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A rare presentation of glossal lymphangioma

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ABSTRACT

Lymphatic malformations is a commonly encountered congenital vascular malformation in clinical practice. We report a case of lymphatic malformation involving the tongue which presented with enlargement following a systemic infection. The lesion was managed by surgical excision. The histopathological features and treatment options for lymphatic malformations are also discussed.

INTRODUCTION

Lymphatic malformations (LM) of the skin result from hyperplasia of lymphatic network. They are hypothesised to be originating from a primitive lymph sac that fails to connect with the rest of the normal lymphatic system during its embryonic development[1]. They are divided into macrocystic, microcystic and combined types. Macrocystic lesions comprise of cavernous lymphangiomas and cystic hygromas. Microcystic type called lymphangioma circumscriptum are small abnormal lymphatic channels presenting with superficial tiny tense vesicles (frog spawn appearance) [2]. Combined microcystic and macrocystic LMs have both superficial and deep component. They tend to commonly enlarge with the growth of the child.

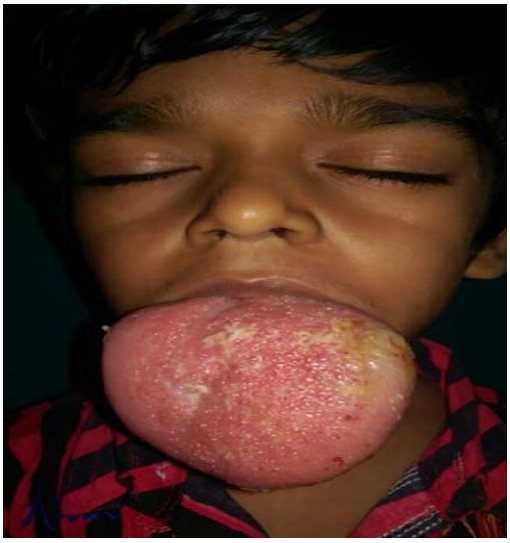
Lymphatic malformations are most common in children with 75% of them present at birth. They can occur anywhere in the body. Most common sites are chest, proximal limb, head and neck region. We present here a case of intraoral combined type of lymphatic malformation presenting with a unique complication.

CASE REPORT

A 7 year old boy presented to the outpatient department of Dermatology with painless massive enlargement tongue for a duration of three months. The boy had a history of enlargement of tongue with little difficulty in closing the mouth since birth. The enlargement of tongue had been confined more to left half of the tongue with adaptation of chewing habits to the other half. There had been no history of bleeding or ulcer over the tongue. Three months earlier, he had an episode of low grade fever which was followed by rapid enlargement of tongue with complete inability to close the mouth. The fever subsided after 3 days. There were no evident features of infective foci.

On clinical examination, anterior two-thirds of the tongue was protruding out of the mouth in the resting state (Fig 1). The protruding portion of the tongue was measuring 14 cm in length and 9 cm in width. The left half was more enlarged than the right with surface showing pebbling and yellowish plaques. Submental and submandibular lymph nodes were enlarged. They were firm, discrete and nontender. There were no dental and mandibular abnormality. There were no skin lesions anywhere else in the body. The clinical differential diagnoses included lymphangioma, hemangioma, neurofibroma and granular cell tumor.

Figure 1 - Massive enlargement of tongue with surface showing pebbling on the left side



The massive enlargement of tongue was causing difficulty in mastication and speech. It was also a threat to airway maintenance. So a definitive surgical procedure was planned. Under nasotracheal intubation, wedge-shaped partial glossectomy of anterior two-thirds of the tongue (Modified Kole's technique) with reconstruction was done. (Fig 5) This enabled an almost complete removal of lesion and full thickness of enlarged tongue[3,4]. The patient recovered well and his tongue healed. Feeding, speech and cosmesis improved remarkably. He has been on follow up for the past 2 months and is doing well.

Macroscopic examination of the resected tongue revealed multiple cystic spaces. Histopathology showed multiple dilated lymphatic channels filled with uniform proteinaceous eosinophilic material and few erythrocytes involving full thickness of the tongue (Fig. 2 & Fig. 3).The cystic spaces were lined by a single layer of endothelial cells with flattened nuclei and surrounded by fibroblasts and lymphoid aggregates. (Fig. 4) The histopathological features were consistent with lymphangioma of the tongue.

Figure 2- Lymphatic dilatations extending on to the epidermis

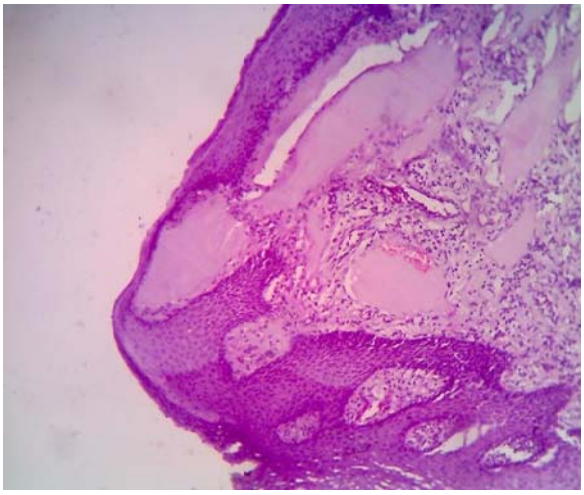


Figure 3- Cystic spaces in between striated muscle of tongue along with lymphoid aggregates

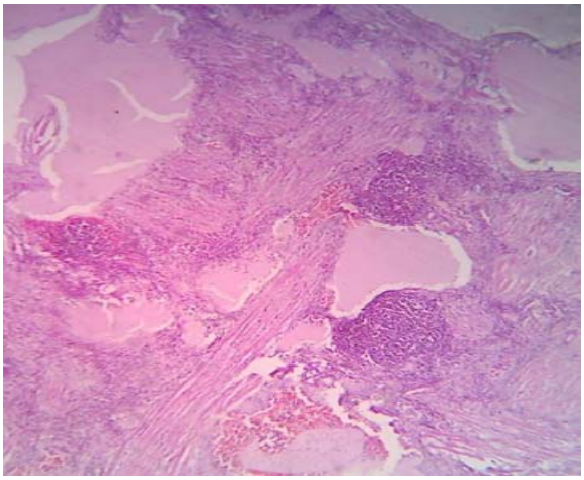


Figure 4- Cystic spaces lined by a single layer of endothelial cells with flattened nuclei

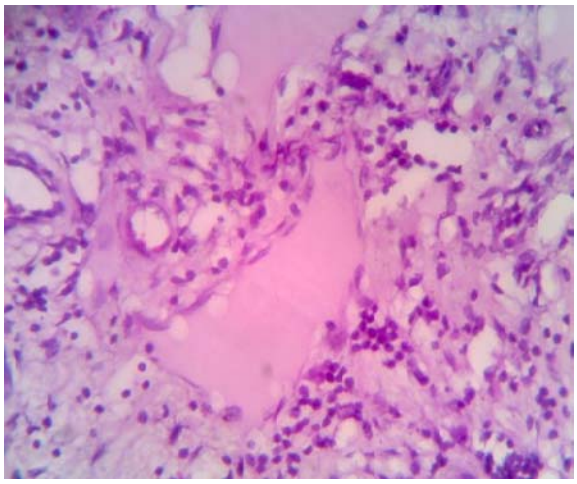
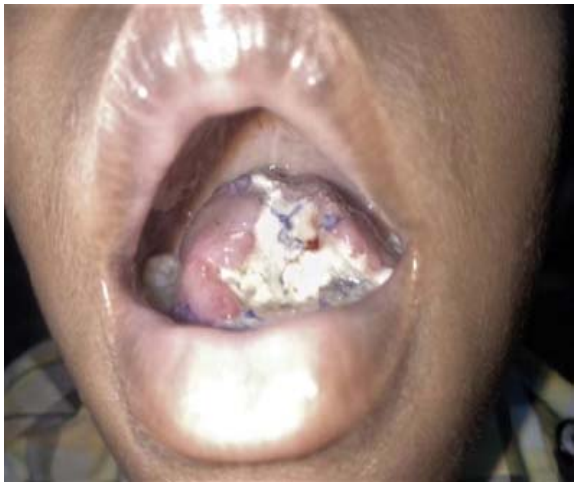


Figure 5- Postoperative day 5



DISCUSSION

Combined lymphatic malformations commonly occur in the cervicofacial region with bone involvement. There may be prognathism and mandibular out growth[5]. The most common site for intraoral combined LMs is the tongue. They can be complicated by inflammatory flares or spontaneous bleeding resulting in sudden expansion of the lesions especially tongue[6]. This can occur following upper respiratory or dental infections. In our case, there was no associated bony abnormality but the rapid growth was preceded by an episode of fever.

Treatment options for microcystic LMs include cryotherapy, CO2 and diode laser, sclerosant injection such as sodium tetradecyl sulfate, pure ethanol, doxycycline, bleomycin, hypertonic saline, 25% Dextrose, OK- 432 (Picibanil) etc[7,8]. This results in inflammatory reaction leading to fibrosis and shrinkage of the treated cyst.

For larger lesions with involvement of deeper structures and functional impairment, surgical excision is the treatment of choice.

CONCLUSION

Intraoral combined cystic lymphangiomas commonly occur over the tongue. Complication with such massive enlargement is uncommon. Surgical correction is the treatment of choice. Recurrence can occur due to incomplete excision or underestimation of the extent of the LM. Prevention of dental caries and other infections is an important measure to prevent recurrence.

REFERENCES

1. Brennan TD, Miller AS, Chen SY. Lymphangiomas of the oral cavity: A clinicopathologic, immunohistochemical and electron-microscopic study. J Oral Maxillofac Surg 1997;55:932-5

2. Bloom DC, Perkins JA, Manning SC. Management of lymphatic malformations. Curr Opin Otolaryngol Head Neck Surg 2004;12:500-4

3. Taher A. Surgical correction of the enlarged tongue. Internet J Head Neck Surg 2008;3(1)

4. Gupta S, Vegad K. Lymphangioma of tongue a rare entity: A case report. IJSS Journal of Surgery. 2015;1:23-25.

5. Edwards PD, Rahbar R, Ferraro NF, Burrows PE, Mulliken JB. Lymphatic malformation of the lingual base and oral floor. Plastic and reconstructive surgery. 2005 Jun 1;115(7):1906-15.

6. Dinerman WS, Myers EN. Lymphangiomatous Macroglossia.The Laryngoscope 1974;291-294.

7. Peters DA, Courtemanche DJ, Heran MK, Ludemann JP, Prendiville JS. Treatment of cystic lymphatic vascular malformations with OK-432 sclerotherapy. Plastic and reconstructive surgery. 2006 Nov 1;118(6):1441-6.

8. Leboulanger N, Roger G, Caze A, Enjolras O, Denoyelle F, Garabedian EN. Utility of radiofrequency ablation for haemorrhagic lingual lymphangioma. International journal of pediatric otorhinolaryngology. 2008 Jul 31;72(7):953-8.