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Case Report

Congenital macroglossia: case report of a rare disease

Sandesh Srivastava, Rahul Kumar, Money Sachdeva, Richa Raj*, Om P. Singh

Department of Pediatrics, Narayan Medical College and Hospital, Jamuhar, Rohtas, Bihar, India

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***Correspondence:**
Dr. Richa Raj,
E-mail: drops0105@gmail.com

ABSTRACT

Macroglossia is a clinical disorder in which the tongue is larger than normal. Enlargement can be true as seen in vascular malformations or it could be due to muscular enlargement The prevalence of congenital macroglossia is less than 5 per 100,000 birth. Various causes include acromegaly, Beckwith-Wiedemann syndrome, congenital hypothyroidism, Down syndrome, and lymphangioma or haemangioma. It may cause significant symptoms in children such as sleep apnea, respiratory distress, drooling of saliva, difficulty in swallowing and dysarthria. Long-standing macroglossia leads to an anterior open bite deformity, increased incidence of upper respiratory tract infections and failure to thrive.

Keywords: Macroglossia, Tongue reduction, Partial glossectomy

INTRODUCTION

Macroglossia or large tongue is commonly seen in vascular malformations or it may be purely a muscular enlargement or hyperplasia of muscle fibres of the tongue.[1] This refers to the protrusion of the tongue beyond the alveolar ridge or teeth.[1,2] It can be divided into two categories: true macroglossia and relative macroglossia, or pseudo macroglossia. "True macroglossia" refers to macroglossia caused by histologic abnormalities within the tongue secondary to an underlying condition, such as muscular hypertrophy and vascular malformation. Relative macroglossia occurs in congenital syndromes like Pierre Robin syndrome and hypotonia in Down syndrome.[2] There is relative macroglossia associated with micrognathia.

Pseudo macroglossia

Allergic reactions to medications that cause the tongue to swell, enlarged tonsils and/or adenoids that displace the tongue, low palate and decreased oral cavity volume, severe maxillary deficiency with narrow palatal arch, severe mandibular deficiency (retrognathism), and local oral tumor that displaces the tongue may all cause macroglossia.

True macroglossia

Congenital causes

Idiopathic tongue muscle hypertrophy, salivary gland tumour. haemangioma, lymphangioma, syndromes e.g. Beckwith-Wiedemann (most common cause in childhood), Behmel, Laband, lingual thyroid, mucopolysaccharidoses - Hunter and Hurler syndromes, and hamartomas.

Acquired causes

Metabolic/endocrine conditions e.g. hypothyroidism, cretinism, amyloidosis-most common cause in adults, acromegaly, and myxoedema.

Inflammatory/infectious causes i.e. Ludwig angina, pemphigus vulgaris, diphtheria, tuberculosis, syphilis, actinomycosis, sarcoidosis, candidiasis, and neoplastic

conditions such as lymphangioma, haemangioma, carcinoma, plasmacytoma, and lymphoma.

In children, it may cause significant symptoms such as sleep apnea, respiratory distress, drooling, difficulty in swallowing and dysarthria. Long-standing macroglossia leads to an anterior open bite deformity, mucosal changes, exposure to potential trauma, increased incidence of upper respiratory tract infections and failure to thrive. Tongue movements and sounds may be affected by the macroglossia. Speech articulation may also be affected.[1-4]

The goals of surgery in a case of macroglossia include restoration of the size and shape of the tongue for function and preserve the existing functions of the tongue including articulation, deglutition, and taste. The surgical procedure should result in a tongue that remains behind the lower dental arch at rest, yet can wet the lips on protrusion.[2,3]

CASE REPORT

7-year-old male child, presented to the hospital with enlargement of tongue since birth. With increasing age, it gradually increased over time and he started having difficulty in feeding, not able to close his mouth properly and was drooling saliva from his mouth. He began to have feeding problems, he was not able to close his mouth properly and was constantly drooling. He had noisy breathing at rest. The child also gave history of burning sensation while chewing food and was having difficulty in breathing in supine position.

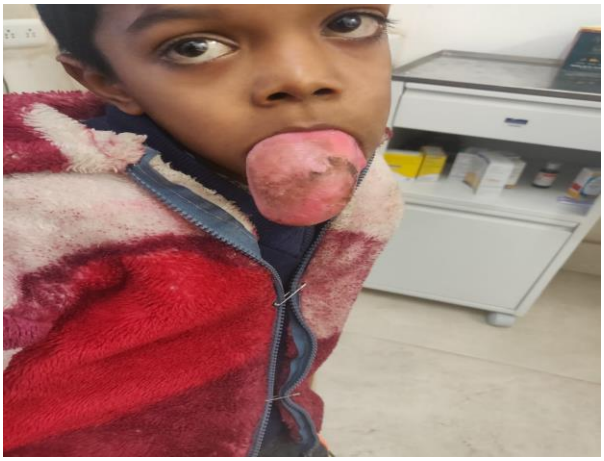

Figure 1: Seven year old male child with congenital macroglossia.

On general examination patient was conscious, oriented to time, place and person. Pulse rate was 92/min, respiratory rate was 22/min. Patient's SpO_2 was 96% on room air, and extremities were warm. In supine position the child was having respiratory distress in the form of tachypnoea (RR-32/min) and was having intercostal and subcostal retraction. Per abdomen examination revealed child's abdomen was soft, non-tender, non- distended. Bowel sound were present. Central nervous system examination revealed child was conscious, oriented with Glasgow coma scale (GCS) of 15/15. All the reflexes were normal. Bilateral pupils were normal in size, equal and reactive to light. Examination of others systems were unremarkable. No facial dysmorphism were present.

On physical examination, the tongue was increased both in length and in width and was large globular roughly measuring 7×5×2 cm with anterior open bite and impression of the lingual surface of mandible molars on the edges of the tongue. No tissue infiltrate were seen.

Investigations

Hemoglobin (Hb) was 11.5 gm%, total leucocyte count (TLC) was 9,800/cmm with differential leucocyte count (DLC) was $N_{64}\,L_{28}\,M_3\,E_2\,B_0$, platelets were 2.4 lakh/cmm, haematocrit was 37.7%. C-reactive protein (CRP) was 2 mg/l. Triiodothyronine (T3) was 1.23 ng/ml, thyroxine (T4) was 9.40 ug/dl, and thyroid stimulating hormone (TSH) was 3.42 uIU/ml. Insulin like growth factor-1 (IGF-1) levels were normal. Ultrasonography (USG) Doppler study of the tongue was normal.

The child was subjected to partial glossectomy with very good results.

DISCUSSION

Management of a case of macroglossia depend on the cause. Medical therapy for macroglossia is useful only when the etiology of the disease is a clearly defined, medically treatable entity such as hypothyroidism, infection, or amyloidosis.[5] No medical treatments have proven useful when the etiology is unclear or the histology reveals simple hypertrophy or hyperplasia.

Causes like congenital hypothyroidism, acromegaly, Down's syndrome can be managed with appropriate medical treatment. Surgical aim while managing a case of macroglossia is to reduce the tongue globally while preserving the taste, sensation and mobility of the tongue. There are numerous techniques recommended. There is no consensus in the surgical management of a case of macroglossia.[6] Various procedures for macroglossia lead to improved cosmesis, speech, mastication and feeding.[6]

CONCLUSION

Congenital macroglossia is an uncommon condition. It is important to achieve uniform global reduction of the enlarged tongue for functional as well as aesthetic reasons. The management of a case with this disorder requires a dedicated team approach. The team comprises of paediatrician, plastic surgeon, dental surgeon, speech therapist along with continuous psychological support.

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