Macroglossia combined with lymphangioma: a case report

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A four year old white female with a clinical presentation of macroglossia is described. Speech disturbances and occasional episodes of traumatic injury to the tongue with severe bleeding brought the patient to seek dental care. Lymphangioma was diagnosed after incisional biopsy. The differential diagnosis of tongue enlargement in children is discussed including review of the literature relevant to the diagnosis and treatment of lymphangioma.
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INTRODUCTION

Macroglossia, enlarged tongue, is a component of numerous syndromes; many of them caused by inherited metabolic anomalies, in which the increase in tongue size is a manifestation of the visceromegaly related to abnormal lysosomal storage of carbohydrates macromolecules. Lysosomal storage diseases, commonly associated with macroglossia, are found in Hurler syndrome, Hunter's syndrome and Maroteaux-Lamy syndrome. Other macroglossia-associated syndromes are Beckwith-Wiedemann, Neurofibromatosis type 1, Hemangiomatosis (Sturge-Weber) and congenital lymphangioma (cystic hygroma).[1]

Macroglossia may be either congenital or secondary in type. Congenital macroglossia is due to an overdevelopment of the musculature, which may or may not be associated with generalized muscular hypertrophy or hemihypertrophy.[2]

Secondary macroglossia may occur as a result of a tumor of the tongue, such as a diffuse lymphangioma or hemangioma, from neurofibromatosis, or occasionally from blockage of the efferent lymphatic vessels in cases of malignant neoplasms of the tongue.[3]

Relative macroglossia is commonly described in conditions in which the jaws are small and the tongue of relatively normal size, giving the protruded appearance of macroglossia (Down syndrome, congenital hypothyroidism, Angelman syndrome). True macroglossia often produces displacement of teeth and maloclusion due to increased pressure exerted by the tongue on the teeth.[1]

Lymphangioma is a benign tumor of lymphatic vessels. Its occurrence is relatively rare and it is characterized by proliferation of lymphatic vessels.[4-5] According to Watson and McCarthy,[6] the majority of cases are present at birth and 95% of the tumors aroused before the age of 10 years with no predilection of sex.[1,6] Prenatal diagnosis is possible through ultrasonographic technology and recently reported.[7]

Lymphangioma of the tongue is a rare condition[8,9] and is the most common cause of congenital macroglossia.[1-3] The dorsal surface of the tongue is the most commonly affected, presenting irregular nodularity with gray and pink and pink projections.[10,11]

The purpose of this case report is to describe a case of macroglossia, secondary to lymphangioma, in a young child and discuss the possible differential diagnosis and treatment options of this phenomenon in children.

CASE REPORT

A four year old girl presented to the Department of Oral Medicine with a chief complaint of an enlarging mass of the tongue. Her mother mentioned that the tongue was enlarged at birth, however, during the last years, the tongue has enlarged significantly causing speech disturbances, occasional severe bleeding on mastication and difficulty in exercising good oral hygiene due to the size and position of the tongue. Medical history was insignificant except that the child was born to both parents at the age of 45.

At examination, the child seemed normal for her age and revealed above average intelligence. The face was symmetrical, however severe macroglossia was noted (Figure 1). The tongue was red and covered by multiple

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Figure 1. Lymphangioma of the tongue.

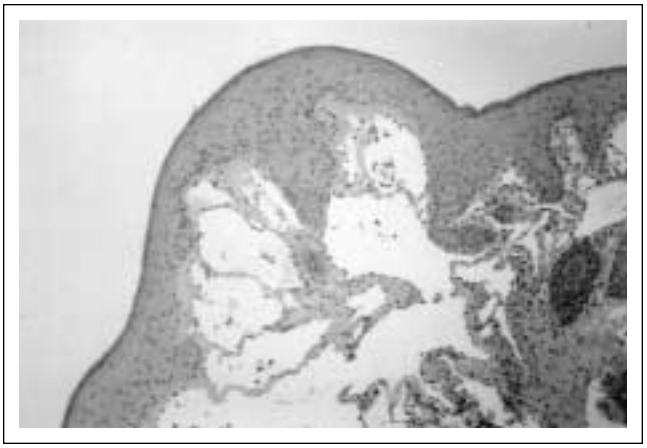

Figure 2. Lymphangioma of the tongue-histological section

bluish vesicle-like lesions on the dorsal aspect. Upon compressing the tissue, no pain was elicited. However, it resulted in marked transient blanching. The complementary oral and extraoral examinations were noncontributory. The patient was caries-free with a slight anterior open-bite.

Differential diagnosis of the lesion included pyogenic granuloma, papilloma, fibroma, hemangioma, rhabdomyoma, neurilomona and lymphangioma. Incisional biopsy from the dorsal aspect of the tongue was compatible with lymphangioma, revealing normal maturation of the epithelium with marked dilation of the lymphatic channels throughout the upper portion of the connective tissue, chronic inflammatory infiltrate predominantly lymphocytic and plasmacytic in the stroma (Figure 2). The treatment recommended and performed was tongue reduction using sclerosing agents.

DISCUSSION

Watson and McCarthy[6] classified lymphangiomas as simple, cavernous, cellular or hypertrophic, diffuse systemic and cystic or hygroma when affecting the neck region. The cavernous type of lymphangioma is the most common.

Lymphangiomas can cause airway interference. Hartl *et al*.[19] studied the results of an outcome survey of 18 cases of pediatric lymphangioma with dyspnea from encroachment on the tongue base, parapharyngeal space, and/or larynx. Eighteen patients were treated. The average age at initial surgery was 22 weeks (median, 5 weeks). All presented with at least unilateral suprahyoid and infrahyoid cavernous (microcystic) lymphangioma.

The tongue base was involved in 11 patients, the parapharyngeal space in 12, and the larynx in 8. Neck dissection was performed initially in all patients. Tracheotomy was performed in 9 patients (50%). Macroglossia was treated by V glossoplasty. Parapharyngeal extensions were treated by cervicotomy or endoscopy, and larynx and tongue base extensions by carbon dioxide laser photocoagulation. Supraglottic laryngectomy was performed in 2 patients. The average follow-up was 4 years postoperatively. One postoperative death occurred.

Sixteen (94%) of the remaining 17 patients had residual lymphangioma. Eight (89%) of the 9 patients with tracheotomy underwent decannulation (average duration, 22 months). Ten patients had persistent symptoms, and 6 were asymptomatic.

Involvement of the upper airway seems to be the determining prognostic factor in extensive lymphangioma. Patients with dyspnea by external compression of cervical lymphangioma on the airway responded well to surgery. Aggressive surgical treatment did not seem to significantly improve the prognosis in patients with intrinsic involvement of the upper airway. Less aggressive, symptomatic therapy may be an alternative to avoid mutilating surgery in patients with intrinsic involvement of the airway.

Lymphangioma lesions are not tender or painful and unlike hemangiomas, do not regress spontaneously. Lesions can also be seen on the palate, buccal mucosa, gingival and lips. Growth usually ceases at puberty. Inflammation from trauma or infection results in excessive lymph tissue formation, severe pain and may lead to swallowing difficulties and airway obstruction.[5] Spontaneous regression is rare and surgical excision is probably the treatment of choice.[12] Surgical techniques include Nd-YAG laser,[13,14] CO_2 laser,[8,15] and injection of sclerosing agents like OK-432.[16]

Recently, a new and more conservative approach was suggested using radiofrequency tissue ablation techniques.[17]

Reduction in tongue size with preservation of motor and sensory function was best accomplished by edge resection of the tip of the tongue.[5] Recurrence of the removed lesions is common in 41% of the cases.[6]

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Gimeno Aranguez *et al*.[18] studied 145 lymphangiomas in children from birth to 16 years of age. The results from the population studied were lymphangiomas were most frequent during the first year of life, most commonly arising on the head, neck and axilla. Lymphangiomas are divided into cavernous, cystic (cystic hygroma) and mixed. They are composed of lymphatic channels of variable size and have a tendency to occur between the tissue layers. They concluded that most important complications are related to the location and large size. The intra-abdominal lymphangiomas caused intestinal obstruction. The lymphangiomas in the tongue caused macroglossia. The local recurrences are is 6% for incomplete excision in cases of complicated surgery.

Since lymphangioma is primarily a disease of childhood, the pediatric dentist might be the first health care personnel to be encountered with the lesion. An early and prompt diagnosis of the lesion might reduce the severity of different developmental complications such as speech disturbances, avoidance from dental care and bleeding associated with oral trauma.

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