SURGICAL MANAGEMENT OF MACROGLOSSIA IN SYNDROMIC PATIENTS: CASE REPORT

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Recebido em 28/02/2018. **Aceito** para publicação em 03/04/2018

ABSTRACT

The Beckwith Wiedemann Syndrome (BWS) is a genetic disease of low prevalence. It has several clinical manifestations. Macroglossia affects most cases and the recommended treatment is surgical excision. This clinical case reports an 8 month-old boy diagnosed with BWS presenting macroglossia, swallowing restriction, respiratory obstruction and airways maintenance by tracheostomy. The patient underwent a glossectomy surgical technique. The incision was performed in a keyhole shape in the central region with a harmonic scalpel. Intra-operative blood loss due to extensive vascularization of the tongue elevates morbidity of pediatric patients submitted to tongue surgery. Glossectomy and an interdisciplinary treatment enabled reduction of several complications associated to macroglossia. The harmonic scalpel improved postoperative results and reduces the risk of complications and sequels.

KEYWORDS: Beckwith Wiedemann Syndrome, Glossectomy, ultrasonic surgical procedures.

1. INTRODUCTION

The Beckwith Wiedemann Syndrome (BWS) is a rare congenital disease of low prevalence. It was first reported separately by Beckwith in 1963 and Wiedemann in 1964[1-2]. The characteristics of this syndrome include neonatal hypoglycemia, macroglossia, macrosomia, hemihyperplasia, omphalocele, cytomegaly of the adrenal cortex, embryonal tumors, renal abnormalities, visceromegaly, diaphragm defects, hypoglycemia and anterior linear ear lobe creases or posterior helical ear pits[3]. This syndrome has an apparent origin in an alteration in the expression of genes a tumor suppressor[4-6].

Macroglossia is the most common clinical feature of BWS, present in approximately 90%. Muscle hypertrophy is the cause for tongue size increase. It is generally present at birth and early death may occur due to feeding difficulties resulting from macroglossia. Growth rate slows around age 7-8 years. Tongue reduction surgery is frequently indicated in infancy or early childhood[3].

Several surgical techniques have been developed for glossectomy. Most techniques recommend removal of midline lingual tissue in keyhole or wedge shape[7-9]. The harmonic scalpel provides accurate cutting capacity at low temperature, less hemorrhage and less post-surgical discomfort[10-13].

The present work aims to report a case of a child diagnosed with magroglossia due to Beckwith Wiedemann Syndrome, submitted to partial glossectomy using an harmonic scalpel.

2. CASE REPORT

Figure 1. Preoperative appearance. Tongue protrusion.

An 8-month-old boy (Figure 1) was referred to the Oral and Maxillofacial Surgery Department of Porto Nacional Regional Hospital (Porto Nacional, Tocantins,

Brazil) for assessment of macroglossia related to subjacent diagnosis of Beckwith Wiedemann Syndrome. The patient had at the first appointment a tracheostomy performed at the first week of life. He had respiratory hypersecretion and needed continuous aspiration. The mother reported the child was feeding well. He had not evidence of obstructive symptoms.

The parents' main concern was the appearance of his tongue. It was enlarged and protruding. They also complained the tracheotomy prevented speech development.

Over the course of two months, the patient had feeding problems and repeated episodes of lingual trauma. Tongue reduction surgery was the proposed treatment option. Surgical treatment and the publication of the case report was authorized by the mother.

Figure 2. Keyhole shape incision drawing with methylene blue.

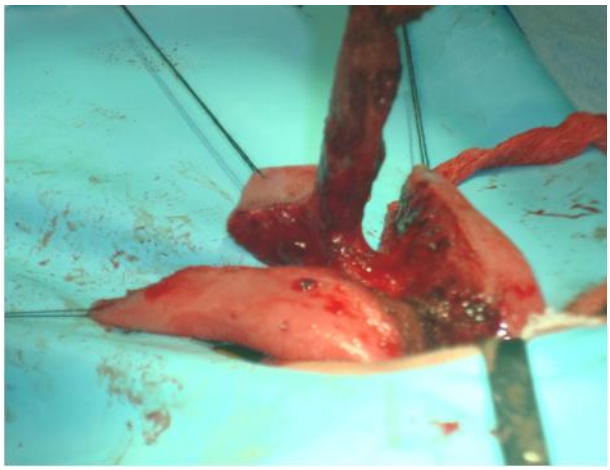

Figure 3. Tongue incision with ultrasonic scalpel.

A partial glossectomy was the chosen treatment. The tongue was symmetrically fixed and tensioned using 3-0 nylon wires (Shalon Suturas, Goiânia, Goiás, Brazil). The procedure used a surgical a keyhole shape[14]. This shape was marked with methylene blue (Fórmula Mais, Palmas, Tocantins, Brazil) in the tongue dorsum for incision planning (Figure 2). The incisions were made in total thickness of the tongue central region by a harmonic scalpel (Ethicon Inc., Somerville, New Jersey, USA) and allowed removal of a tongue segment preserving the lateral tissue (Figures 3). The procedure led to minimal bleeding, without lesion of nerve tissue. The suture was made in anatomical planes with resorbable wire (Vicryl, Ethicon, Somerville, New Jersey, USA) (Figure 4). Minimum pain and edema occurred at immediate post-operative period. There were no signs of dehiscence or infection. Patient discharge was on the third post-operative day (Figure 5).

Figure 4. Suture of the tongue in anatomical planes.

Figure 5. Postoperative follow-up after 8 weeks

Two months of phonoaudiological and psychological follow-up of the patient and the family showed enhanced tongue functionality. The parents reported significant improvement in tongue movements, swallowing and a reduction in salivary secretion. They reported satisfaction with the tongue appearance after surgery. There was no

sensorial compromise after surgery and no further episodes of tongue trauma. The patient had medical appointments for evaluation of tracheotomy removal after glossectomy. Follow-up appointments proved significant reduction of tongue size and increased function.

3. DISCUSSION

The Beckwith-Wiedemann Syndrome is a genetic disease of excessive growth with several clinical symptoms. Macroglossia, neonatal hypoglycemia, renal alterations, visceromegaly, nevus flammeus in the frontal region and a higher incidence of childhood neoplasias are the main manifestations[5-7].

Tongue increased could lead to obstruction of the airways, problems with swallowing, facial growth deficit, dental deformities, speech problems, lingual trauma, abundant and persistent salivation. Tongue reduction by glossectomy is an effective treatment option to relieve these symptoms. Glossectomy should maintain a narrow tip of the tongue with sufficient length to allow normal tongue movements. It is important to cause no harm to the sensorial functions of the tongue[7-9].

Several surgical techniques for glossectomy have been described in the literature, but Morgan's technique[14] is the most used. The incision shape is similar to a keyhole. This shape allows to preserve neurovascular tissues[7-9]. The tongue has extensive vascularization and hemorrhage is a great concern. Pediatric patients have an increased risk for bleeding[10-13].

Glossectomy traditional techniques use a cold scalpel or monopolar cauterization. Several recent reports indicate the use of the harmonic scalpel. It produces a hemorrhage-free procedure and considerably reduces operative time. The use of the harmonic scalpel also reduces postoperative pain and edema compared to monopolar cauterization. These signs and symptoms are reduced due to ultrasonic homeostasis at a lower temperature (50-100 ◦C) than monopolar cauterization, (150-400 ºC)[10-14]. The harmonic scalpel provided better operative and postoperative features in this case.

BWS patients require an adequate treatment plan to obtain long-term positive results. Tongue surgical reduction is not able to achieve clinical improvements without an interdisciplinary approach. dentoskeletal alterations demand orthopedic and orthodontic treatment, combined to physiotherapy, phonoaudiology and psychological support[6].

4. CONCLUSION

Macroglossia is highly prevalent in Beckwith Wiedemann Syndrome patients. Glossectomy and an interdisciplinary treatment enabled reduction of several complications associated to macroglossia. The harmonic scalpel improved postoperative results and reduces the risk of complications and sequels.

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