CASE REPORTS

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Coblation for Congenital Macroglossia in Beckwith-Wiedemann Syndrome

ABSTRACT

Objective: To present a rare case of congenital macroglossia managed with radiofrequency ablation.

Methods:
Design: Case report
Setting: Tertiary government hospital
Patient: One

Results: A case of a congenital macroglossia in a 4-year-old female with Beckwith-Wiedemann Syndrome is presented. Neither breathing nor swallowing difficulty was associated with the enlarged tongue. Coblation-assisted ablation of the tongue deformity was performed. There was minimal bleeding, pain and swelling postoperatively. Tongue mobility and taste sensation were unaffected.

Conclusion: A new and more conservative approach to surgery for congenital macroglossia using radiofrequency ablation (coblation) has been described. Coblation-assisted ablation of lingual tissue may be an effective therapy for patients with macroglossia providing satisfactory functional and cosmetic outcome.

Key words: *macroglossia, Beckwith-Wiedemann Syndrome, coblation*

A large tongue poses a therapeutic challenge. Reduction of tongue size and improvement of function are the goals of management. Medical therapy may suffice if tongue enlargement is due to systemic disease, but surgical reduction offers the best functional and cosmetic results. Standard surgical procedures directly remove a wedge of tongue muscle and mucosa and are associated with significant morbidity.[1] The challenge has been to find a conservative treatment with low morbidity and better results than those achieved with cold steel or diathermy excision. Recent studies have advocated the use of a plasma-mediated radiofrequency device (coblation) for tongue reduction. It provides the ability to remove tissue at a low temperature, thereby causing less tissue destruction and resulting edema.[1]

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The ArthoCare EVac 70 Xtra Plasma Wand and Coblator II Surgery System were provided by ArthoCare Corporation, Sunnyvale, CA, USA.

Dr. Vicente signed a disclosure that he is a speaker for the ArthroCare Corporation and does not receive honoraria and travel allowances except for the use of the Coblator II System loaned to him for research purposes. Other than this, he does not have any proprietary or financial interests with ArthroCare or with any organization that may have a direct interest in the subject matter of this manuscript, or in any product used or cited in this study.

The other authors signed a disclosure that they have no proprietary or financial interest with any organization that may have a direct interest in the subject matter of this manuscript, or in any product used or cited in this article.

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We report a rare case of congenital macroglossia successfully treated with coblation.

CASE REPORT

A 4-year-old female was brought for evaluation of macroglossia. The patient was born term, large for gestational age (11.5 lbs) via Cesarean section to a 26-year-old primiparous mother**.** Maternal and family histories were unremarkable. A large tongue was noted during physical examination at birth. The left side was slightly larger than the right and one-third of the entire tongue protruded from the mouth. There was no breathing difficulty associated with the enlarged tongue. Newborn screening and thyroid function tests were both normal. Abdominal ultrasound revealed enlarged kidneys. Unfortunately, due to financial difficulties she was unable to continue follow-up visits until she reached four years of age.

The patient's tongue grew in size as she aged, with the anterior 1/3 still protruding from the mouth. She was unable to close her mouth without notable effort and the tongue would always protrude from the mouth, with minimal drooling. Speech was intelligible and there was no impairment in swallowing. She was a kindergarten pupil with no observed developmental delay. However, the unsightly physical appearance of the large tongue evidently affected interaction with her peers.

Physical examination showed a normocephalic child, with growth and development at par with age. Intraoral examination revealed a 7 x 4 x 1.5 cm pinkish, smooth, enlarged tongue with good tone and mobility *(Figures 1 a, b)*. The left side was prominently larger than the right, with preferential lateralization of the tongue tip to the right upon protrusion. Examination of dentition showed an open bite with Class III malocclusion *(Figure 1 c)*. Inspection of the ears demonstrated posterior helical pits and creases on both earlobes. Abdominal findings revealed an omphalocoele *(Figure 1 d)*. The left upper and lower extremities were observed to be larger than the right. The rest of the physical examination findings were normal.

Karyotyping was 46 XX (normal female). Non-contrast and contrast-enhanced 64-multislice CT images of the oral cavity revealed a markedly enlarged tongue, left side more than the right, measuring approximately 7.6 cm in length on the unprotruded study and about 8.4 cm on protrusion *(Figures 2-4)*. The width of the left side measured about 3.6 cm while the right measured 1.4 cm *(Figure 4)*. The left lingual artery and left masseter muscle were noted to be more prominent compared to the right. Sleep cine MR sagittal T1 images demonstrated significant glossoptosis with near-complete effacement of the hypopharynx but with intermittent opening. No apneic episodes were noted on limited sleep study.

Because of the prominent tongue and the associated psychological problems, the patient's parents decided to seek surgical intervention. She underwent coblation-assisted ablation of lingual tissue under

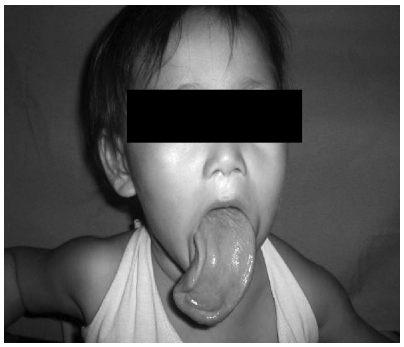


Figure 1. A. Protruded tongue

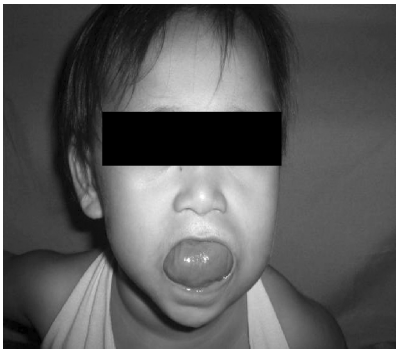


Figure 1. B. Incomplete mouth closure

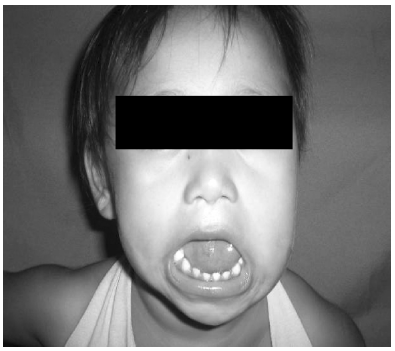


Figure 1. C. Malocclusion

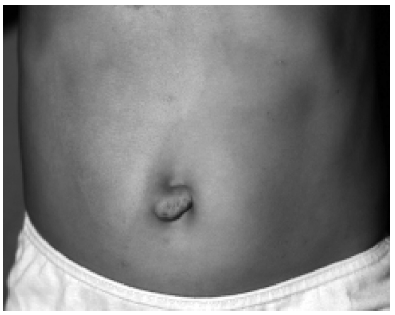


Figure 1. D. Abdominal wall defect

general anesthesia.

Pre-operative markings delineated the course of the lingual arteries and the desired extent of tongue reduction *(Figure 5)*. A midline incision was made 8 cm from the tongue tip. An ArthoCare EVac 70 Xtra Plasma Wand attached to the Coblator II Surgery System (ArthoCare Corporation, Sunnyvale, CA, USA) at an ablation setting of 7 was inserted into the midline incision and held in place for 10 seconds. It was then advanced 5 mm deep along the midline, with care to stay medial to the marked boundaries of the lingual artery. The tongue incision was left open. The same coblator wand was then introduced into the area demarcating the desired tongue length 5 cm from the tip. Trimming of the edges was carried out in full thickness until the desired tongue length was achieved. The tip was closed using absorbable sutures.

Postoperative bleeding, pain, and edema were minimal. Oral antibiotics, steroids and paracetamol were continued until 5 days after discharge.

On the 3rd post-operative week, the tongue size on protrusion was 4 x 4 x 2 cm, with good cosmetic result *(Figure 6)*. Minimal drooling was noted. There was no sensory deficit and no limitation of tongue mobility. The patient did not report any difficulties with swallowing and in manipulating an intra-oral food bolus. There was no change in taste sensation. Three months after the operation, tongue size was measured at 3.5 x 3 x 1.5 cm, the left side thicker than the right *(Figure 7)*. She had full tongue mobility with no alteration in swallowing, speech and taste sensation. The tongue now fit comfortably in the mouth (Figure 8). The parents were satisfied with the result and overall evaluation of the patient's condition was considered very satisfactory.

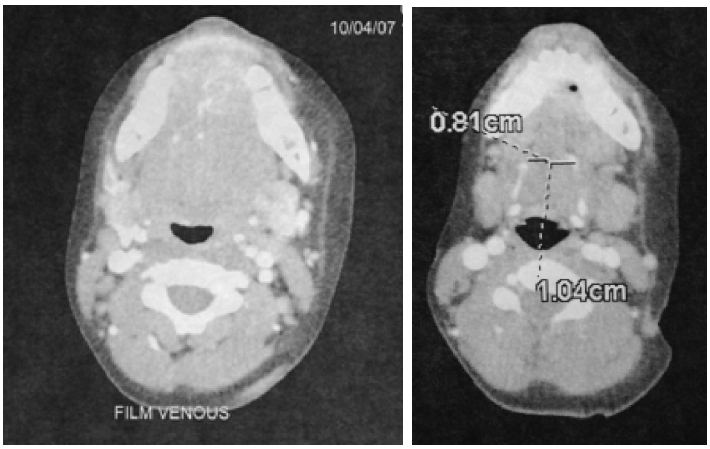
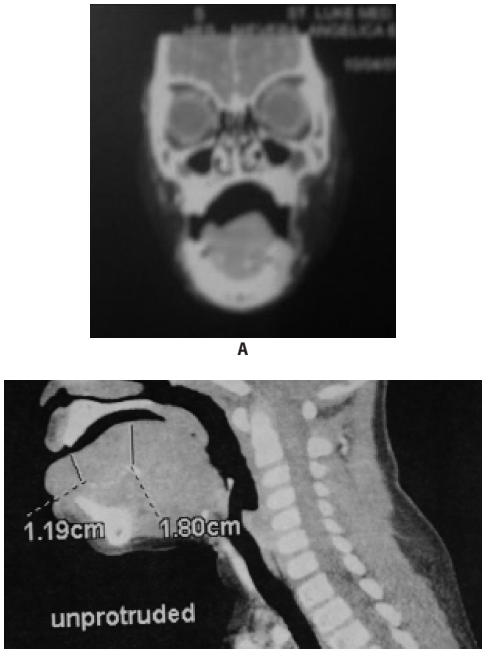


Figure 2. Axial CT scan of the Oral cavity showing prominent tongue.



A

B

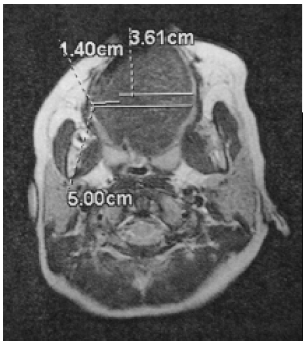
Figure 3. Coronal and sagittal CT scan of the unprotruded tongue.

DISCUSSION

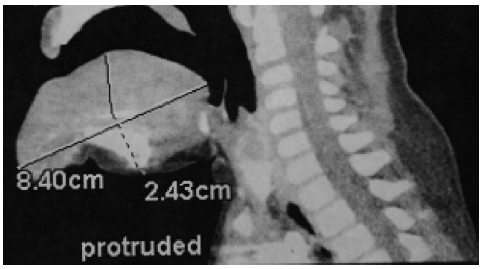
Overgrowth disorders such as Beckwith-Wiedemann syndrome (BWS) can present with an enlarged tongue. The disease was initially described by Beckwith in 1963 and Wiedemann in 1964. Its prevalence is estimated to be approximately 1 in every 17,000 live births, and 97.5% of these patients have macroglossia.[3] The syndrome results from chromosomal changes in the imprinted 11p15.5 region that cause increased levels of the fetal growth factor insulin-like growth factor 2.[4] The most common clinical findings are the triad of macroglossia, abdominal wall defect (omphalocoele, umbilical hernia, diastasis recti) and macrosomia (prenatal gigantism, postnatal gigantism or both). Other clinical findings include neonatal hypoglycemia, renal abnormalities, visceromegaly, hemihypertrophy, nevus flammeus of the forehead, ear anomalies (anterior linear earlobe creases, posterior helical pits) and an increased incidence of childhood neoplasms.[4-6] The diagnosis can be established if at least three diagnostic findings are present.[4]

The effects of an enlarged tongue include airway obstruction, impairment of normal speech and swallowing, maxillofacial abnormalities, and ulceration and necrosis of the exposed tongue. Moreover, the psychological consequences arising from the patient's physical appearance can result in the false impression of mental deficiency, leading to poor social development and low self-esteem.

Medical management of macroglossia may suffice if it is due to systemic disease. However, surgical reduction offers the best functional and cosmetic result and minimizes morbidity. The indications for surgical tongue reduction include airway obstruction, sleep apnea,



A



B

Figure 4. Axial and sagittal CT scan with tongue measurement on protrusion.

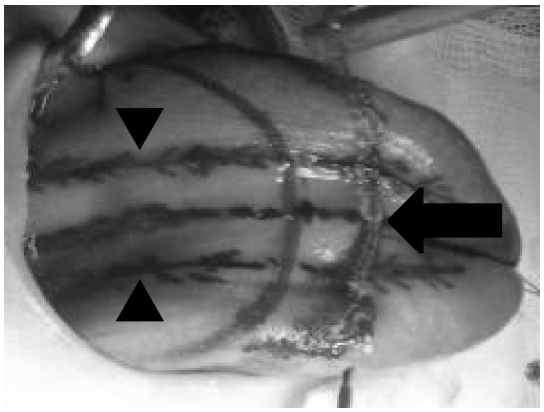


Figure 5. Pre-operative markings outlining the course of the lingual arteries (arrowheads) and the desired extent of tongue reduction (big arrow).

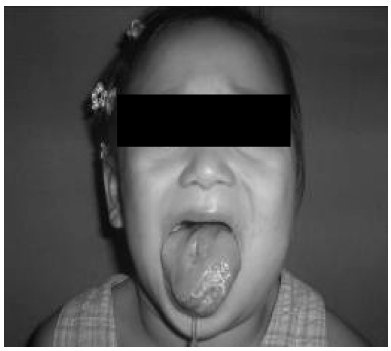


Figure 6. 3 weeks post-op

speech difficulties, dysphagia, recurrent local trauma, and cosmetic concerns.[9] The goal of surgery is to reduce tongue size and improve function, but extensive resection risks damage to the neurovascular bundles and the global nature of the macroglossia poses difficulties in creating a normal tongue.

Many surgical strategies have been proposed to reduce tongue size. Standard surgical procedures are invasive and directly remove a wedge of tongue muscle and mucosa.[9-15] Significant pain and morbidity are encountered with large incisions in the oral cavity. Recent studies have advocated the use of a plasma-mediated radiofrequency device (coblation) for tongue reduction.[1,16-19] Coblation technology provides the ability to remove tissue at a low temperature, thereby causing less tissue destruction and resulting edema. [1,16,17,19] Although most of the present literature center on coblation as a novel surgery for tongue base reduction in obstructive sleep apnea patients, it has been shown to safely reduce lingual volume in a porcine model.[17] It has also been used for the treatment of lymphatic malformation in the tongue and lip.[16, 19]

Following our experience, coblation-assisted ablation of lingual tissue may be an effective therapy for patients with macroglossia providing satisfactory functional and cosmetic outcome.

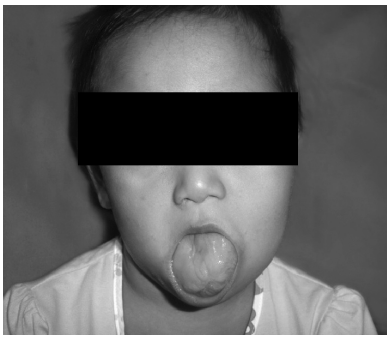


Figure 7. 3 months post-op

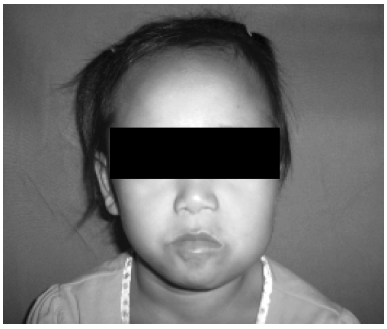


Figure 8. mouth closed post-op

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