Case Report

Generalized Lymphangioma of the Tongue: A Rare Cause of Macroglossia

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Received: 06 October, 2018.
Revised: 23 February, 2019.
Accepted: 12 April, 2019.
Published: 27 November, 2019.

Abstract

Generalized lymphangioma of tongue is a rare cause of macroglossia in children. It causes mechanical discomfort and functional and psychological disturbances. We report a case of macroglossia due to generalized lymphangioma in a 10 year old where partial glossectomy was done to achieve symptomatic relief.

Keywords: *Lymphangioma tongue, macroglossia, partial glossectomy*

Introduction

Lymphangiomas are uncommon benign tumors of childhood occurring as a result of congenital malformations of the lymphatic system. They are commonly (50%–70%) located in the head and neck region and rarely occur in the oral cavity.[1] Almost half of these lesions are noted at birth, and around 90% develop by the age of 2 years. Only 6% of all lymphangiomas occur in the tongue.[1] Intraoral lymphangiomas are most frequently seen on the dorsum of the tongue followed by palate, buccal mucosa, gingiva, and lips. Lymphangioma of the tongue in children was associated with difficulty in swallowing and mastication, speech disturbances, airway obstruction, mandibular prognathism, open bite, and maxillofacial deformities.[1] Large lymphangiomas may result in airway obstruction and death. We share our case of generalized lymphangioma of the tongue in a 10-year-old child who needed a partial glossectomy.

Case Report

A 10-year-old female child presented with a generalized diffuse swelling of the tongue since birth. The parents had noted her to have an apparently large tongue since birth which gradually increased in size. The child also developed multiple nodules over the tongue which spontaneously burst with watery discharge [Figure 1 - Left picture]. This was associated with excessive salivation, inability to completely close the mouth, chewing and feeding difficulty, and slurring of speech over the past 5 years. There was no history of dyspnea. On examination, the child had diffuse nontender enlargement of the tongue with multiple vesicle-like projections over the dorsal aspect, giving it an irregular granular experience. She also had mandibular prognathism. The rest of the oral cavity and neck were normal.

With a clinical diagnosis of generalized lymphangioma of the tongue, a magnetic resonance imaging scan was done. This showed a large diffuse ill-defined altered signal intensity lesion involving tongue and sublingual regions on either side of the midline. There was no intracranial or intraorbital extension. In view of the symptoms, physical findings, and investigations, a partial glossectomy was planned to provide symptomatic relief to the child.

The area of excision was marked, which was middle one-third of dorsum to the junction of anterior two-third and posterior one-third including the tip and a portion of ventral surface. After nasal intubation, the tongue was infiltrated with 2% lignocaine with optimal dilution of adrenaline. Following excision, the tongue was reconstructed in layers with 3-0 and 4-0 polyglactin sutures [Figure 2]. The child was given nasogastric tube feeding for 10 days following surgery after which

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How to cite this article: Shah AA, Mahmud K, Shah AV. Generalized lymphangioma of the tongue: A rare cause of macroglossia. J Indian Assoc Pediatr Surg 2020;25:49-51.

Access this article online

Quick Response Code:

Website: www.jiaps.com

DOI: 10.4103/jiaps.JIAPS_210_18

Figure 1: Preoperative photograph showing the enlarged tongue and the postoperative photograph after surgery

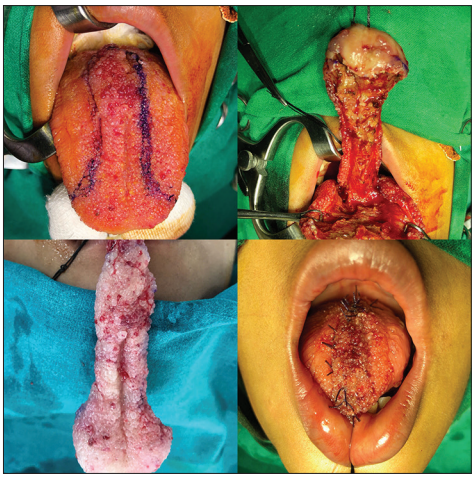

Figure 2: Peroperative photograph showing the line of incision, part of the tongue excised, and the immediate postoperative result

oral feeds were gradually introduced, which were tolerated well. Histopathology confirmed lymphangioma circumscriptum characterized by numerous channels filled with lymphatic fluid in the papillary dermis causing it to expand. Large caliber lymph filled vessels were also seen in the deeper plane.

The child had significant relief from her preoperative symptoms following surgery [Figure 1 - Right picture].

Discussion

Lymphangiomas are benign, idiopathic hamartomas of the lymphatic system that arise from either congenital obstruction or sequestration of the primitive lymphatic enlargement. Fifty percent of these lesions are noted at birth and around 90% develop by 2 years of age. Seventy-five percent of lymphangiomas involve the head and neck region. Three types of lymphangiomas have been described: (a) lymphangioma simplex (capillary lymphangioma), (b) cavernous lymphangioma, and (c) cystic lymphangioma (cystic hygroma).[1,2] The clinical appearance depends on the extent of the lesion. Superficial lesions form elevated nodules, whereas deeper lesions are diffuse.[3] Lymphangiomas rarely affect the oral cavity. Affected sites in the oral cavity may include the tongue, palate, gingiva, lips, and alveolar ridge of the mandible.[4] Lymphangioma of the tongue forms only 6% of all lymphangiomas in the body.[1] It is an important cause of macroglossia in children. These patients may have problems in feeding, speech disturbances, poor oral hygiene, and bleeding from the tongue secondary to trivial trauma. Macroglossia with an irregular and nodular surface with gray and pink projections is the pathognomonic feature of lymphangioma of the tongue.[5] Lymphangioma of the tongue may cause esthetic, occlusal, functional, and psychosocial disturbances in the child.[6] Deep-seated lesions may result in upper airway obstruction. Infected lymphangiomas at the base of the tongue can occasionally result in Ludwig's angina. Most intraoral lymphangiomas are cavernous with large dilated lymph channels, large macroscopic cystic spaces, and small capillary-sized vessels.[7] The lymphatic vessels are located just beneath the epithelium, often replacing connective tissue papillae. The differential diagnosis includes hemangioma, amyloidosis, congenital hypothyroidism, neurofibromatosis, and primary muscular hypertrophy.[5]

The various treatment modalities for lymphangioma are surgical excision, radiation therapy, cryotherapy, electrocautery, sclerotherapy, steroid administration, embolization, laser surgery with Nd-YAG, CO_2, and radiofrequency tissue ablation technique.

The management of generalized lymphangioma of the tongue is indeed challenging. The aim of the treatment should be restoration of the tongue size for articulation, preservation of taste, correction of mandibular and dental deformities, and overall cosmetic improvement. In the present case, looking at the extent of the lesion, symptoms, and secondary cosmetic deformities, surgical excision was the treatment of choice. Because of the age and infiltrating nature of the lesion, a partial glossectomy was planned to give the child symptomatic relief and improve her quality of life.

Laser photocoagulation has been reported useful in controlling the tongue size and removing superficial lymphangioma in some cases. However, the role of laser photocoagulation in such an extensive lesion is debatable. Intralesional sclerotherapy also has a high recurrence rate because of the site and the complexity of the lesion.[1]

Good postoperative care and nursing are prudent in these cases where postoperative edema, infections, cellulitis, seroma formation, and lymphatic fluid leakage are known to occur.[4]

The child is presently 14 months postoperative and in regular follow-up. There is a significant symptomatic improvement in her speech and cosmetic appearance. There are residual vesicles along the lateral border of the tongue, but they are asymptomatic. In the event of any local symptomatic recurrence, the option of using sclerosants is available, and it can be used judiciously and in small doses.

Conclusion

Although lymphangiomas are encountered less frequently in the oral cavity, early recognition, initiation of timely and optimal intervention, and good postoperative and nursing care are important to have optimal results and minimize complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Alqahtani A, Nguyen LT, Flageole H, Shaw K, Laberge JM. 25 years' experience with lymphangiomas in children. J Pediatr Surg 1999;34:1164-8.
2. Kheur Supriya M, Samapika R, Yashwant I, Desai RS. Lymphangioma of tongue: A rare entity. Indian J Dent Adv 2011;3:635-7.
3. Goswami M, Singh S, Gokkulakrishnan S, Singh A. Lymphangioma of the tongue. Natl J Maxillofac Surg 2011;2:86-8.
4. Stănescu L, Georgescu EF, Simionescu C, Georgescu I. Lymphangioma of the oral cavity. Rom J Morphol Embryol 2006;47:373-7.
5. Hancock BJ, St-Vil D, Luks FI, Di Lorenzo M, Blanchard H. Complications of lymphangiomas in children. J Pediatr Surg 1992;27:220-4.
6. Uguru C, Edafioghor F, Uguru N. Lymphangioma of the tongue with macroglossia: A case report. Niger J Med 2011;20:166-8.
7. Tasca RA, Myatt HM, Beckenham EJ. Lymphangioma of the tongue presenting as Ludwig's angina. Int J Pediatr Otorhinolaryngol 1999;51:201-5.