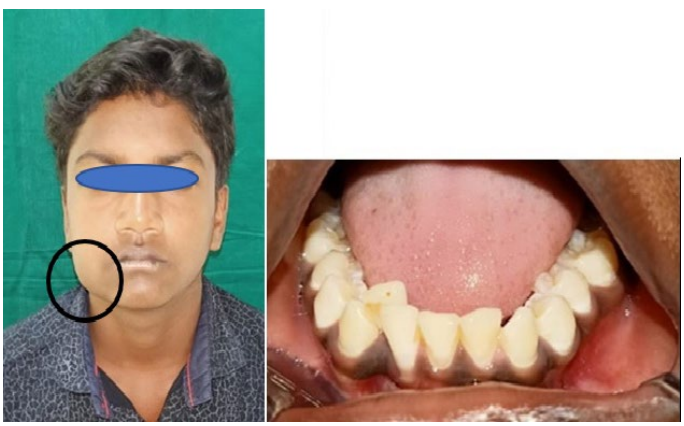

Figure 1. Clinical photography of swelling in the right side of the face.

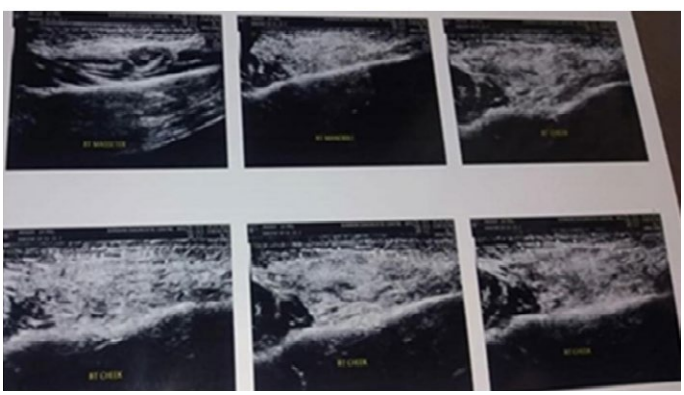

Figure 2. Ultrasonography showing presence of focal nodular thickening at right cheek of the face.

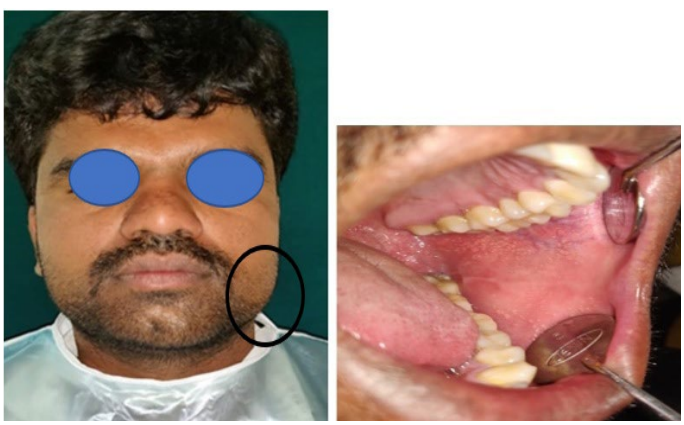

Figure 3. Clinical extra-oral and intra-oral photography of swelling in the left side of the face.

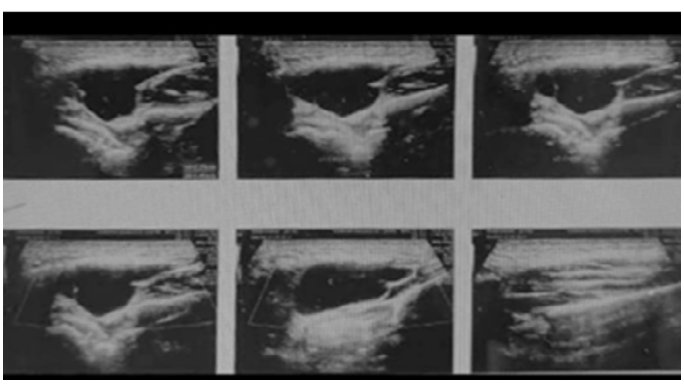

Figure 4. Ultrasonography showing presence of cyst with clear fluid at left cheek with irregularly septate and situated below the subcutaneous fat.

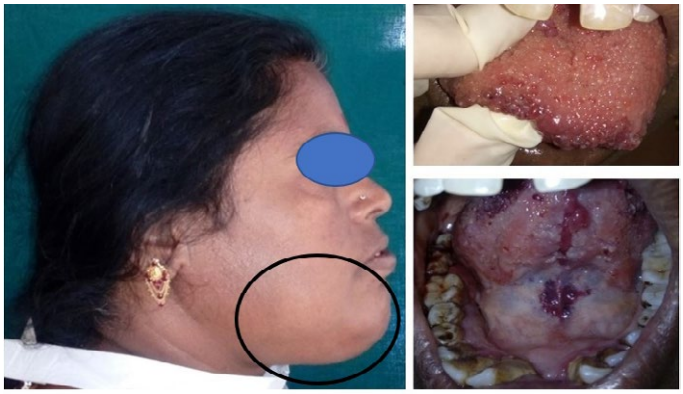

Figure 5. Clinical Extra-oral and Intra-oral photography of swelling on the dorsum of tongue, ventral surface of the tongue and floor of the mouth.

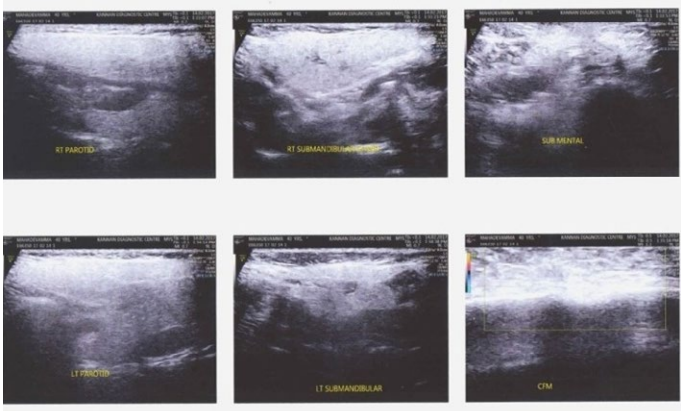

Figure 6. Ultrasonography showing presence of extensive thin-walled cystic mass noted in the submental region.

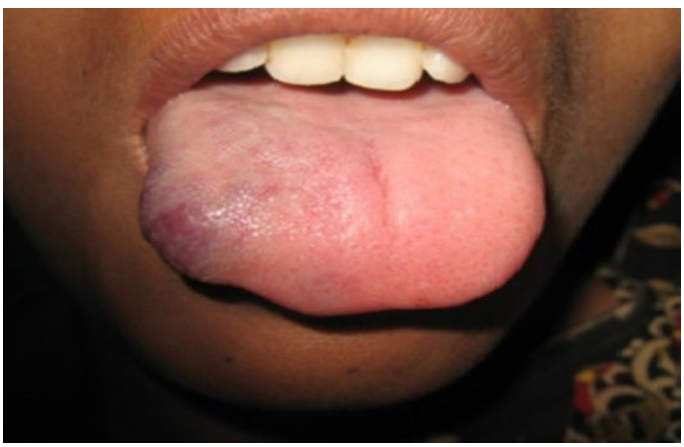

Figure 7. Intra-oral photography of swelling on the lateral surface of the tongue.

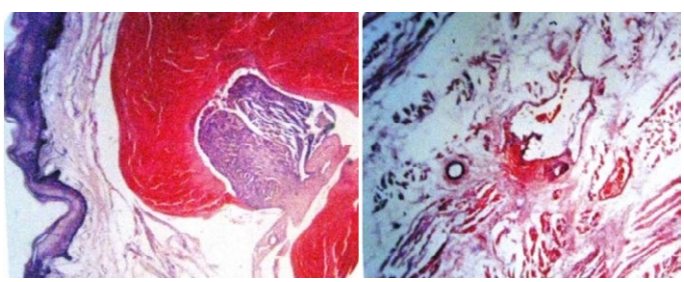

Figure 8. Photography of H&E tissue section showing endothelial lined lymphatic channels with eosinophilic lymphatic fluid (10X magnification).

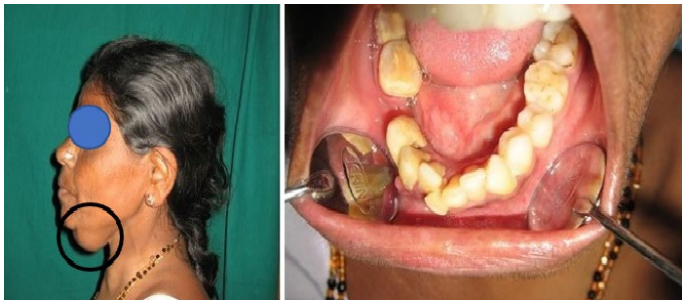

Figure 9. Clinical Extra-oral and Intra-oral photography of swelling on the floor of the mouth .

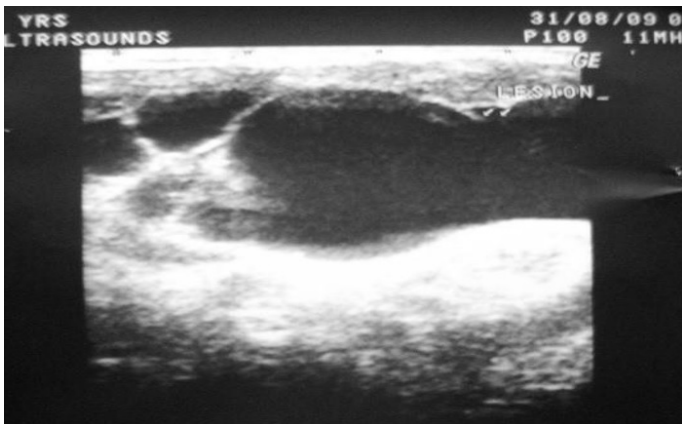

Figure 10. Ultrasonography showing extensive thin-walled cystic mass lesions at cervicomandibular area.

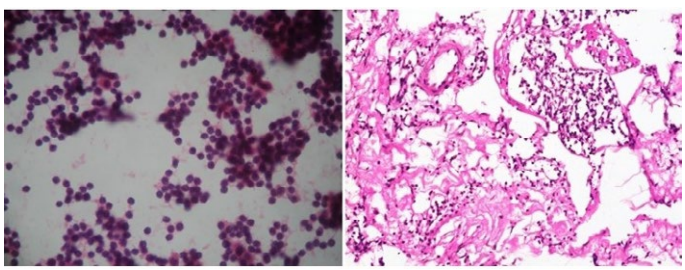

Figure 11. Photography of FNAC and H&E tissue section showing endothelial lined lymphatic channels with eosinophilic lymphatic fluid (10X magnification).

Therapeutic intervention:

The lesion in Cases 2 and 4 remained asymptomatic, with no signs of progression or change, and was kept under observation. The patient had undergone intralesional sclerotherapy before any surgical procedure due to the proximity of the facial nerve, and a cystic fluid effusion was drained under local anaesthesia in Case 1. Hemi-mandibulectomy with surgical removal of a lesion was performed in Case 5. In Case 3, the patient was not willing to undergo surgical intervention.

Follow-up and outcomes:

All cases showed uneventful healing, with 90% shrinkage and no signs of recurrence of the lesion. The patient was followed every 3 months for a year and remained asymptomatic except for case-3. [Table-2]

Table 2. Timeline of event.

History given by patient	Case-1	Case-2	Case-3	Case-4	Case-5
Patient-reported to our OPD	2 years back	5 years back	4 years back	2 years back	10 years back
Clinical features at the time of reporting	Swelling in the right side of the face for 1 month	swelling in the left cheek for 8 months	A diffuse swelling in the submandibular and submental region	swelling on the right lateral dorsum of the tongue for 5-months	Swelling in the submandibular and submental region
Diagnosis (Based on clinical, radiological, and histopathological examination)	A simple type of lymphangioma	A simple type of lymphangioma	Cystic type of lymphangioma	Cystic type of lymphangioma	Cystic type of lymphangioma
Therapeutic intervention	The patient underwent to get intralesional sclerotherapy before any surgical procedure due to the proximity of peripheral branches of the left facial nerve and cystic fluid effusion was drained from different places under local anesthesia	The treatment was not initiated and kept under observation.	The patient was not willing to undergo surgical intervention	The treatment was not initiated and kept under observation.	Hemi mandibulectomy with surgical excision of tumor
Recall and follow-up: a week	There was uneventful healing with 90% shrinkage and no signs of recurrence of the lesion.	There was no progression or change seen and the lesion remained asymptomatic	A patient has not reported back	Patients who remained asymptomatic	Healing was uneventful and No sign of recurrence or complication
First follow-up: a 3month later	There was uneventful healing with 90% shrinkage	Lesion remained asymptomatic		Lesion remained asymptomatic	No signs of recurrence of the lesion.
Second follow up: 6 months later	No signs of recurrence of the lesion.	Lesion remained asymptomatic		Lesion remained asymptomatic	No signs of recurrence of the lesion.
Third follow up: 9 months later	No signs of recurrence of the lesion.	Lesion remained asymptomatic		Lesion remained asymptomatic	No signs of recurrence of the lesion.
Fourth follow up: 12 months later	No signs of recurrence of the lesion.	Lesion remained asymptomatic		Lesion remained asymptomatic	No signs of recurrence of the lesion.

Discussion

Virchow, in 1854, defined and coined the term "*lymphangioma*," but Redenbacher, in 1828, described it. The etiological factors can include trauma, infections, iatrogenic injuries, and environmental factors[5,6].

Lymphangiomas are soft, slow-growing, painless, translucid plaques that give frog eggs or a "tapioca pudding" appearance due to their small, thin-walled vesicles[7]. If the lesion is a superficial mass, the color of the lesion is red or blue due to capillary rupture into lymphatic space. If the lesion is deeper, the lesion represents mass with a routine color[5]. In this series, we are exhibiting one superficial lesion and three cases of deeper lesions.

The patient may exhibit difficulty chewing and swallowing with impairment of speech, poor oral hygiene, malocclusion like an open bite, and mandibular prognathism[8,9]. Although history and clinical presentation can sometimes suggest the diagnosis of lymphangiomas, it is necessary to differentiate them from other lesions like Haemangioma, Amyloidosis, Lipoma. Neurofibroma, Granular Cell Tumor[9].

OPG, USG, CT, and MRI may be necessary for radiographic examination. USG is a very useful diagnostic aid to evaluate the solid or cystic nature of mixed vascular malformations[7]. In ours, in all three cases where we used ultrasonography to confirm the clinical diagnosis.

Histopathologically, lymphangiomas consist of dilated lymphatic vessels lined by endothelial cells, which contain proteinaceous material or spaces[4,8]. The histologic picture of this lesion is unique in its presentation, which always helps us arrive at a final diagnosis. FNAC of aspirated cystic fluid shows straw-colored fluid with few inflammatory cells. Based on the clinical and histopathologic features, the lesion was classified as shown in Table 3.

Lymphangiomas are treated differently depending on their size, location, and extent of invasion into adjacent tissue. For small, non-expansion lesions, continuous observation is the first step. If the lesion is a large and poorly defined border, surgery is the treatment of choice with the surrounding border of normal tissue. Other procedures include sclerotherapy, radiation therapy, cryotherapy, and laser surgery (CO_2, Nd:YAG, and diode lasers)[1,8,10].

The recurrence rate is around 39% because of incomplete resection of the lesion due to its infiltrative nature into neighbouring structures. Therefore, surgery associated with a sclerosing agent like OK-432 is recommended[5]. The postoperative complications of lymphangiomas include infection, pseudocyst formation, haemorrhage, and recurrent cellulitis[8].

Conclusion

Lymphangiomas are rarely encountered in the oral cavity at the adult stage, and the present case series is shown with various clinical and imaging presentations. The responsibility of the clinician is to examine and diagnose the lesion with the help of other diagnostic aids like ultrasonography, FNAC, Biopsy. The challenge is early detection, which allows for proper treatment initiation and continuous monitoring to prevent recurrence or other complications.

Acknowledgment

We are grateful to all the patients who gave us written consent to compile this scientific typescript and to herald the same. We'd like to thank our co-workers for helping us with this project.

Table 3. Classification of Lymphangioma.

1. Based on the histopathological feature [10]
Types-I: Lymphangioma simplex or capillary lymphangioma consisting of small, thin-walled capillary-sized lymphatic vessels
Type-II: Cavernous lymphangioma, comprising large, dilated lymphatic vessels with surrounding adventitia.
Type III: Cystic lymphangioma or cystic hygroma exhibiting large macroscopic cystic lymphatic spaces surrounded by fibrovascular tissues and smooth muscles
Type IV: Lymphatic channels appear to be dissecting through dense collagenic bundles.
2. ased on Size of the lesion [12]
Type-1: Macrocystic (>2 cm),
Type-2: Microcystic (<2 cm)
Type-3: Mixed (combination of both)
3. Based on clinical presentation [13,14]
Superficial lesion:
Lymphangioma simplex or capillary lymphangioma (skin),
lymphangioma circumscripta (extremities and genitals), and
lymphangioma cavernous type. (Mouth, lips, cheek, tongue)
Deeper lesion: Cystic lymphangiomas type (Neck and axilla)
4. Based on the anatomical involvement [15]
Stage/class I, infrahyoid unilateral lesions;
Stage/ class II, suprahyoid bilateral lesions;
Stage/class III, suprahyoid or infrahyoid unilateral lesions;
Stage/ class IV, suprahyoid bilateral lesions;
Stage/class V, suprahyoid or infrahyoid bilateral lesions;
Stage/class IV, infrahyoid bilateral lesions
5. Based on the histopathologic findings, has been proposed by Watson and McCarthy [16]
Simple lymphangioma,
Cavernous lymphangioma,
Cellular or hypertrophic lymphangioma,
Diffuse systemic lymphangioma and
Cystic lymphangioma or hygroma.

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Mini Curriculum and Author's Contribution

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