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Spontaneous Regression of Lymphatic Malformation -A Rare Case of Transient Macroglossia

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ABSTRACT

Lymphatic malformation (LM) is a rare benign proliferation of dilated lymphatic vessels with inappropriate communication, filled with lymphatic fluid and lined by endothelial cells. Its occurrence is rare in the oral cavity; however, inside the oral cavity most common site is anterior two-thirds on the dorsum of the tongue, presenting as macroglossia. Macroglossia compromises the airway, often leads to obstructive sleep apnea, malocclusion, and speech and mastication difficulty. The main objective of tongue LM treatment is to preserve the taste sensation, restoration of the tongue size for articulation and cosmesis. We report a rare case of transient intermittent macroglossia due to LM. The patient presented with painful swelling of the tongue, relative macroglossia. There was spontaneous regression of LM with antibiotics and corticosteroids. LM can grow suddenly due to infection or hemorrhage and can shrink spontaneously. Upon treatment of the infection, there is spontaneous regression, total or partial. Therefore, expectant management should be considered in the treatment of tongue LM, to preserve the functions of tongue.

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Introduction

LM are rare tumors that present as localized masses of lymphatic fluid owing to poor communication within the lymphatic channels [1]. They attribute to about 6% of benign tumors in the pediatric age group [2]. 75% of cases arise in the head and neck region, and about 50% of these lesions appear at birth, and around 90% develop by two years of age.Superficial lesions may present as an elevated, transparent group of vesicles, becoming red or purple owing to hemorrhage. In contrast, deeper lesions are soft and diffuse masses with standard color [3]. Local infections along the course of lymphatic drainage will cause LM to swell, protrude, and sometimes become painful. An increase in swelling causes airway obstruction and is a life-threatening pediatric emergency.

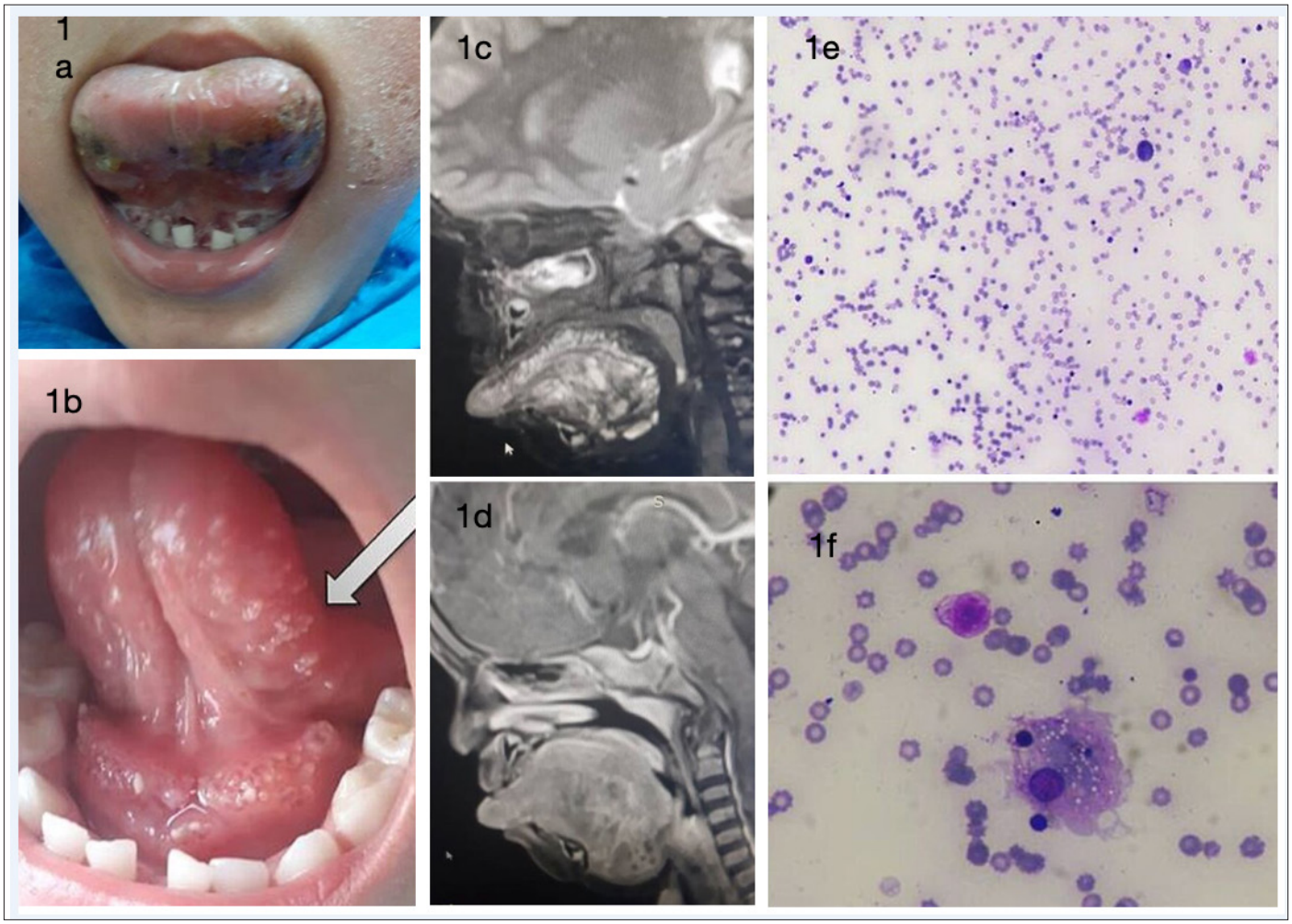
Case presentation

A 4-year-old girl presented to the pediatric outpatient department with difficulty in closing her mouth, a large painful protruded tongue, and drooling of saliva. There was difficulty in eating, speaking, and breathlessness on lying down. In the past, there were two episodes of a sudden increase in the size of the tongue, which was self-limiting with spontaneous regression of tongue size to normal. Swelling of the tongue first developed at two years of age. Upon examination of the oral cavity, the tongue was diffusely enlarged and protruding, keeping the mouth permanently open with hemorrhagic ulceration on the ventral aspect of the tongue with xerosis of the dorsum of the tongue; the floor of the mouth was not visible owing to significant tongue swelling. Palpation of the tongue revealed a soft, cystic, and diffusely tender swelling involving the tongue and floor of the mouth. (Figure 1a). Mild respiratory distress was present in the lying-down position. She was afebrile and had bilateral cervical lymphadenopathy. Her anthropometric evaluation revealed a weight of 13 kilogram (Kg) [between −1 standard deviation (S.D) and −2 S.D], a height of 95.4 centimetre (cm) (between −1 S.D and −2 S.D), and a head circumference of 49.5 cm (50th percentile; WHO-World Health Organisation; MGRS-multicentre growth reference study). The rest of the systemic examination was insignificant.

Ultrasonography of the neck and the oral cavity revealed a large cystic LM measuring 3.5 cm × 3 cm, involving bilateral submandibular and submental spaces with multiple internal septations. No color flow in the cystic cavity on doppler imaging. Magnetic resonance imaging (MRI) revealed a well-defined heterogeneously enhancing mass lesion measuring (4.3 × 4.7 × 4.4) cm, involving the entire tongue, bilateral submandibular, and

submental spaces. It extended through the floor of the mouth up to subcutaneous tissue on either side and caused a mass effect in the form of inferior displacement of mylohyoid muscle (Figure 1c,d). Fine needle aspiration cytology (FNAC) was performed, which revealed the presence of cystic lymphangioma of the tongue with secondary infection (Figure 1e,f).

The patient received intravenous (IV) fluids, antibiotics, analgesics, and steroids; as a result, macroglossia regressed significantly (Figure 1b) with the restoration of the tongue to standard intraoral size.



1a: Enlarged swollen tongue with areas of haemorrhagic and necrotic tissue
1b: Tongue regressed to intraoral size; multiple tiny vesicular and cystic lesions present at the base and the lateral border of the tongue
1c: Sagittal T2/STIR image showing hyperintense lesion with multiple hyperintense loculi in inferior aspect with few showing fluid level within.
1d: Sagittal T1 fat sat post contrast image showing heterogeneous enhancement of the lesion with multiple well-defined nonenhancing areas.
1e: Cytological picture 10 X Magnification suggestive of lymphocytes (benign nature of swelling)
1f: Cytological picture 40 X Magnification suggesting a cystic macrophage

Discussion

LM are slow-growing tumors resulting from premature cessation of the expected growth of the primitive lymph channels during embryogenesis, usually diagnosed in infancy and early childhood. They are associated with Turner's syndrome, Noonan's syndrome, trisomies, cardiac anomalies, fetal hydrops, fetal alcohol syndrome, and Familial pterygium coli. Clinically, these malformations are classified as macrocystic (>2 cm), microcystic (<2 cm), and mixed (combination of both) [4]. LM has a marked predilection for the head and neck region but is rare in the oral cavity. Intraoral LM presents as a localized or diffuse growth most commonly on the anterior two-thirds of the dorsal surface of the tongue, presenting as macroglossia.

Macroglossia is defined as the tongue that protrudes beyond the teeth and alveolar ridge in a resting posture. Primary disorders of the tongue tissue lead to true macroglossia, and when affected secondarily due to tissue infiltration, it is referred to as relative macroglossia [5]. Difficulty with speech and deglutition are frequently associated symptoms, while infection and bleeding are owing to trauma and, in severe cases, airway obstruction is a common complication. With each episode of infection, the lymphatic channels dilate, which leads to progressive swelling of the tongue.

MRI has become the investigation of choice since it depicts the extent and invasion of these lesions. In our case, the diagnosis of LM was confirmed with ultra- sonography and magnetic resonance imaging.

Spontaneous regression is rare and may occur in 1.6–16.0% of cases and is more likely in macrocystic LM with fewer than five septations [6, 7]. LM may regress wholly or partially after a bout of infection in rare instances.

Kato et al., proposed that patients who showed regression had intact out-flow; however, the in-flow sometimes suddenly increased and overcame the out-flow drainage capacity, resulting in temporary expansion of the lymphangiomas [8]. The increase in lymph in-flow was mainly associated with intra-cystic bleeding or infection [9].

The appropriate time to start treatment for LM is unclear. The management of LM depends upon its size, type, anatomical structures, and surrounding tissue involvement. Kato et al., suggested that macro-cystic and mixed lymphangiomas, which suddenly occur in patients aged two years or older, could be observed for a couple of months without treatment to avoid possible complications and unnecessary expenses associated with drainage, sclerotherapy, medication, and/or surgery. Furthermore, microcystic LM should be treated earlier because of the limited possibility of spontaneous regression [8].

Microcystic lesions are diffuse and difficult to eradicate because they do not follow the tissue planes, while macro-cystic lesions are more localized and easy to excise. The treatment objectives are preserving taste and restoring tongue size for articulation and better cosmetic appearance. Oosthuizen et al.advocated expectant management for the first two years in asymptomatic lesions because of chances of spontaneous regression [10].

Our case is an addition to a meager number of the cases reported to have spontaneously regressed. The index case's distinctive features are one having a recurrent course and the second spontaneous regression.

Surgical excision is the modality of choice [4]. However, surgical resection can be challenging due to ill-defined borders and association with major structures. Other treatment options include radiation therapy, cryotherapy, electrocauterization, sclerotherapy (bleomycin, tetracycline, OK-432), embolization, laser therapy; neodymium yttrium aluminum garnet (Nd-YAG, CO2), steroid administration, ligation and radiofrequency tissue ablation, and m-tor inhibitor sirolimus [11].

Conclusion

LM can grow suddenly due to infection or hemorrhage and can shrink spontaneously. Upon treatment of the infection, there is spontaneous regression, total or partial. Therefore, expectant management should be considered in the treatment of tongue LM, to preserve the functions of tongue.

References

1. Rathan JJ, Vardhan BG, Muthu MS, Venkatachalapathy, Saraswathy K, et al. (2005) Oral lymphangioma: a case report. J Indian Soc Pedod Prev Dent 23: 185-189.
2. A Alqahtani , L T Nguyen, H Flageole, K Shaw, J M Laberge (1999) 25 years experience with lymphangiomas in children. J Pediatr Surg 34: 1164-1168.
3. Goswami M, Singh S, Gokkulakrishnan S, Singh A (2011) Lymphangioma of the tongue. Natl J Maxillofac Surg 2: 86-88.
4. Usha V , T Sivasankari , S Jeelani , G S Asokan , J Parthiban (2014) Lymphangioma of the tongue - a case report and review of literature. J Clin Diagn Res 8: ZD12-ZD14.
5. Vogel JE, Mulliken JB, Kaban LB (1986) Macroglossia: A review of the condition and a new classification. Plast Reconstr Surg 78: 715-23.
6. Qin Zhou, Jia Wei Zheng, Hua Ming Mai, Quan Feng Luo, Xin Dong Fan, et al. (2011) Treatment guidelines for lymphatic malformations of the head and neck. Oral Oncol 47: 1105-1109.
7. Perkins JA, Maniglia C, Magit A, Sidhu M, Manning SC, etal. (2008) Clinical and radiographic findings in children with spontaneous lymphatic malformation regression. Otolaryngology–Head and Neck Surgery 138: 772-777.
8. Ling Xiu Ngui, Shashi Gopalan, Chiun Kian Chai (2016) A rare case of spontaneous regression of huge neck lymphangioma post primary infection, Egyptian Journal of Ear, Nose, Throat and Allied Sciences 17: 99-102.
9. Ha J, Yu Y, Lannigan, F (2014) A review of the management of Lymphangiomas. Current Pediatric Reviews 10: 238-248.
10. Oosthuizen J, Burns P, Russell J (2010) Lymphatic malformations: A proposed management algorithm. International Journal of Pediatric Otorhinolaryngology 74: 398-403.
11. Curry S, Logeman A, Jones D (2019) Sirolimus: A Successful Medical Treatment for Head and Neck Lymphatic Malformations. Case Rep Otolaryngol 2019: 2076798.

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