Surgical management of macroglossia in children: two case reports

Tratamento cirúrgico de macroglossia em crianças: relato de dois casos

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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; Case reports [Publication type]

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).

Two cases of macroglossia in children seen from 2002 to 2005, at the Hospital Infantil Darcy Vargas, São Paulo, SP, are reported, addressing surgical technique and the results obtained.

CASE 1

A one-year-old female patient with increased tongue volume and protrusion since birth (figure 1) associated to difficulty 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 (figure 2).

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Magnetic resonance angiography of the head did not show anomalous vessels towards the tongue tumor and mouth floor (figure 3).

The child underwent partial glossectomy by means of a keyhole resection of the tumor and mouth floor with a good result (figure 4).

The pathological examination revealed a cystic hygroma of tongue and mouth floor. Currently she has excellent cosmesis, with spontaneous reduction in prognathism and slight difficulty in phonation.
CASE 2

A nine-month-old female patient with Beckwith-Wiedemann syndrome presented a significant increase in tongue volume and protrusion, difficulty swallowing, presence of breathing sounds and excessive salivation (figure 5).

She was submitted to tracheotomy one week before glossectomy. Partial glossectomy was performed with a keyhole resection (figure 6) with good final aesthetic results (figure 7).

Figure 5. Preoperative aspect

Figure 7. Postoperative aspect

Figure 6. Surgical procedure

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).

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 (4). 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 (5-7), and in most cases they involve its anterior portion, as
observed in case 1. The purpose of treatment in such patients is to excise the lesion, preserving phonation and swallowing and favoring an appropriate orofacial development. Partial glossectomy with lymphangioma resection achieved its goal at an early age, as described in case 1.

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(9). 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(10-13), 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 in both cases 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, such as in cases of macroglossia due to lymphatic malformation (case 1) or associated to Beckwith-Wiedemann syndrome (case 2).

Endotracheal intubation 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 insensitive 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.

CONCLUSION

Keyhole resection showed to be an effective treatment for cases of macroglossia in children and, in the two cases reported presently, improved facial aesthetics and airway function were observed, with no changes in speech or swallowing.

REFERENCES