

CASE REPORT

MACROGLOSSIA – A RARE INDICATION FOR TRACHEOSTOMY IN A CHILD: A CASE REPORT

Shankar Tati¹, Manish Kumar², Geetha C³, Ranganath Swamy D⁴, Anjani Kumari⁵

HOW TO CITE THIS ARTICLE:

Shankar Tati, Manish Kumar, Geetha C, Ranganath Swamy D, Anjani Kumari. "Macroglossia – A Rare Indication for Tracheostomy in a Child: A Case Report". Journal of Evolution of Medical and Dental Sciences 2014; Vol. 3, Issue 56, October 27; Page: 12853-12858, DOI: 10.14260/jemds/2014/3708

ABSTRACT: Macroglossia is a disorder in which the Tongue is larger than normal, is usually caused by an increase in the amount of tissue on the Tongue. This condition can be seen in certain inherited or congenital disorders, we are here presenting a case of Macroglossia, which were demanded for an Elective Tracheostomy in a child of 03 years. The patient was referred from Nizams Institute of Medical Sciences for securing airway by Tracheostomy since patient was in stridor, before starting the sclerotherapy. It is an emergency condition when patient presents with stridor Tracheostomy indicated, no treatment required for mild cases or cases with minimal symptoms. Speech Therapy may be beneficial, surgery to reduce the size of the Tongue (Reduction Glossectomy) is useful and recommended in severe cases. Recently the cobulator is used for reduction glossectomy.

KEYWORDS: Tracheostomy, Macroglossia, Beckwith – Wiedemann syndrome, Amyloidosis, Dyspnea, Sialorrhea, Down's syndrome, stridor.

INTRODUCTION: Macroglossia is uncommon and usually occurs in children, it is usually clinically diagnosed, and macroglossia has been reported to have a positive family history in 6% of cases. The most common cause of tongue enlargement is vascular malformations (eg. Lymphangioma & Hemangioma) and muscular hypertrophy. (Beckwith-Wiedemann syndrome and Hemihyperplasia). It has an extensive list of possible causes, severe enlargement of the tongue can cause cosmetic and functional difficulties including in speaking, eating, swallowing, and sleeping, treatment is depend upon the exact cause, Its treatment has been largely surgical in the modern era.

CASE REPORT : A Male child aged about 03 years Resident of Bhupalpally, Warangal Dist, referred from Nizams Institute of Medical Sciences for Elective Tracheostomy before starting Sclerotherapy for reduction in size of the Tongue, patient was suffering with large tongue since birth but increased in size gradually and attained the present size.

The patient approached at Gandhi Hospital doctors who referred him to Plastic Surgery department of Nizams Institute of Medical Sciences, before planning for surgery / Sclerotherapy the patient was referred for Elective Tracheostomy. Since the tongue was extremely large and protruding out of the oral cavity difficult intubation was anticipated

The patient was admitted, after all necessary investigations the Elective Tracheostomy was done under General Aneasthesia, Jockson's Metallic Tracheostomy tube No. 16 was inserted patient was observed for 24 hours and sent back to NIMS for further management.

CASE REPORT

MRI CHECK PHOTOS:



Figure 1

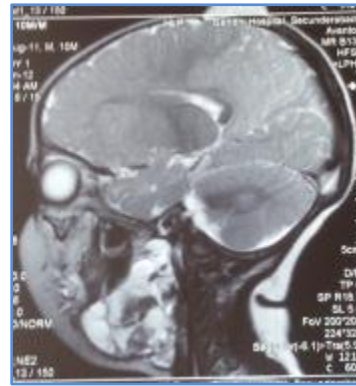


Figure 2

MRI Check: Evidence of large lobulated diffuse soft tissue mass is present in the right pterygoid space. The mass shows multiple cystic areas with variable contents. Some of the cysts show fluid filled levels. The mass extends medially involving the paratonsillar region, laterally extends into infra temporal space. There is no intra cranial extension of the mass. Screening of brain is normal.

Impression: Multiplaner of Soft Tissue Diffuse Hemangioma Involving Right Fascial Spaces.

MRI NECK PHOTOS:



Figure 3



Figure 4

MRI Neck :- Large lobulated T1 iso, T2 STIR hyperintense lesion noted in right side of neck extending from level of C6-C7 vertebrae to right temporal fossa. There are multiple T1 hyperintensity and fluid – fluid levels seen with in the lesion – S/o bleed. Evidence of multiple septa and phleboliths. The lesion measures 5x8cm approximately. Superiorly the lesion is involving subcutaneous plane and temporalis muscle extending inferiorly involving right parotid gland, right submandibular gland, masseter muscle, medial pterygoids extending into the infra temporal fossa and parapharyngeal spaces. Lesion is involving the base of tongue and at this level it is extending to opposite side.

CASE REPORT

However carotid & jugular vessels are indented, there is no indentation over Oropharynx, Naso & Laryngopharynx.

Impression: Feature Suggestive of Vascular malformation - ? venous / lymphatic in the right side of the Neck.

DISCUSSION: Macroglossia is the medical term for an unusually large tongue, it is un common, usually occurs in children. Judging the size of the tongue is subjective as there are no existing measures for assessing tongue size; evaluation of the enlarged tongue is based on both clinical and functional assessment of the tongue. The most common causes of tongue enlargement are vascular malformations (e.g. Lymphangioma or hemangioma) and muscular hypertrophy (e.g Beckwith – Weidemann syndrome or hemihyperplasia)⁽¹⁾. Enlargement due to lymphangioma gives the tongue a pebbly appearance with multiple superficial dilated lymphatic channels.

Amyloidosis is an accumulation of insoluble proteins in tissues that impedes normal function.

This can be a cause of macroglossia if amyloid is deposited in the tissue of the tongue, which gives it a nodular appearance. Beckwith – Wiedemann syndrome is a rare hereditary condition, which may include other defects such as omphalocele, visceromegaly, gigantism or neonatal hypoglycemia ⁽²⁾, the tongue may show a diffuse, smooth generalized enlargement.

The face may show maxillary hypoplasia causing relative mandibular prognathism. Apparent macroglossia can also occur in down syndrome⁽³⁾. The tongue has a papillary, fissured surface. Macroglossia may a sign of hypothyroid disorders.

The common causes of Macroglossia in children Down Syndrome, Beckwith – Wiedemann syndrome, Amyloidosis, Sandhoff’s disease, Cretinism, Hemangioma, Congenital Hypothyroidism, Lymphangioma, Myxedema, Pompe disease. The secondary effects of the macroglossia is dependent to some extent on the size and shape of the tongue⁽⁴⁾.

1. Changes to Appearance:

- The tongue may protrude out of the mouth for some of the time, most of the time and in some children all of the time.
- The degree of tongue protrusion may vary in different situations, For example, it may protrude more when the child is tired or concentrating. There is also a tendency for tongue protrusion to increase while the child is babbling / talking ⁽⁵⁾.
- The tongue may rest inside the lower lip causing the lip to protrude and appear “floppy”.
- It may cause increased spacing between the teeth (“splaying of the teeth”) ⁽⁶⁾.
- It may cause an anterior open bite which is a gap between the upper front teeth and the lower teeth.
- It may cause the lower jaw to protrude that the top jaw.

2. Feeding difficulties:

- In infants, breast or bottle feeding may be difficult if the child has breathing difficulties.
- The baby may be unable to make a good seal around the teat of a bottle or nipple because of the tongue size.

CASE REPORT

- Movement of the tongue during eating may be restricted making it difficult to chew and control the food within the mouth during eating.
- As the child develops teeth, the child may sometimes bite the tongue during eating if the tongue rests over the margins of the teeth, may need a referral to a speech and language therapist who specializes in feeding difficulties.

3. Speech difficulties:

- Surprisingly, macroglossia has little impact on the development of speech unless it is extremely large. However, it does affect the way the child looks when he/she talks. Here are some examples. In the normal speaker, tongue tip sounds /t, d, n, l, s, z/ are produced with the tip of the tongue behind the upper teeth. When the tongue is large, it can be difficult to place the tongue tip behind the upper teeth and the child produces these sounds with the body of the tongue against the bottom of the top teeth instead. Normal production of the sounds f and v may be difficult. Some children with an enlarged tongue are unable to produce the sounds p, b and m with both lips together which is the normal way to produce these sounds and they make these sounds with the tongue against the top lip. All the changes to speech production mentioned above will not affect the clarity of speech but will change the way that the child looks when talking.

4. Respiratory difficulties:

- The child may have to sleep with his / her mouth open which can cause the tongue to become dry to cracked.
- Child may be more susceptible to upper respiratory tract infections.
- A few children have difficulties with breathing caused by thickening at the base of the tongue as this can block the upper airway.
- Macroglossia is usually clinically diagnosed. Sleep endoscopy and imaging may be used for assessment of obstructive sleep apnea. The initial evaluation of all patients with macroglossia may involve abdominal ultrasound and molecular studies for Beckwith – Wiedemann syndrome.⁽⁷⁾

Treatment options for Macroglossia:

1. Speech therapy: if a child has a true macroglossia, speech therapy aimed to encourage the child to keep his / her tongue within the mouth is not a management option. However, the child may have poor oral motor control, speech and feeding difficulties for other reasons. If the child has any concerns at all about speech, oral motor skills or feeding skill the child should be referred to a speech and language pathologist.

2. Surgery: Surgery is not always recommended in all cases of macroglossia. In some cases the macroglossia may be too mild to warrant surgery. If the child has other medical problems surgery may not be advocated. Additionally, some parents feel that they do not wish their child to undergo an operation of this nature.

CASE REPORT

Tongue Reduction Surgery ^(8,9,10): Tongue reduction surgery aims to reduce the length, width and bulk of the tongue. After surgery, the tongue should be able to rest inside the mouth behind the front teeth but be able to protrude and moisten the lips. A successful operation should leave no visible scarring on the tongue and good mobility of the tongue for feeding and speech.

There is no consensus as to the best age to carry out the surgery but generally under the age of two years is thought to be favorable to prevent the lower jaw from being pushed forward.

CONCLUSION: Macroglossia means a large tongue; the most common causes of Tongue enlargement are vascular malformations and muscular hypertrophy. It is an uncommon condition, and usually occurs in children. Treatment and prognosis of macroglossia depends up on its cause and also up on the severity of the enlargement and symptoms it is causing. In our case the tongue was very largely enlarged causing respiratory obstruction, hence it was advised for elective tracheostomy, doing tracheostomy for macroglossia is a rare situation. Hence we are presenting this case report.

REFERENCES:

1. Bouquot, Brad W.Neville, Douglas D.Damm, Carl M. Allen, Jerry E. (200). Oral & Maxillofacial Pathology (2.ed.ed). Philadelphia: W.B. Saunders. PP. 9-10.
2. Dios, Pedro Diz; Posse, Jacobo Limeres; Sanroman, Jac into Fernandez; Garcia, Emma Vazquez (September 2000). "Treatment of macroglossia in a child with Beckwith-Wiedemann syndrome." Journal of oral and maxillofacial surgery: official journal of the American Association of Oral and Maxillofacial Surgeons 58 (9): 1058-61.
3. Guimaraes CV, Donnelly LF, Shott SR, Amin RS, Kalra M (October 2008). "Relative rather than absolute macroglossia in patients iwht Down syndrome: implications for treatment of obstructive sleep apnea". Pediatr Radiol 38 (10): 1062-7.
4. Prada, CE; Zarate, YA; Hoplin, RJ (February 2012). "Genetic causes of macroglossia: diagnostic approach." Pediatrics 129 (2):e431-7.
5. Tei E, Yamataka A, Komuro Y.Huge lymphangioma of the tongue: a case report. Asian Surg.Oct2003; 26 (4): 228-30.(Medline).
6. Topouzelis, N; Iliopoulos, C; Kolokitha, OE (April 2011). "Macroglossia." International dental journal 61(2): 63-9.
7. Perkins, JA (December 2009). "Overview of macroglossia and its treatment." Current Opinion in Otolaryngology & Head and Neck Surgery 17 (6): 460-5..
8. Balaji SM. Reduction glossectomy for large tongues. Ann Maxillofac Surg. Jul 2013; 3(2):167-172. (Medline).
9. Matsumoto K, Morita Kl, Jinno S, Omura K. Sensory changes after tongue reduction for macroglossia. Oral Surg Med Oral Pathol Oral Radiol. Aug 15 2012; (Medline).
10. Morgan WE, Friedman EM, Duncan NO, Sulek M.Surgical management of macroglossia in children. Arch Otolaryngol Head Neck Surg. Mar 1996; 122 (3):326-9. (Medline).

CASE REPORT

AUTHORS:

1. Shankar Tati
2. Manish Kumar
3. Geetha C.
4. Ranganath Swamy D.
5. Anjani Kumari

PARTICULARS OF CONTRIBUTORS:

1. Professor, Department of ENT, Osmania Medical College, Hyderabad.
2. Assistant Professor, Department of ENT, Osmania Medical College, Hyderabad.
3. Assistant Professor, Department of Anaesthesia, Osmania Medical College, Hyderabad.
4. Assistant Professor, Department of ENT, Osmania Medical College, Hyderabad.

5. Assistant Professor, Department of ENT, Osmania Medical College, Hyderabad.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. T. Shankar,
12-5-149/6/A,
Flat No. 201, Sajjas Ambiance,
Vijayapuri, Tarnaka,
Hyderabad-500017,
Telangana, India.
Email: drshankar_ms@yahoo.com

Date of Submission: 13/10/2014.
Date of Peer Review: 14/10/2014.
Date of Acceptance: 22/10/2014.
Date of Publishing: 27/10/2014.