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Original Research Article

Macroglossia and outcome of severity based treatment regime

Umesh Kumar, Vijaykumar Huded*, Sudipta Bera, Pradeep Jain, Arnab Sarkar, Yasharth Sharma

Department of Plastic and Reconstructive Surgery, Institute of Medical Sciences, Banaras Hindu University, Varanasi Uttar Pradesh, India.

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***Correspondence:**
Dr. Vijaykumar Huded,
E-mail: drvijayh21@gmail.com

ABSTRACT

Background: This study aims to categorize macroglossia patients into mild, moderate, and severe groups and formulate a treatment plan depending upon the severity of tongue involvement.
Methods: Eight patients presented with macroglossia between 2018 and 2020 are reviewed retrospectively. The patients were categorized into three subgroups depending upon the clinical presentation and subjected to either sclerotherapy or surgical debulking. The clinical outcome as a reduction of size and symptomatic improvement were analyzed and categorized after a minimum of 6 months follow-up.
Results: Eight patients (5 males and 3 females) aged 10-40 years with a mean age of 28.25 (SD 10.29) years were included in the study. Of eight patients, four cases were of vascular malformation, three of neurofibroma, and one was due to amyloidosis. Four patients were treated with surgery, three with sclerotherapy while one patient was managed with combined modalities. On average, 58% and 28% volume reduction were achieved with surgery and sclerotherapy respectively. Excellent, very good, and good results were obtained in 1, 3, and 4 cases respectively. Pain (2/8), edema (2/8), and distal congestion (1/8) were noted as a complication.
Conclusions: Macroglossia results from various causes and the common cause being VM. Surgery and sclerotherapy are the mainstay treatment for such a condition. They remain effective when used alone or in combination and also in a staged manner depending upon the severity of macroglossia.

Keywords: Macroglossia, Severity, Key-hole, Sclerotherapy, Surgical debulking

INTRODUCTION

Macroglossia is defined as the enlarged tongue that protrudes beyond the alveolar ridge.[1] The tongue is a vital structure for speech and swallowing. Enlargement of the tongue may hamper these functions and cause dental, muscular, and bone problems. In extreme cases, it may cause breathing difficulties during sleep. Correct identification of the underlying cause of macroglossia and choosing an appropriate treatment modality is essential for effective reduction of tongue volume and relieving the symptoms.[2]

Various classification systems had been described earlier to categorize the condition. The treatment plan also varies depending upon the aetiologies. Surgical debulking has been described as the gold standard for such conditions.[3]

The previous classifications mainly address the aetiologies, but the severity of the disease may also have a role in the surgical outcome. We hereby propose a classification system that is based on clinical findings and formulated a treatment plan based on the etiology and severity of macroglossia.

Aim and objectives

Current investigation aims to assess the treatment outcome based on this treatment protocol for macroglossia. The main objectives of the study were to determine the outcome of our treatment protocol in form of the amount of volume reduction and functional improvement.

METHODS

Current retrospective study was carried out with eight patients presenting with macroglossia between 2018 and 2020. Patient-related variables are collected from the hospital record section and analyzed. Patients with the complaint of a large tongue were examined clinically and included in the study after a provisional diagnosis of macroglossia. The patients were subjected to a color Doppler study and vascular malformation (VM) as etiology was confirmed. Tissue biopsy was obtained in unidentified etiologies. The patients were categorized into mild, moderate, and severe grade of macroglossia as following: mild; enlargement of the tongue which can be accommodated within the oral cavity. Moderate; enlargement of the tongue which can be accommodated within the oral cavity with extra effort. Severe; enlargement of the tongue which protrudes outside the oral cavity even during resting. It cannot be accommodated within the oral cavity by any means.

Based on the grades of macroglossia and etiology treatment modalities were planned. Mild and moderate forms of macroglossia with VM were treated with sclerotherapy. Patients who did not respond with three to five sittings of sclerotherapy and moderate grade were planned for surgical debulking. Severe forms of macroglossia and cases other than VM were treated by debulking surgery. The mild form of macroglossia other than VM was reassured and kept on regular follow up (Figure 1).

Sclerotherapy

The patients with mild to moderate macroglossia and VM were treated with foam sclerotherapy. Foam sclerotherapy was done by directly injecting the foam of sodium tetradecyl sulphate (STS) into the lesion. The maximum recommended dose of STS is 2 mg/kg of body weight. The subsequent doses of sclerosant were given after 3 weeks for a minimum of three sittings and more if required.

Surgical technique

The surgical reduction glossectomy was done with the "keyhole" technique. The goal was to reduce the tongue in width and length and to convert a severe grade of macroglossia into a lesser grade. The second stage debulking was done where clinical symptoms persisted. The surgery was done under general anaesthesia (GA) with nasotracheal intubation and the tongue was pulled out of the oral cavity through a 3-0 silk suture fixed to the surgical field to maintain the symmetry between the sides. A 'keyhole incision' was marked over the dorsum of the tongue with methylene blue to facilitate the planned incision. The tongue was infiltrated with 2% lignocaine with 1:200000 epinephrine. Excess tongue tissue was excised maintaining hemostasis. The tongue was closed in layers with polyglactin 3-0. The stay suture of the tongue was left for initial 24 to 48 hours to prevent tongue fall in the initial post-operative period.

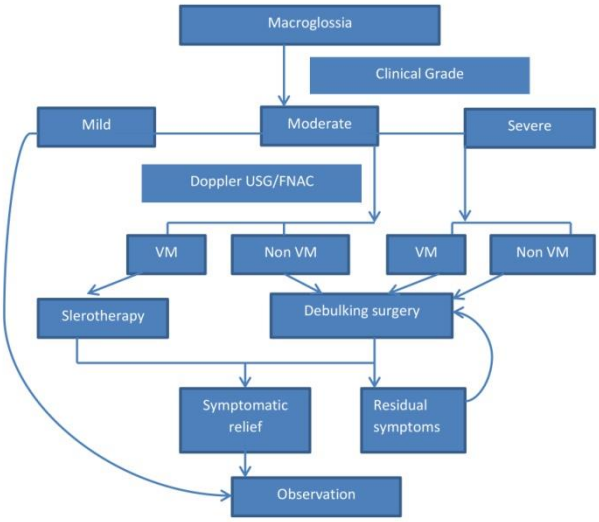

Figure 1: Schematic flow diagram showing the treatment plan based on severity grade for different etiologies of macroglossia.

Patients were followed up weekly for 6 weeks followed by every month for 6 months. During follow-up visits, patients were assessed for speech, sensory function, motor movements, and complications. The approximate volume of reduction was measured subjectively by clinical assessment and comparing serial photographs by two independent observers. The reduction is graded as excellent, good, fair, and no change. The clinical outcome is measured based on reduction of size, improvement of tongue movement, and taste sensation on a scale of 0 to 4 subjectively.

Statistical analysis

All the data were tabulated and statistical analysis was done with Graph Pad Prism version 5 software. Data are expressed as a percentage (%), mean, standard deviation (SD), and standard error of mean (SEM).

RESULTS

Eight patients (5 males and 3 females) aged 10-40 years with a mean age of 28.25 years (SD-10.29, SEM 3.63)

were included in this study. Among different etiologies in our patients, 4 were VM, 3 were neurofibroma (NF) and 1 was due to amyloidosis. Most of the patients (7/8) were moderate to severe grade as per our classification (Table 1).

Table 1: Demographic profile, classification treatment modalities and clinical outcome.

Age (years)/sex	Severity	Etiology	Treatment	Approx reduction (%)	Outcome (0-5)	Complications
25/Female	Moderate	Vascular Malformation	Sclerotherapy	20-30	Excellent	Edema, pain
10/Male	Severe	Neurofibroma	Surgery	50-60	Very Good	Edema
35/Male	Mild	Vascular Malformation	Sclerotherapy	20-30	Very Good	-
18/Male	Moderate	Vascular Malformation	Sclerotherapy	30-40	Very good	Pain
40/Female	Moderate	Amyloidosis	Surgery	50-60	Good	Distal congestion
38/Male	Severe	Neurofibroma	Surgery	60-70	Very Good	-
32/Female	Moderate	Vascular Malformation	Sclerotherapy and Surgery	50-60	Good	-
28/Male	Moderate	Neurofibroma	Surgery	50-60	Good	-

Four cases were treated with a surgical reduction only. Three patients responded after sclerotherapy and did not require any further treatment. Three, four and five sittings of sclerotherapy were required in these cases respectively. One patient required surgical reduction after three sittings of sclerotherapy. Average reductions were 58% and 28% after surgical excision and sclerotherapy as single modality respectively. Three patients with moderate to severe macroglossia were treated with surgical debulking and the outcome was good in most cases in terms of reduction in size and functional improvement. After 3 sessions of sclerotherapy, there was approximately a 30% reduction in the size of VM and hence the size of the tongue. The complications noted in our study were post-operative edema, pain, and distal congestion. Edema and distal congestion were noted in one case each after surgical debulking. Pain and edema were noted in three and one case respectively in the patient treated with sclerotherapy. All the complications subsided with conservative treatment. The duration of sclerotherapy was 17, 12, and 10 weeks respectively (mean 12.2 weeks, SD 3.30 weeks). Surgical debulking was done in a single sitting in two cases and two sitting in one case.

A 10 year old boy presented with the complaint of gradually increasing size of an tongue that was protruded outside the mouth since birth. There was difficulty in chewing, swallowing, and speech but no pain. He also complained of drooling of saliva almost all the time. On local examination, the tongue was approx 5 cm outside the mouth from the incisors teeth and was of severe grade as per our classification (Figure 2a-b). Tongue appeared to be dry and scaly. On palpation, it was non-compressible. USG of abdomen and pelvis revealed a normal study. The patient was found to be euthyroid, had no eye lesion, no bone pathology, and no café au lait spots or frecklings of the skin.

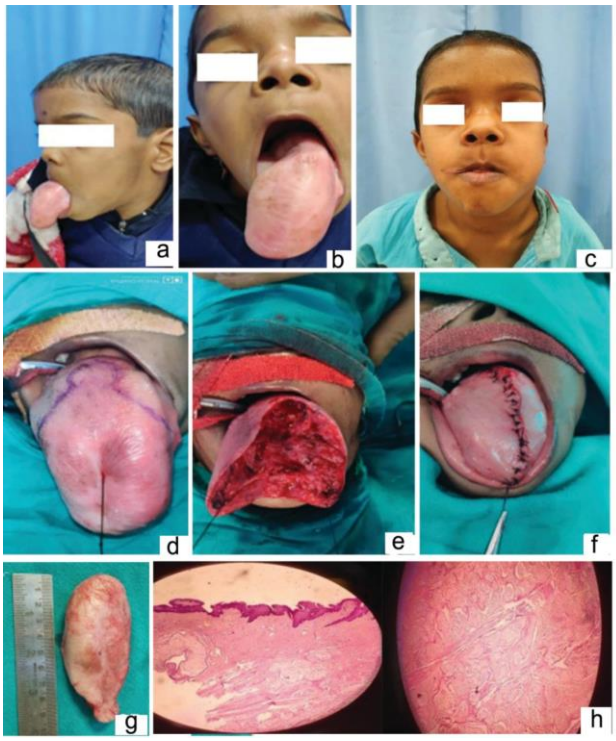

Figure 2: Case of severe macroglossia due to Plexiform Neurofibroma, treated with surgical debulking; preoperative photographs a) lateral view b) front view c) postoperative photograph after 3 weeks d) 'keyhole incision' marking e) Intraoperative photograph after debulking f) immediate postoperative photograph g) excised specimen h) appearance on histopathological examination.

The 'keyhole technique' was planned for surgical debulking (Figure 2d-f). The length and width of the tongue were reduced preserving the neurovascular bundle. The severity of macroglossia was converted to moderate grade. There was no necrosis and loss of tongue functions. The histopathology report of the excised tongue revealed isolated plexiform NF (Figure 2g-h). The patient had edema of the tongue in the immediate post-operative period which subsided after 3 weeks. He was advised mouth wash, soft bland diet, and was discharged on the 5th postoperative day (Figure 2c).

An 18 year old male patient presented with swelling over the tongue since birth which was increasing in size for the last 8 years. He also complained of intermittent bleeding on trivial trauma to the tongue. On examination, the patient had a diffuse swelling involving anterior 2/3 and dorsum of the tongue (Figure 3a). The swelling was compressible and the diagnosis was confirmed with USG. The patient was treated with foam sclerotherapy with STS. After 5 sessions of sclerotherapy at 3 weeks interval, the swelling reduced in its size approximately 30% of its initial size and improvement of symptoms (Figure 3b). The only complication noted was pain which subsided with analgesics.

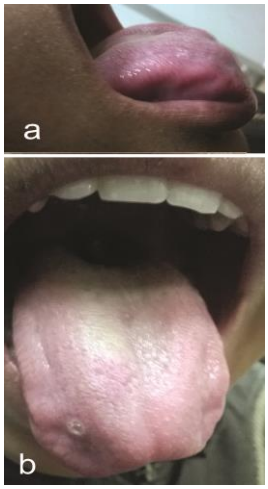

Figure 3: Moderate grade macroglossia treated with sclerotherapy, a) preoperative photo b) 3 week post-procedure photograph.

DISCUSSION

Macroglossia may be caused by muscular hypertrophy, vascular malformation, metabolic diseases, and idiopathic causes or as part of a syndrome such as Down's, Beckwith Wiedemann syndromes.[1,2] Various classifications have been proposed to describe macroglossia. According to Vogel et al macroglossia may be true macroglossia or relative.[4] True macroglossia is either congenital or acquired tongue hypertrophy. Relative macroglossia is due to a small oral cavity or neurological dysfunction causing protrusion of the tongue due to hypotonia of muscles as seen in Down's syndrome.[5] Myer et al classified macroglossia based on etiology and classified them into generalized or localized, and further divided into congenital, traumatic, inflammatory, metabolic and neoplastic groups.[6] Balaji has described an etiologic classification consisting of four categories as tissue overgrowth, tissue infiltration, relative, and inflammatory. Tissue overgrowth macroglossia is seen in Beckwith Wiedemann syndrome, hypothyroidism, chromosomal abnormalities, hemihyperplasia, mucopolysaccharidosis, whereas infiltration types are seen in lymphatic or venous malformations, hemangiomas, neoplasms, and neurofibromatosis. Relative macroglossia is seen in Downs syndrome-micrognathia, muscular hypotonia, angioedema.[7] But none of the earlier classifications has described macroglossia for therapeutic purposes.

We have proposed a classification system based on clinical examination and graded the hypertrophy for therapeutic purposes. We observed that grading the hypertrophy is essential for a rough estimation of reduction, thus the allocation of treatment modalities. Surgical debulking may reduce the maximum volume in a short period, whereas volume reduction is less in sclerotherapy and time-consuming. Moreover, response to treatment roughly depends upon the volume of reduction, thus the outcome can be co-related.

Macroglossia is of diverse etiologies and has influences on the treatment plan. VM is the most common etiology of macroglossia. Lymphangioma is commonly seen in children and presents with nodular or blister swelling. Macroglossia due to haemangioma is commonly seen in children and is known for varying levels of tongue involvement. Tongue as large as 9 cm beyond incisor has been reported.[8] These types of macroglossia are managed according to the flow characteristics. The high flow lesions are treated with embolization of feeding vessels followed by surgical reduction. Slow flow lesions are amenable to sclerotherapy, Cryosurgery, or laser. The non-compressible lesions, lesions at difficult locations and functionally compromising one often requires surgical debulking.

Other causes requiring surgical management are NF, amyloidosis, etc. Macroglossia may present as a manifestation of neurofibromatosis-I, as reported in previous literature.[9,10] But isolated neurofibromatosis involving only the tongue has been reported extremely rarely to date. We have noted in one out of three neurofibroma cases in our series. Our case illustrated this extremely rare variety of plexiform NF of the tongue in which etiology is difficult to differentiate due to lack of other clinical findings. Amyloidosis due to deposition also may rarely cause macroglossia. In these cases, the diagnosis is confirmed by fine needle aspiration cytology (FNAC) and postoperative histopathology. The treatment remains surgical debulking in these cases. Hypothyroidism may also cause macroglossia but treatment is mainly medical.

The optimum age for glossectomy has been described as between four to seven years unless there are any complications.[5] Surgical debulking is the gold-standard modality for optimum reduction of size.[3,11-13] Two types of reduction have been described, either peripheral or central. In peripheral reduction hypomotility and alteration of shape to globular appearance is common, thus central reduction is the favored approach these days. In central reduction, the shape and mobility are retained well. Reduction with a central V-shaped incision reduces the length, but the width remains the same. We adopted a modified central reduction with a 'keyhole incision' as described by Balaji.[7] The combination of a V and elliptical incision reduces both length and width with preservation of tongue margins and tip, which are important for tongue motility, taste sensation. This also maintains the shape by preventing fibrosis. Alteration of the salt, bitter taste and tongue movement have been reported with this approach.[3,13] We have not encountered such complications in our cases. The only complication seen was postoperative edema of the tongue, which subsided using antiedema measures. The cases were staged sufficient time apart to allow proper healing between the stages.

Sclerotherapy is a useful modality for VM, which remains the most frequent cause of macroglossia. The drawbacks of gold standard surgical debulking are excessive bleeding, volume loss, incomplete excision, and recurrences which may be avoided with staged sclerotherapy.[14] Though with only sclerotherapy volume reduction is less, we have noted combining sclerotherapy followed by surgery has better control over volume reduction in one case in our series. We have used STS as a sclerosant in our cases. Foam STS was prepared using the modified Tessari method.[15] No complication related to sclerosant was observed in current investigation.

The proposed classification scheme enabled us to quantify the excess tongue volume and helped us to assess the volume of reduction as well. In the study, we have noted that sclerotherapy, though did not reduce the size significantly in moderate grade macroglossia, but helped us to downstage effectively the grade of macroglossia with combined surgery in one case. The surgical reduction was most effective for severe cases with a reduction of significant volume in a single sitting. Resection to one stage down effectively helped to maintain the vascularity and minimize the complications like tip necrosis and restriction of movement. Our management scheme helped to effectively titrate the reduction, stage the procedure, and combine the modalities.

Limitations

Current study is mostly limited by small sample size due to low incidance of macroglossia. Current study thus may be used for references and further multi institutional and larger population study may be required for a definitive conclusion.

CONCLUSION

Severity based classification of macroglossia apart from etiology helps in formulating a treatment plan. Surgical debulking is the preferred modality in severe macroglossia, which can be staged if required. The multimodal treatment combining sclerotherapy and surgery is effective in macroglossia caused by slow-flow vascular malformations.

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Ethical approval: The study was approved by the Institutional Ethics Committee

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