A 8-years old girl presented with huge tongue, drooling of saliva, difficulty in swallowing, mastication, dysarthria and anterior open bite

Asaduzzaman, Md Wares Uddin, Helal Uddin, Nazia Mehanaz, Suvasish Das, Faria Tabassum Tanni

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Department of Oral and maxillofacial surgery, BSMMU, Dhaka (A, MWU); Department of Orthodontics, BSMMU, Dhaka (HU, FTT); Department of Pedodontics, BSMMU, Dhaka (NM, SD)

For Correspondence:
Asaduzzaman
asaduzzamanomfs@lbsmmu.edu.bd

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Presentation of case

Dr. Kaniz Fatima Munny (MS Resident): A-8-years old girl hailing from Bagerhat, Bangladesh was admitted in our department with complaint of huge tongue, drooling of saliva, difficulty in swallowing, difficulty in mastication, dysarthria and anterior open bite for last 3 years. Her father gave history that she was apparently normal up to two months after birth then a growth was observed on the dorsal surface of her tongue. Initially the growth was small but gradually become large and interfering with closing of mouth, mastication, swallowing and speech. Her father did not give any relevant family history. On inspection, the swelling involved the whole tongue causing protrution from mouth. Tip of the tongue had multiple pebble like vesicles which also spreaded to dorsum surface. Right lateral border of tongue appeared blue and shiny (Figure - 1). On palpation, the measurement of protruding tongue from upper incisor was almost 9 cm. Tongue was soft pebbly and non-tender in nature. Tongue movement and mouth opening was normal but there were anterior open bites. There were difficulty in talking, eating, drooling of saliva but no dyspnea or respiratory distress. On general examination patients all parameter are within normal limit but mentally depressed. Laboratory data showed CBC within normal limit (Table - I), MRI of tongue showed macroglossia but no arteriovenous malformation, FNAC from lesion showed mature lymphocyte with proteinaceous background. Histopathological exam from lesion showed dilated lymphatic channel. (Table - II)

Provisional diagnosis: Macroglossia due to vascular malformation

Differential diagnosis

Dr. Nazia Mehanaz (*Assistant Professor*): Vascular malformation is a generalized term which encompassess a group of lesions. They mainly present at birth, by forming an anomaly of angiovascular or lymphovascular structures. 1% of births may present vascular malformations and majority of these patients do not present for treatment.[1] When vascular malformation involved in tongue, they may cause macroglossia**.** As in our case, the type of macroglossia is a localized form and not related to any syndrome; differential diagnosis of macroglossia should include local congenital anomalies like lymphatic malformation, arteriovenous malformation, hemangioma, congenital cyst etc.

Lymphatic malformation:

In 1854 Virchow First described that Lymphangioma is a rare, benign, congenital disease of unknown etiology that originates from lymph vessels.[2] In our case, lymphatic malformation may cause macroglossia.

Arteriovenous malformation: Though arteriovenous malformation usually presents at birth but manifest in childhood or adolescence. This malformation can occur at any part of body. But in the oral cavity, most commonly occur on anterior two-thirds of the tongue, palate, and gingival and buccal mucosa.[3]

Hemangioma: Four major type of pediatric vascular tumors are usually benign and consist of infantile hemangioma, congenital hemangioma, kaposiform hemangioendothelioma (KHE) and gynogenic granuloma. They may involve anywhere in the body and present during infancy or childhood. They can cause both local complications (bleeding, destruction of tissue, obstruction, and pain) and systemic problems (thrombo- cytopenia, congestive heart failure, and death).[4] Hemangioma of tongue also may cause macroglossia.

Congenital cyst: Congenital tongue base cysts are uncommon and may cause airway obstruction. An approximate annual incidence of 1.82 per 100000 live births in an oriental population was reported. [5] This may elevate tongue and cause macroglossia.

Table-I

CBC findings of the case

Name of the investigation	Results and comments	Reference
Hb%	14gm/dl	15 ± 2 gm/dl
ESR	15mm in 1st hour	0-20 mm/1st hr
WBC	7.1×10^9 /L	7.0 ± 3.0 × 10^9 /µL
RBC	5.1 × 10^12/L	5 ± 0.5 × 10^12/L
Platelet	350× 10^12/L	150-450 × 10^12/µL
Neutrophils	72%	40-80 %
Lymphocyte	21%	20-40 %
Monocytes	4%	02-10%
Eosinophils	3%	01-06%
PCV	PCV: 47.8%	45 ± 05%
MCV	MCV: 97.5 fl	92 ± 9 fl
MCH	MCH: 30.6 pg	29.5 ± 2.5 pg
MCHC	MCHC: 33.1 g/dl.	33 ± 1.5 gm/dl

Table-II

MRI, FNAC and histopathological findings of the case.

Name of the investigation	Results and comments
MRI	Macroglossia of tongue but no vascular malformation
FNAC	Cholesterol crystals, lymphoid cells mainly small lymphocytes in variable number and endothelial cells are present.
Histopathological examination	Mature lymphocyte with proteinaceous background
	Dilated lymphatic channel.

Diagnosis: Dr. Faria Tabassum Tanni: Macroglossia due to lymphatic malformation.

Regarding diagnosis of the case

Dr. Helal Uddin (Assistant professor): Patient needs more investigations for evaluation of general condition like chest X-ray, serum creatinine, liver function test, coagulation profile and specific investigations were performed for the tumor itself like FNAC followed by incisional biopsy with histopathological examination and MRI. Firstly MRI was done to evaluate any vascular communication then FNAC was done for cytological evaluation from the lesion which was in favor of

Figure 1 : Show pre-operative (a) frontal view (b) lateral view

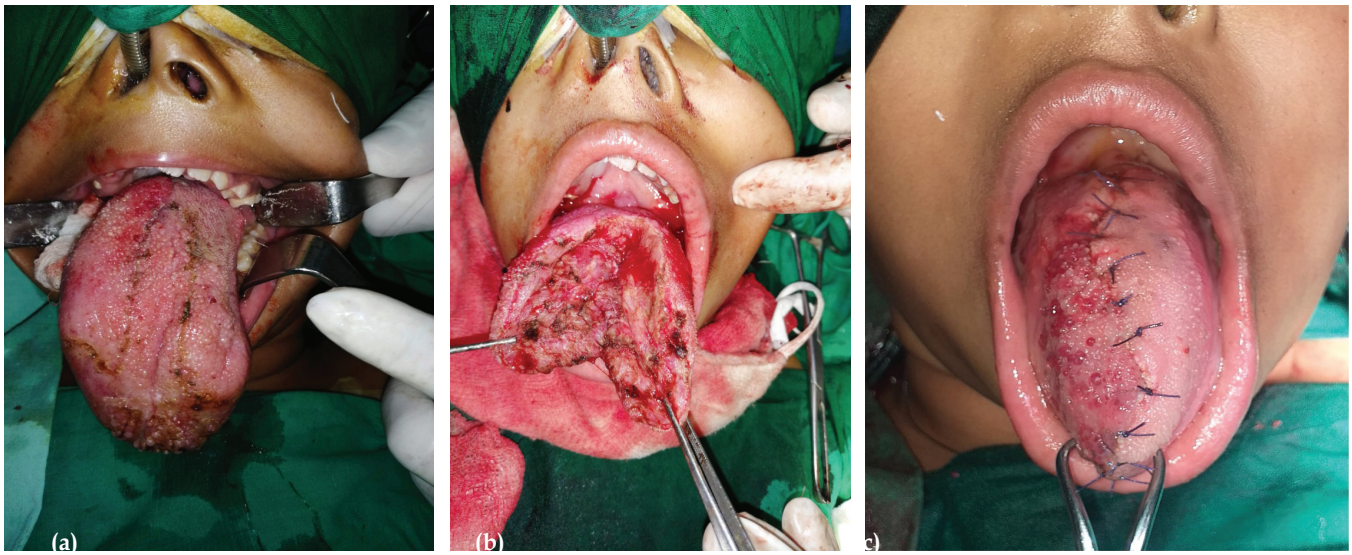

Figure 2 show intra-operative (a) excisional marking (b) modified keyhole subtotal glossectomy (c) Final closure

lymphatic malformation. Finally incisional biopsy was taken from the lesion to confirm diagnosis. The final report was lymphatic malformation.

Discussion

Dr. Md. Wares Uddin (Professor): Balaji[6] stated that "Macroglossia is a generalized term used to describe the tongue that protrudes beyond the teeth during natural resting posture. It creates an artificial, persistent, impression of teeth on lateral borders of the tongue when the patients slightly open their mouth". In the present case, the lesion was involved whole of the tongue causing protruding from mouth. Most commonly, the usual time of occurring of lymphatic malformation in the oral cavity is the first decades of life and usually occurs in the tip, dorsal and lateral border of tongue. Sometimes it may arise from lip, gingiva, buccal mucosa, palate and base of tongue. [7] One of the common causes of macroglossia is lymphangioma of anterior two-thirds of tongue.[8]

Clinically the lesion varies its appearance according to location whether is it deep or superficial. When the lesion is superficial, it presents as papillary lesion with pebbly surface due to translucent vesicles. The vesicles may be same color of adjacent mucosa or may be of mild reddish hue or sometimes may be given tapioca pudding or frog eggs like appearance.[9] But when the lesion is deeper it presents as diffuse nodules with soft consistency and negligible alteration of color or texture. [10] The progression of deep seated lesion may cause extrusion of tongue, inability to close the mouth, failure to chew, increase salivation, jaw deformity, speech problem, poor oral hygiene and sometimes airway obstruction.[11]

In this case, the patient had papillary lesion with pebbly surface, extrusion of tongue, inability to close the mouth, failure to chew, increase salivation, jaw deformity, speech problem, and poor oral hygiene. So this case had both superficial and deep lesion. Superficial lesion mainly present in the tip of tongue and deep lesion present within anterior two-thirds of tongue.

Regarding treatment plan

Dr. Asaduzzaman (*Assistant Professor*): It was suggested that surgery is the primary treatment of this type of malformation. Usually the name of surgical procedure is tongue reduction surgery. The objectives of treatment of lymphangiomatous macroglossia include stoppage of drooling of saliva, restoration of articulation preservation of taste, mandibular and dental deformities correction, and esthetic care.[12]

No single ideal tongue resection procedure has been documented in the literature. Considering the developmental aspect, etiology, age, gender, existing dimension, the procedure have to be customized. [13] The primitive surgical designs were to reduce the extensions of the tongue in terms of length and width. Peripheral trimming and reduction techniques were aimed at reducing the length and the width of the tongue. Modification of Kole's incision, Davalbhatka, and Heggies were introducing other techniques that aimed at preserving the function at the same time reducing the bulk of the tongue in all the three dimensions. This procedure is called key-hole procedure. Where anterior two-thirds of tongue is enlarged by involving disease, tip of tongue sacrifice shall be considered. Patient should be informed regarding the postoperative loss of taste sensation when the tip of the tongue has to be excised. [6]

In our case we used modified key-hole sub-total glossectomy to reduce the tongue three dimensionally. As the anterior two-thirds of tongue were elongated, sacrifice of the tip of tongue was done with properly informed of postoperative complication.

Regarding surgical procedure:

Dr. Asaduzzaman (Assistant Professor): The surgery was done under general anesthesia. With all aseptic preparation under general anesthesia draping was done. Marking of tongue with key-hole shape on the dorsum of tongue was done (Figure: 2a). Local anesthesia was infiltrated along the incision line. Incision of mucosa was done with scalpel. Carefully identify and dissection of muscle was done with diathermy for better control of bleeding (Figure: 2b). Closing layer by layer with 3/0 vicryl (Figure: 2c).

Follow up:

Dr. Suvasish Das (*Assistant Professor*): Patient was followed- at an interval of 3 months. History of presenting complains were evaluated. Function and esthetic of tongue were also evaluated. Her complaints of huge tongue, drooling of saliva, difficulty of swallowing, difficulty of mastication, dysarthria and anterior open bite all were solved (Fig: 3).

Final diagnosis: Macroglossia due to lymphatic malformation

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