Macroglossia Surgical Correction : A Case Report

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Abstract

Congenital macroglossia is associated with a variety of syndromes, most commonly Down syndrome and Beckwith-Widemann syndrome. Clinically, macroglossia may impair the airways, cause dysphagia and poor cosmesis. A variety of treatments have been proposed and surgical resection is the most common. We review management of patients and describe a surgical technique, consisting of a keyhole resection to decrease the width and length of the tongue. Patients had improved cosmesis and better function of upper airways, with no change in speech and feeding. Keyhole resection is an effective treatment for macroglossia.

Keywords: Macroglossia, congenital, Macroglossia, surgery.

Introduction

Macroglossia is a rare condition in pediatric patients; however, when present, there are significant symptoms such as airway obstruction, difficulty feeding and aesthetic deformities[1].

The first report of macroglossia was a description of oral lymphatic malformation, in 1854, by Virchow and Uber[2].There are many causes of macroglossia in children, which are divided into true and relative. The true cause occurs when histological abnormalities correlate with the clinical findings of tongue enlargement. Vascular malformations, muscle hypertrophy and tumors are the most common causes of true macroglossia. Relative macroglossia includes all cases in which histology does not explain the pathological condition. Down syndrome is the main cause of relative macroglossia[3]. Several treatments have been suggested for patients with significant symptoms and surgery is the most indicated therapy by means of varied procedures[3-4]. A one-year-old female patient with increased tongue volume and protrusion since birth (Fig. 1) associated to difficult swallowing (feeding only by nasogastric tube) and phonation. History of two previous events of tongue trauma with bleeding and sudden increase in tongue size. Upon examination, she presented considerably enlarged tongue and protrusion, deformities in dental arches and a large cystic tumor in the lower portion of the tongue and mouth floor. Tracheotomy was performed under sedation before the imaging exams due to difficult endotracheal tube placement. Computed tomography revealed lesion with attenuation coefficient and heterogeneous uptake of contrast medium, which was predominantly hypodense, with poorly-defined limits, involving tongue and the space under tongue. The lesion shows a marked volumetric increase of the tongue and reduced diameter of the oropharyngeal air column (Fig. 2). Surgical management of macroglossia in children.

Discussion

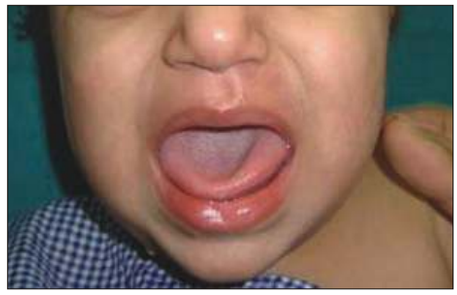
Macroglossia is an uncommon condition with significant morbidity. The characteristic picture is tongue protrusion that may lead to dental and facial abnormalities, mucosal exposure and dryness, exposure to trauma, dysphagia and difficulty phonation, airway obstruction, salivation and growth delay[1,3-4].

Patients with macroglossia should be assessed to identify the primary cause before suggesting any type of treatment. The cause of macroglossia is already identified in many patients with associated syndromes. If there is no apparent cause, patients should be evaluated for metabolic disorders; in that, 24% of patients with hypothyroidism present macroglossia[5]. Imaging studies of the tongue and airways may be beneficial in cases of vascular malformations.

Treatment should be based on severity of symptoms and of macroglossia. When tongue enlargement is small, with minimum symptoms, no treatment is recommended. On the other hand, very affected children may benefit from early interventions, and surgical resection is the most effective treatment.

Lymphatic malformations in tongue are rare[6-7], and in most cases they involve its anterior portion, as einstein. 2007; 5(2):166-169.

The purpose of treatment in such patients is to excise the lesion, preserving phonation and swallowing and favoring an appropriate oro-facial development. Partial glossectomy with lymphangioma resection



achieved its goal at an early age, as described .

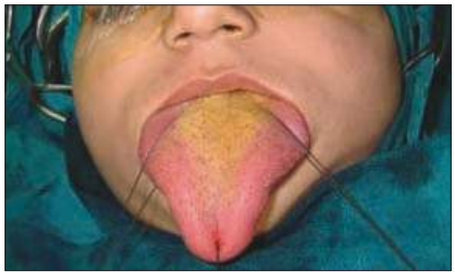
Beckwith-Wiedemann syndrome was first described by Beckwith, in 1963, and later by Wiedemann, in 1964, as exomphalos/ omphalocele, macroglossia and gigantism syndrome (EMG/OMG syndromes). Today it is related to other malformations[(8)]. It is a genetic syndrome of overgrowth that is relatively common, and characterized by congenital abnormalities, such as visceromegaly, macroglossia, abdominal wall defects, pre- and postnatal overgrowth and neonatal hypoglycemia. It is a polymorphous syndrome subject to a variable combination of signs and symptoms. Among the several anomalies mentioned, macroglossia is the most common manifestation of the syndrome, found in 82% to 99% of the individuals affected. It may be associated with a spectrum of craniofacial alterations[9].

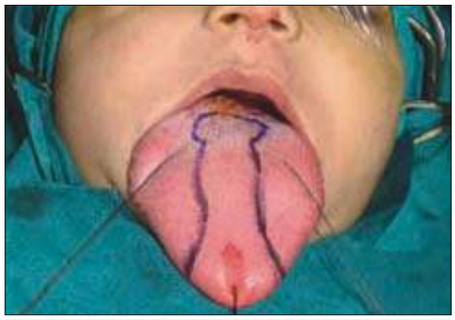
The syndrome may cause difficulty swallowing, phonation and even respiratory problems due to inability to keep the tongue inside the mouth. This may lead to dryness, ulcerations and even infections in the tip of the tongue.

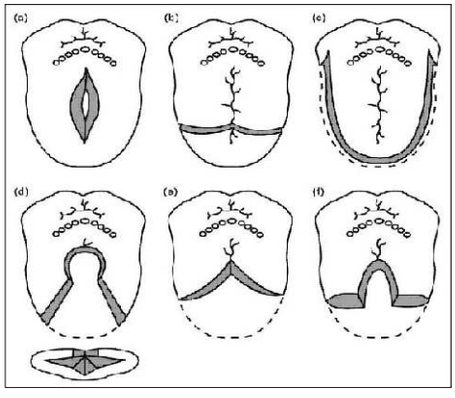
Moreover, untreated macroglossia may result in impaired craniofacial development, with open bite and inclined incisors, leading to a prognathic aspect[9]. The histological exams of macroglossia show muscle hyperplasia or even normal histology. Many authors have a clinical approach of this anomaly and wait the child grow, since the tongue tends to

accommodate inside the mouth as they grow. However, surgical procedure is mandatory in some patients[10-11], like our case 2, in whom the aesthetic deformity was significant and associated to swallowing disorders and respiratory problems. Partial glossectomy is the procedure most often performed in such cases in order to reduce tongue to normal size and preserve its function. Excellent aesthetic and functional results were obtained with this technique, with improvement of ronchi, appearance, feeding and speech after surgery. The child could develop speech and breath adequately, thus avoiding future craniofacial problems.

The surgical technique employed this case recommends keyhole resection of tongue[3]. The large anterior wedge excised provides good width reduction, and the broad circular incision results in decreased

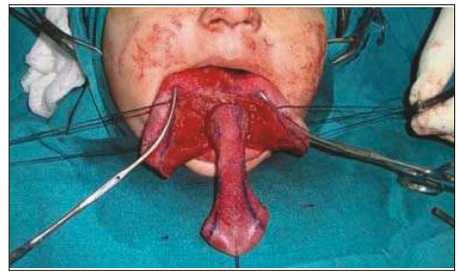


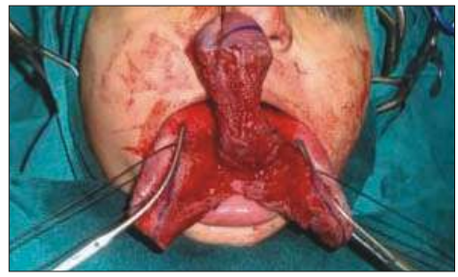


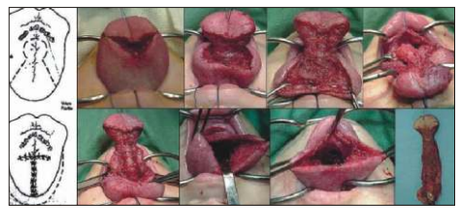


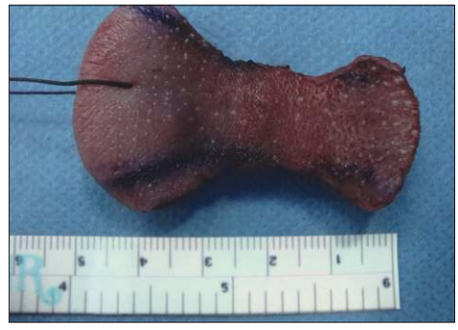
tongue volume and length. It is a versatile resection that may be used in most cases of macroglossia.

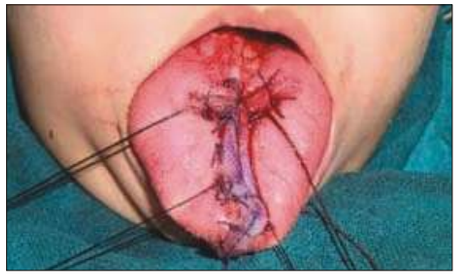
Endotracheal in tubation of these patients is often difficult and we chose to perform tracheotomy in both cases, to avoid

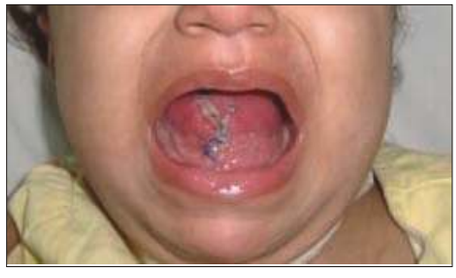












anesthetic risks and local edema with chocking and aspiration in the postoperative period.

The surgical treatment of macroglossia promotes aesthetic and functional improvement; if performed at an early stage, it may prevent dento-alveolar complications.

Speech, deglutition and saliva management also improve. There might be complications related to partial tongue resection, such as ankylosis, globular tongue with an in sensitive tip[12]. The tongue body may remain wide even when the new tongue size is normal. No postoperative complications were observed with the technique employed in both cases presented.

References

1 Horn C, Thaker HM, Tampakopoulou DA, De Serres LM, Keller JL, Haddad J. Tongue lesions in the pediatric population. Otolaryngol Head Neck Surg. 2001;124(2):164-9.

2. Virchow R, Uber. Makroglossic and pathologiscle neubildung quergestreifler muskelfasern. Virchows Arch (Pathol Anat). 1854;7:127.

3. Morgan WE, Friedman EM, Duncan NO, Sulek M. Surgical management of macroglossia in children. Arch Otolaryngol Head Neck Surg. 1996;122(3):326-9.

4. Clauser L, Tieghi R, Polito J. Treatment of macroglossia in Beckwith-Wiedemann syndrome. J Craniof Surg. 2006;17(2):369-72.

5. Grant DB, Smith I, Fuggle PW, Tokar S, Chapple J. Congenital hypothyroidism detect by neonatal screening: relationship between biochemical severity and early clinical features. Arch Dis Child. 1992;67(1):87-90.

6. Balakrishan A, Bailey CM. Lymphangioma of the tongue. A review of pathogenesis, treatment and the use of surface laser photocoagulation. J Laryngol Otol. 1991;105(11):924-30.

7. Lobitz B, Lang T. Lymphangioma of the tongue. Pediatr Emerg Care. 1995;11(3):183 -5.

8. Weng EY, Mortier GR, Graham Jr JM. Beckwith-Wiedemann syndrome. An update and review for the primary pediatrician. Clin Pediatr (Phila). 1995;34(6):317-26. Review.

9. Rimell FL, Shapiro AM, Shoemaker DL, Kenna MA. Head and neck manifestations of Beckwith-Wiedemann syndrome. Otolaryngol Head Neck Surg. 1995;113(3):262-5.

10. Mixter RC, Ewanowski SJ, Carson LV. Central tongue reduction for macroglossia. Plast Reconstr Surg. 1993;91(6):1159-62.

11. Siddiqui A, Pensler JM. The efficacy of tongue resection in treatment of symptomatic macroglossia in the child. Ann Plast Surg. 1990;25(1):14-7.

12 Kopriva D, Classen DA. Regrowth of Tongue Following Reduction Glossoplasty in the Neonatal Period for Beckwith-Wiedemann Macroglossia. J Otolaryngol. 1998;27(4):232-5. einstein. 2007; 5(2):166-169