

DEBILITATING LYMPHANGIOMATOUS MACROGLOSSIA-CASE REPORT.

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Abstract

Objective; Lymphangiomatous macroglossia in children though uncommon may cause significant morbidity which maybe functional or developmental when untreated and also presents a significant surgical challenge when trying to balance complete surgical excision and maintenance of contour and function.

Case report; We report a case of extensive tongue lymphangioma in which surgical excision was performed with good cosmetic and functional results.

Conclusion; Lymphangiomatous macroglossia in children is a debilitating condition not only due to the physical problems it causes but also the attendant psychological issues that arise. Prompt treatment is required to improve the patient's physical and psychosocial condition of Nairobi, Kenya

Keyword; Lymphangiomatous macroglossia

Introduction

Lymphangiomas are considered to be benign congenital tumours of the lymphatic system¹. They exhibit no sex predilection and are usually congenital. Upto 90% of all lymphangiomas are evident by 2 years of age. These most commonly involve the head and neck² but may also occur anywhere and present as a painless, soft mass.

As they enlarge they may compress on vital structures resulting in dysphagia, dyspnoea or stridor³. The initial appearance of lymphangioma in adulthood is less common. The growth rate of this neoplasm is variable. Slow progression followed by pseudoregression eventually gives rise to reappearance. Rapid growth or engorgement with lymph or blood is often associated with direct infection, trauma or a secondary respiratory or skin infection. Spontaneous regression has been reported in as many as 15% of patients with cervicofacial lymphangioma.

Malignant change has not been reported. Important to note is that they are occasionally associated with several congenital syndromes

and developmental deformities such as turners syndrome, down's syndrome, congenital heart abnormalities and hand and foot deformities^{4, 5, 6, 7}.

Whether patients with this neoplasm present in childhood or adulthood, the identification of a mass requires a diagnosis and management plan. Management may be complex and difficult because enlargement often results in functional and cosmetic problems.

Attempts at a classification have been created to differentiate between the capillary, cystic, and cavernous forms of cervicofacial lymphangioma⁵; however, the histologic appearance is not always uniform, which suggests a variable clinical and pathologic appearance of essentially the same disease. Determining location, persistence, growth patterns, and functional and cosmetic problems associated with lymphangioma usually requires a full diagnostic evaluation.

We report a case of extensive tongue lymphangioma in which surgical excision was performed

with good cosmetic and functional results. In September 2012, a 4 year old Kenyan girl presented at the facio-maxillary trauma and reconstructive surgery clinic, Nairobi hospital, with complaints of tongue swelling that had been present since birth and was now associated with speech, breathing and eating difficulties due to recent significant increase in size.

Clinical examination revealed an enlarged right sided tongue swelling with associated derangement in the development of the maxilla and mandible (figure 1A and 1B). Of note was that the tongue could not



Figure 1A: Oral appearance

be withdrawn into the mouth with attendant saliva drooling.

Taste sensation was intact. There were no other associated anomalies. Magnetic resonance imaging (MRI) revealed an intraglossal lesion centered on the right side of the tongue crossing the midline



Figure 1B: Patient at initial presentation

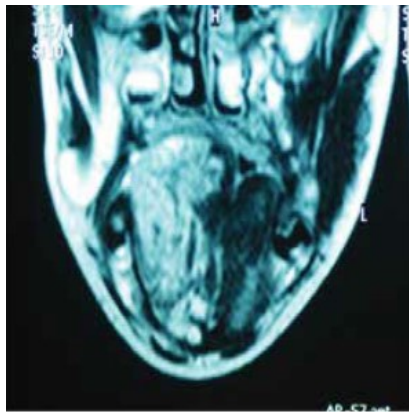


Figure 2A: Coronal view

invading the sublingual and right submandibular fascial spaces with associated left lateral displacement of the oropharynx (figure 2). Also noted was adenoid hypertrophy causing moderate to severe nasopharyngeal narrowing. Histopathologic evaluation showed a tumorous lesion consisting of mature skeletal, fibrous, vascular and peripheral nerve fibre components with no signs of atypia, dysplasia or malignancy.

Based on the above a diagnosis of glosso-pharyngeal lymphangioma was made. Given the extent of the lesion and character of its growth, a management plan was formulated comprising (figure 3).

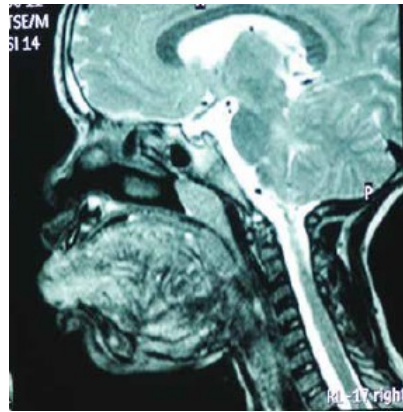
Immediate: Conservative surgery under general anaesthesia:

- ◆ Reduction glossectomy with

reconstruction of the anterior tongue with a rotation flap from the left side.

- ◆ Posterior glosso-plasty. Post operative recovery in HDU with particular attention to airway control.

Secondary: post puberty Glosso-pharyngeal reassessment +/- or-



2B: Sagittal view

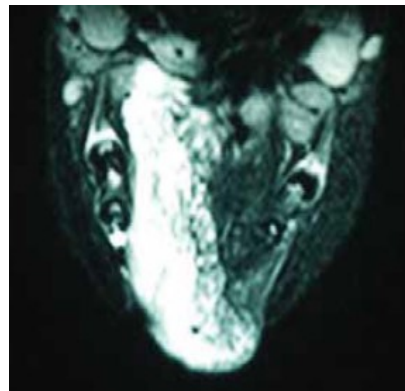


Figure 2: MRI scans of glosso-pharyngeal lesion

thodontic/Orthognathic surgery. Surgical intervention was undertaken successfully with an uneventful post-operative recovery. 3 weeks and 4 months postoperatively, the patients tongue appeared normal morphologically and was located within the oral cavity (figure 4). However she has residual maxilla mandibular deformity (anterior and lateral open bite) that will be treated at a later date.

Discussion

Lymphatic malformation of the tongue often result in macroglossia whose sign and symptoms may vary. In children, lymphangi-

omatous macroglossia is uncommon but may cause significant morbidity which may be functional or developmental. Furthermore, children may develop psychosocial issues due to incessant teasing and bullying brought about by their condition.

The most obvious symptom is tongue protrusion which may have attendant functional effects such as feeding difficulties and speech impediments. In addition to these, developmental abnormalities like maxillo-mandibular deformities may arise due to imbalance in the facial functional matrix⁸. In long standing cases, systemic effects such as failure to thrive, obstruct-



3A: Enlarged tongue



3B: Surgical site marking



3C: Immediate post-operative photo

tive sleep apnoea and in some cases airway obstruction and death, may occur ³. Our patient present-

ed with tongue protrusion, failure to thrive due to feeding difficulty and obstructive sleep apnoea which necessitated urgent intervention.

The management principles of lymphangiomas encompass accurate diagnosis through clinical history of evolution of the condition and histopathologic evaluation. MRI offers the best possible tool for demarcation of this infiltrative lesion within the soft tissues. Finally, definitive treatment encompasses either surgical resection, radiation therapy, cryotherapy, electrocautery, sclerotherapy, steroid administration, embolization,



3D: Adequate tongue reduction



3E: Excised specimen



Before



3 weeks post-operative



4 months post-operative

gation and laser surgery¹.

Overall, surgical excision is the treatment of choice³. In children, surgery is preferred between ages of 3-7 years (spontaneous regression viz-a-viz growth spurt before anterior teeth have been replaced by permanent dentition)⁹. Surgical treatment varies though generally it has been subdivided into two groups: central glossectomy and peripheral glossectomy depending on the resection sites and the sutural margins^{10,11}. Variations including V-shaped wedge resection in which the entire thickness of the tip of the tongue is removed back to the level of the circumvallate papillae, bilateral marginal resection, and U-shaped resection with the open end of the incision facing posteriorly^{12, 13, 14, 15} have been described. Regardless of the method used, the recurrence rate is high due to the non-encapsulated and 'infiltrating' nature and as such there is often a need for further surgical procedures³.

However, the current case presented with a unilateral extensive tongue lesion that crossed them-

li- idline that could not be approached through the above means. Conservative reduction glossectomy with posterior tongue glossoptasty was chosen in our case to achieve macroscopic tumour removal with maintenance of normal tongue contour and function. This highlights some of the difficulty one may face with due to the varying presentation.

Important to note is that surgical principles must be adhered in terms of protection of normal tissue (neurovascular structures, muscular elements) to maintain critical tongue functions post-resection. It cannot be overemphasized on the need for careful long term follow-up not only to assess for recurrence of the lymphangioma but also to address the dentofacial functional and morphologic deformities which may persist and as such require protracted treatment involving orthodontics and orthognathic surgery.

Conclusion

Lymphangiomatous macroglossia in children is a debilitating condition not only due to the physical problems it causes but also the attendant psychological issues that arise. Prompt treatment is required to improve the patients physical and psychosocial condition.

Consent: Informed consent was obtained from the patient's guardians for the publication of the case report.

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Declaration: The authors declare no competing interests and submit that the above case report has not been presented to any other journal for publication. Further, the authors surrender copyright of the article to the journal.

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