DEVELOPMENTAL SEVERE MACROGLOSSIA. MANAGEMENT AND SURGICAL TREATMENT OF A DIFFICULT CASE.

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ABSTRACT

Objective:

Presented an extremely rare case of developmental Macroglassia in a female child with significant functional and cosmetic symptoms. The condition was worsening progressively despite persistent conservative treatment necessitating surgical management.

Case Report:

Severe macroglassia is a rare condition, usually presents as an enlarged tongue causing not only the functional symptoms like airway, speech or dietary difficulties but also presents cosmetic and social issues. Such lesions are rare but well documented in the medical literature. Our case is a female patient who was asymptomatic by birth and thereafter, but started to have progressive increase in her tongue size at the age of two, with obstructive symptoms. The patient was initially treated by conservative therapy and other alternative minimally invasive modalities of treatment. After no significant improvement over a period of almost two years, the size kept on increasing and ultimately the massive size, the cosmetic & social stress necessitates the surgical option. The macroglossia was treated successfully by surgical excision relieving all the symptoms.

Conclusion:

Different aetiologies of the condition have been mentioned in the medical literature. The treatment depends on the diagnosis, the size, the location of the tongue involved and the symptoms caused by them. If the tongue is massively enlarged and the conservative treatment fails, the surgical excision is one of the best recommended treatment options.

Keywords:

Macroglossia, Lingual tumours, Haemangioma tongue, Lingual abnormalities, Lingual Hypertrophy

INTRODUCTION

The tongue swelling or hypertrophy is commonly called macroglossia traditionally defined as a resting tongue that protrudes beyond the teeth or the alveolar ridge. Usually it is the benign tongue enlargement due to hypertrophy or growth of the lingual tissues due to a variety of causes. The exact incidence is unknown. The enlargement can be localized or else it can involve the whole or a major part of tongue as a diffuse swelling. Macroglossia can be pseudo-enlargement (displacement of the tongue created by anatomic factor other than tongue size alone) [1] or a true swelling. The enlargement can be divided as mildly, moderately or severely enlarged. Similarly the severity of the symptoms is often due to the aetiology and the volume of the tongue. Diffuse or posterior part of the tongue involvement would cause more airway and dysphagia symptoms. If the anterior two-third is involved the tongue will be pushed out of the oral cavity and makes difficult for the patient to retrieve it back into the mouth easily. So due to the constant exposure of the tongue to the environment the surface of the tongue starts drying and cracking. It further aggravates the symptoms leading to crusting or bleeding, pressure symptoms, visible deformity and even the social and psychological symptoms not only for the patient but for the family as well.

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AETIOLOGY

There are multiple aetiologies for the macroglossia. In pseudo enlargement cases the tongue size is normal but due to the associated anatomical or physiological pathologies, it is pushed to present as an enlarge size. True Macaroglossia can be classified in different ways. A classification by Meyer et al divides into generalised and localised based on the extent of involvement. He divides localised lesions into four categories the congenital, inflammatory, traumatic and neoplastic. The common congenital or developmental conditions are lymphangioma and haemangioma, lingual thyroid, Beckwith-Wiedmann Syndrome, Downs Syndrome etc.[2] The common acquired aetiologies of the macroglossia are enormous and can be divided as metabolic, inflammatory, systemic, traumatic or neoplastic. As the tongue enlarges in size its muscle bulk increase and occupies most of the oral and oropharyngeal cavities and its movements are restricted, causing airway obstruction, breathing difficulties, speech symptoms and dysphagia of various intensities. As the child grows, the size of the tongue hinders normal facial growth causing mandibular and maxillofacial deformities resulting in open bite deformities and dental malocclusion.

CASE REPORT

We presented a case of developmental macrophagia in two year old female village girl, a daughter of a poor peasant, who was born normal with normal breathing, feeding and eating habits. At the age of two she developed a progressive increase in her tongue size. Initial physical examination revealed a normal child with mild increase in the tongue size, with normal oral cavity, with drooling and minimal speech and swallowing difficulties with no associated head & neck or systemic symptoms. The rest of the otolaryngological & head and neck examination was unremarkable. The birth history, past medical history were clear and no family history of any congenital cause. As the tongue size and protrusion kept increasing over a period of two years, she also started to have progressive increase in her obstructive symptoms. Initially started to have difficulty in speech (like a hot potato in the mouth) and mild dysphagia to solids only but gradually it progressed to liquids as well due to the massive bulk of the tongue.

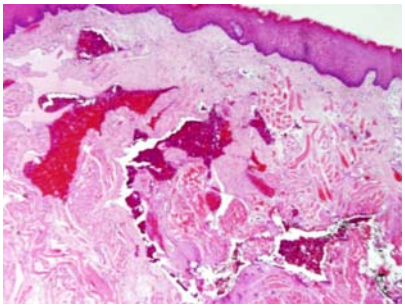
As a result she had to use only liquid and pure diet, because opening mouth and mastication was not so easy. The diffuse tongue enlargement mainly involved the anterior two-third. It filled the oral cavity and the main bulk of the tongue was always protruding out of dental line. The oropharynx was not so obstructed and never threatened the airway seriously throughout her management, because the huge extra-oral part of the tongue prevented the posterior tongue to slip back. The massive size caused a gross deformity of her mandible in the form of open bite, malocclusion and a persistently large protruding tongue mass, which was not retrievable back into the oral cavity causing constant drooling. Due to the specific local health system, the poor family was under severe financial constrains. Clinically the immense psychological stress was also very obvious on the patient and her family. She was too shy to face the general public with her deformity and most of the times kept her lower face covered.

MANAGEMENT

All routine lab investigations including complete blood count, LFTs, renal function tests, thyroid function tests, were normal. ECG was normal. X-ray chest and lateral soft tissue neck were normal and revealed the posterior one-third of the tongue minimally enlarged, with normal naso-oropharynx and normal upper and lower airways. Thyroid scan was normal. CT scan revealed normal head and neck, with enlargement of the anterior two-third of the tongue with diffuse enlargement of the soft tissue mass query consistent with fibro-haemangioma of the tongue. The fine needle aspiration revealed a lymphangioma.

Initially the patient was treated medically with poor outcome. Repeated controlled periodic sclerosing agents (Ethenolamine Oleate, steroids) were locally injected in the tongue under general anaesthesia with only temporarily improvement initially. After regular follow ups and repeated sittings

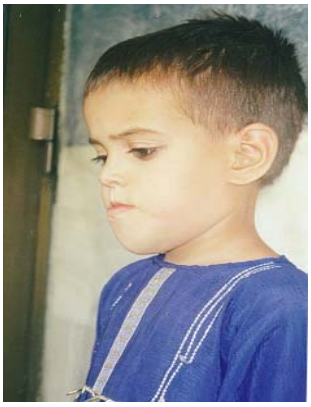
Sever Macroglossia / Obstruction / Cosmetic deformity



High Power view (H& E)
The Lesional Blood Vessels are seen
deep within Skeletal
Ms Fibers of the Tongue

Preparation for Sclerotic
Injection under GA

Immediate Post Op



Late Post Op

of sclerosing agents the results were still not promising. She was further followed up and was treated with two sessions of local cryotherapy to the affected parts of the tongue as an alternative modality of treatment but again with no significant improvement. The condition kept progressing with each follow up and no improvement was noted in the progression of the disease process. During the late visits, the family was found to be under extreme economical, psychological and social stress. All social worker's efforts and counselling seemed to be ineffective. As a result of all these developments, a partial reduction glossectomy was planned. After complete work up and informed consent, the surgery was performed under general anaesthesia with nasotracheal intubation, as the family did not agree for the tracheostomy. Of course the difficulty airways were anticipated, but due to the preoperative detailed assessment and radiological assistance the anaesthesiologist were successful to introduce controlled nasotracheal tube. Complete tongue and oral cavity measurements were taken preoperatively to safeguard the neurovascular bundle as much as possible. A triangular wedge of the anterior half of the tongue was resected and the two lateral flaps were stitched together developing a new tip of the tongue. Due to the previous sclerotherapy and cryotherapy the bleeding was much less than expected. The surgery and the post operative intensive care period were uneventful and the overall outcome was a pleasant surprise for the health care team, the family and of course for the young patient herself.

RESULTS AND ANALYSIS

During the immediate post operative period, the patient and the family was exited, but there was an obvious mandibular deformity resulting in sever open bite deformity and malocclusion Fig xi). Maxillofacial consultations were sought and wait-and-see policy was adopted with minimal biting exercises. Over a period of three month the deformity was almost completely corrected the bite and occlusions were satisfactory. The patient was followed up for seventeen months with full improvement, and then lost for follow ups.

HISTOLOGY

Histological studies of the tongue specimen revealed diffuse angioma of the tongue. The stroma had numerous dilated blood vessels. The immune histological studied confirmed the diagnosis and C.

Immunostain confirmed the histopathology, with no malignant cells seen.

DISCUSSION

This condition is well documented in the literature. The benign oral masses as a whole are studied by different authors at different times concerning the incidence, aetiology, frequency, nature and distribution. In a large recent study the non neoplastic oral lesions were 96% of the total. Benign lesions include traumatic, inflammatory, infective, cystic and developmental. The developmental lesions were only 9% of the total. According to the author and others, the pathogenesis of vascular lesions of the oral cavity in general and the tongue lesions in specific are debatable, whither they are developmental malformations or hamartomas of blood vessels.[3] In another recent large series of 2161 cases of oral and maxillofacial cases combined, 59% were claimed to be benign, 33% were malignant and 8% were tumours like leisions.[4] Macroglossia unlike other oral cavity benign lesions, when present can cause a number of functional, aesthetic and psychological issues for the patient himself and for the family as well. The treatment of this unusual pathology is challenging and controvercial.[5]

In addition to the above mentioned symptoms occasionally the rare symptoms like traumatic bleeding are severe enough to threaten the airway and the patient life.[6]

The management starts with the gold standard history and physical examinations, laboratory and radiological investigations. CT and MRI together or alone are considered to be valuable preoperative investigation tools. Some claim MRI to be the ideal tool for vascular malformations.[7] In a recent case report three dimensional CT angiography's role is highlighted in the treatment of macroglossia with large venous malformations.[8] Treatment options in-

clude, observation, orofacial therapy, medical therapy and surgical therapy. Even in moderate or severely enlarged macroglossia there is a wide spread consensus that the treatment should be specific according to the aetiology. The orofacial therapy uses a palatal device to stimulate the muscular tone and the proper tongue position. The right medical and orthodontic therapy should be considered first and the surgical treatment should only be adopted when the medical therapy fails. Also the surgical options much be selected in accordance with the functional results desired and must be the most conservative technique to preserve the neurovascular bundles as much as possible.[9]

Regarding the modalities of the treatment, in addition to the specific medical therapy the other medical options are limited. Corticosteroid can be life saving both in acute airway obstruction and are useful postoperatively to reduce oedema. Sclerotherapy is agreed upon by the majority. The aim of the sclerotherapy is targeted elimination and damages the vessel walls and transforms it into a fibrous cord that can not be recanalized. It is simple, cost effective and aesthetically acceptable modality for both therapeutic and aesthetic purpose.[10] The type of sclerosing agents used are controversial and should be selected according to the individual cases. The steroid or sclerotherapy injections are always painful, so the analgesics are used to minimise the pain. The types of analgesia used are the personal choice and usually the conventional analgesics are used, but on occasions other options have been claimed beneficial, like 24% hypertonic sucrose solution given via needleless syringe to the anterior tip of the tongue in combination with a pacifier as an analgesic during intralesional injection of infantine hemangioma.[11] *The* indication of surgery are severe macroglossia, airway obstruction, speech difficulties, dysphagia and cosmetics. The goals of the surgery are to reduce the tongue size and to improve the functions. A variety of surgical approaches have been mentioned. Almost all modalities of surgeries are performed under general anaesthesia. Due to the protruding large size of the tongue difficult airway is a major issue in these cases, especially in paediatric population with diffuse macroglossia due to lymphangiomas or hemangiomas. Therefore preoperative tracheostomy is a safe and recommended option in these cases.[12] Pediatric hemangiomas is rare in the oral cavity. Among the most frequent location are the lip, followed by tongue and the treatment of choice is surgical excision of the leision.[13]

The most practical surgical options used for these cases are reduction partial glossectomy. The fundamentals of the surgical reduction are to preserve the neurovascular supply to the remaining intrinsic tongue muscles to safeguard the long term impact on airway, swallowing, speech and articulation.[14] The common surgical techniques discussed in the literature are the wedge resection, the stellate and double stellate tongue reduction[15], the modified key hole technique[16], Kole glossectomy,[17] Grabb glossectomy, Dingman glossectomy and Jian glossectomy.[18] The other rare surgical options are laser glossectomy[19], Hormonal scalpel surgery[20], submucosal minimal invasive techniques and combination embolization-surgery[21] etc. The submucosal resection is a relatively recent minimal invasive technique in which the endoscopic surgery is used and incorporates coblation method with ultrasonography for children who need tongue base reduction.[22]

CONCLUSION

Macroglossia is a rare benign oral cavity lesion with a diverse aetiology. Heamangiomas usually develop in the paediatric population but lingual hemangiomas are even rare. Macroglossia causes pressure symptoms and aesthetic, psychological and social problems for the patient and the family. The medical treatment should be tried first and if they fail the surgical option should be adopted. The surgery in each case must be evaluated with extreme caution and planning and the most conservative therapy to save the neurovascular supply should be adopted. Post surgical conservation of the normal physiology and cosmetics should be the aim of surgery. Post operative appropriate rehabilitation and long term follow up is needed for successful outcome.

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