

Case report

Cavernous hemangioma presented with gigantic tongue

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Abstract:

Lingual hemangioma is a benign congenital vascular tumour that may cause macroglossia. It can present with airway obstruction, speech disturbance, swallowing difficulties and mandible dental deformities. It usually occurred during childhood. Females are more affected. It usually poses difficulties in diagnosis and management. This case illustrated a rare case of lingual hemangioma in a 5 year old boy presented with tongue enlargement associated with swallowing difficulties and significant tongue bleeding which required blood transfusion. Surgical intervention in the form of tongue reduction surgery was successfully done to him. Histopathological examination revealed a cavernous hemangioma.

Keywords: cavernous hemangioma; recurrent bleeding; tongue reduction surgery

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Introduction:

Hemangioma is a common case occurring in childhood and adult. However lingual hemangioma that present with a gigantic tongue is rarely encountered. It can often be found in the skin and mucosa of the head and neck region, with involvement of the oral cavity including lips, buccal mucosa, or tongue. Hemangioma in the oral cavity causes discomfort and potentially serious clinical problems. This lesion in the tongue can cause particular problems such as recurrent hemorrhage, biting of the lesion, pain and difficulty with speaking, mastication or deglutition. Being a mobile structure organ, tongue hemangioma may cause difficult problem to the patient because it is more prone to trauma and lead to further complications such excessive bleeding. Either scenario may happened in which it may remain indolent or it may produce obstructive symptoms or alarming hemorrhage.

Case Summary:

A 5 year old Malay boy presented with recurrent tongue bleeding associated with difficulty in swallowing, speech difficulty and mild shortness of

breath for 5 days prior to admission. The bleeding was moderate in amount and persistent. On examination, patient was pale (Hb 7.9gdL), there was huge tongue mass protruding out of the mouth, purplish red in colour. There were multiple hemangiomas spots with crusting and area of blackish mucosa (Figure 1). The tongue were fully occupied the entire interdental space, along with the upper alveolar ridge, leaving a groove on the hemangiomas tongue surface. Patient was unable to close his mouth due to the obstructing mass. The nasopharynx and oropharynx were normal. There was diffuse soft right neck swelling extending to right submandibular area. Patient was transfused with 2 pints of packed cell in view of severe anemia. The bleeding was controlled by manual compression.

Previous medical history revealed that the patient had undergone tracheostomy and excision surgery for cystic hygroma with upper airway obstruction at 14 days of life. At present, there was no evidence of recurrence. He was still on tracheostomy tube.

MRI showed hemolymphatic malformation and diffuse hemangiomas involvement of the tongue

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Figure 1: Hemangiomatous tongue before tongue reduction surgery



Figure 2: MRI saggital view showed the hemolymphatic malformation and diffuse hemangiomatous involvement at the base the tongue and pushed tongue anteriorly

with extension to the bilateral supraclavicular and left superior mediastinal area. Oropharynx and hypopharynx were almost totally occluded (Figure 2). In view of difficulty in swallowing and the risk of bleeding, surgical intervention was planned. Tongue reduction surgery was performed. The anterior part of the hemangiomatous tongue mass was excised (Figures 3 A&B).



Figure 3: Surgical marking for the incision site (A) & subcutaneous tissue undermined and partially excised (B)

No significant intraoperative heamorrhage and overall operative procedure was uneventful. Protruding tongue mass successfully removed (Figure 4).



Figure 4: Tongue hemangioma after tongue reduction surgery

Gross examination of the specimen showed fleshy tissue with clots on cross section. Postoperative period was uneventful. Histopathological examination of the excised tongue tissue showed numerous dilated vascular channels (some with muscular wall) in a loose fibrous stroma, interspersed between skeletal muscle bundles in keeping with cavernous hemangioma (Figure 5).

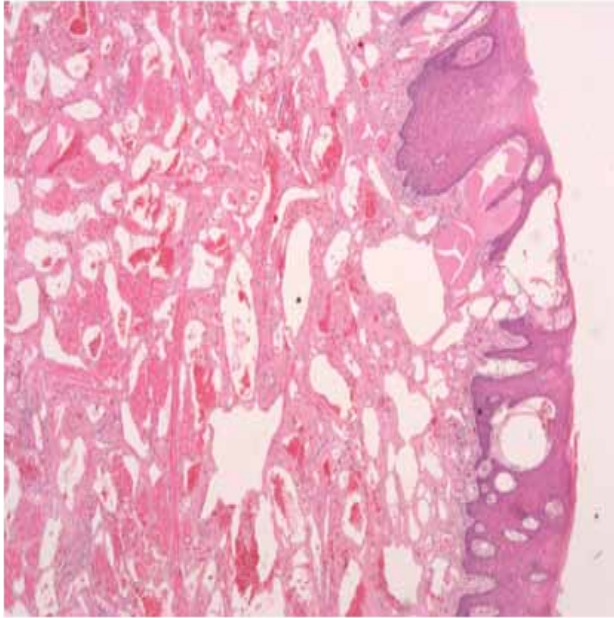


Figure 5: Numerous dilated vascular channels (some with muscular wall) in a loose fibrous stroma, interspersed between skeletal muscle bundles in keeping with cavernous hemangioma.

There was involvement of resection margin. However there was no evidence of malignancy. After 8 months follow up there was no recurrence. He was able to take orally and no recurrent tongue bleeding.

Discussion:

Hemangioma is a common benign vascular tumor in childhood^{1,2}. There is female predilection with ratio female to male is 2-3: 1³. Histologically there are classified into capillary, cavernous and miscellaneous forms⁴. Capillary hemangiomas further classified into juvenile, pyogenic granuloma, and epithelioid hemangioma⁴. Capillary hemangioma is composed of many small capillaries lined by a single layer of endothelial cells supported in a connective tissue stroma of varying density. Cavernous hemangioma is formed by large; thin walled vessels, or sinusoids lined by epithelial cells separated by thin layer of connective tissue septa⁵.

The majority of hemangioma involves the head

and neck region. However, they are rare in the oral cavity but may occur on tongue, lips, buccal mucosa, gingiva, palatal mucosa, salivary glands, alveolar ridge, and jaw bones^{3,4}. Clinically, hemangioma appears as soft mass, smooth or lobulated, and sessile or pedunculated and may vary in size from a few millimeters to several centimeters⁵. It is very uncommon to see such hemangiomatous gigantic tongue.

Pathogenesis of hemangioma remains incompletely understood. Various theories have been proposed. First, an aberrant and focal proliferation of endothelial cells, although the underlying cause remains unclear⁶. Second, placental theory of hemangioma, as various histology and molecular markers such as GLUT 1, Lewis Y Antigen, Merosin, CCR6, CD15, IDO, FC, and gamma Receptor II had been studied⁷. GLUT 1 positive is considered highly specific and diagnostic, and it is useful for making differential diagnosis between hemangioma and other vascular lesions⁷. The third theory is the somatic mutational events in gene involved in angiogenesis³. Growth factors specifically involved in angiogenesis such as VEGF, b-TGF, and IGF are often increased during the proliferation phases of hemangioma growth^{8,9}.

This patient presented with huge tongue mass and persistent bleeding which require blood replacement. Although it is a rare presentation, it poses a life threatening condition to the patient as airway may be jeopardized. Persistent bleeding may end up with significant blood loss and may lead to subsequent complication such as hypovolemic shock. Apart from that, there is also high risk of aspiration. If further oedema occurs secondary to trauma, it will further swell and obstruction to the food passage and airway embarrassment could not be avoided. The lymphangiomatous component at base of tongue was shown to be originated from the base of tongue, as shown by the MRI. This made the tongue was pushed forward and appeared as a gigantic tongue.

Treatment wise can be categorized into medical and surgical intervention. Medical intervention includes systemic or intralesional corticosteroid¹⁰. Among the medically treated cases, only 30% respond to the therapy. If medical treatment failed, surgical intervention is an option. Surgical resection can be offered with or without pre operative embolization. In selected cases embolization alone can achieve the desired outcome^{11,12}. However, in this case pre embolization was not carried out to the patient because

it was not feasible in our center. However if pre embolization is really needed to do in selected cases, referral to appropriate center will be made. Resection can be employed to remove the hemangomatous tissue but to leave as much as possible normal tongue tissue including tongue tip for the functional purpose. An important note is that surgery to the highly vascularized lesion such as in tongue hemangioma is not without complication. We must be cautious and anticipate the profuse bleeding intraoperatively

and special consideration and precaution is taken beforehand and meticulous surgical technique should be applied to avoid the complication. Apart from that, LASER photocoagulation is also available as an alternative surgical treatment and provides a good result in selected cases^{13, 14}. Although LASER has less complication, the repeated number of treatment, variable response and recurrence of the lesion are all disadvantage of this modality.

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