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HEMANGIOLYMPHANGIOMA OF TONGUE, REPORT OF A RARE CASE

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ABSTRACT
Hemangiolymphangioma is a variant of lymphangioma showing vascular component. Lymphangiomas are further subclassified microscopically into capillary, cavernous, cystic and lymphangioendothelioma, depending upon their histopathological features. Lymphatic malformations or lymphangiomas are uncommon congenital malformations of the lymphatic system, usually occurring in the head and neck region, characterized by collections of ectatic lymph vessels that form endothelial lined cystic spaces. Malformations of vascular nature originate as anomalies caused due to errors in vasculogenesis. Early recognition is of utmost importance for initiation of proper treatment and avoiding serious complications. These tumors are generally broadly classified into vascular tumors (hemangiomas) and vascular malformations (venous malformations, arteriovenous malformations, lymphatic malformations). Advancements in the knowledge of such vascular malformations are continuously changing their treatment protocols. Herewith, we present a rare case of vascular malformation diagnosed as hemangiolymphangioma in a two year six months old boy, along with a review of literature regarding its categorization.

KEYWORDS: Hemangiolymphangioma, vascular malformation, lymphangioma.

INTRODUCTION
Hemangiolymphangioma is a variant of lymphangioma showing vascular component.[1] The need for categorization of anomalies and congenital aberrancies formed due to developmental vascular defects produce identifiable birthmarks of the skin and mucosa and a variable degree of underlying soft tissue abnormalities.[2] Involvement of the oral cavity is common but frequently requires unconventional treatment strategies for its management.

Though previously termed “angiomas” or vascular “birthmarks”, vascular anomalies are divided into two main categories: vascular tumors and vascular malformations. Infantile hemangiomas comprise the majority of vascular anomalies and are considered the predominant vascular tumor type composed of rapidly proliferating endothelial cells.[3]

Blood vessel architecture is incomplete and surrounded by hyperplasic cells in hemangiomas and other vascular tumors. In contrast, vascular malformations do not contain hyperplasic cells but consist of progressively enlarging aberrant and ectatic vessels composed of a particular vascular architecture such as veins, lymphatic vessels, venules, capillaries, arteries or mixed vessel type.

The latter comprises lymphangiomas or lymphatic malformations which are congenital collections of ectatic lymph vessels that form endothelial lined cystic spaces.[4] The pathogenesis of these tumors could be of importance in thoroughly understanding the mode of these varying histopathological presentations. Lymphangiomas are benign, relatively rare tumors characterized by proliferation of lymphatic vessels. They represent about 6% of the total number of benign tumors of the soft tissue in patients aged less than 20 years.[5]

Regarding gender distribution of lymphangioma, it is equally divided between males and females, with about 50% of the lesions being noted at birth and 90% developing by 2 years of age.[6] Oral lesions may occur at various sites but they form most frequently on the anterior two thirds of tongue. They may increase in size, producing macroglossia which may lead to difficulties in mastication, deglutition, and speech; and displacement of the teeth, with a resulting malocclusion. They may interfere with normal breathing, particularly during sleep, produce sleep apnea, and in certain instances, produce a life-threatening upper airway compromise.[7]

They can also be present in the palate, buccal mucosa, gingiva and lip.[5] The tumor is superficial in location and demonstrates a white pebbly surface that resembles a cluster of translucent vesicles. The deeper lesions could

mimic various soft tissue tumors since the color which is classically used for diagnosing such tumors would seem to be irrelevant.

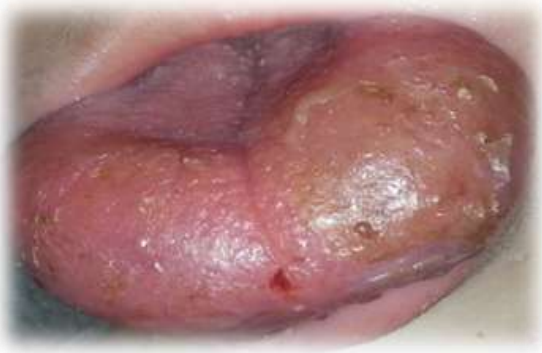
CASE REPORT

Our patient is a two years six months male patient, born to nonconsanguineous parents by cesarean section due to previous one; he is product of full term to a pregnancy complicated by maternal depression. He was born with a protruded tongue. Admitted at the age of one week to ICN for observation because of refusal of breast feeding and difficult to suck bottle, where was observed for two weeks then discharged home with no diagnosis. For also difficulty of feeding readmitted to the Hospital at the age of two months with tongue external protrusion and edema. CT angiography of neck revealed macroglossia associated with enlargement of the submandibular glands (filled with fluid density, no solid masses) and their ducts, most likely representing obstruction secondary to compression of the opening of the Wharthon ducts at the floor of the mouth. Ultrasound thyroid and neck showed septated cystic structures identified bilaterally, displacing the submandibular salivary glands, most likely representing dilated salivary ducts. In keeping with the previously mentioned diagnosis of cystic enlargement of the submandibular salivary gland ducts secondary to macroglossia. At four months of age, patient's tongue passed through 3 stages: 1-first week: eruption of small red spots associated with severe pain that prevented our patient from sleep. During this stage patient manifests fever up to 39 degrees associated with chills. 2- Second week: tongue became edematous; spots enlarge then burst pus and blood followed by resolution of fever. 3- Third and Fourth weeks: tongue continues to discharge blood and pus until it becomes necrotic and sloughs completely off with resolution of the edema and pain. During this period patient received antibiotics, after which he was admitted for surgical drainage of a cyst at the level of the tongue's frenulum, which contained blood and pus. Also underwent a tracheostomy, to secure and protect the airway.

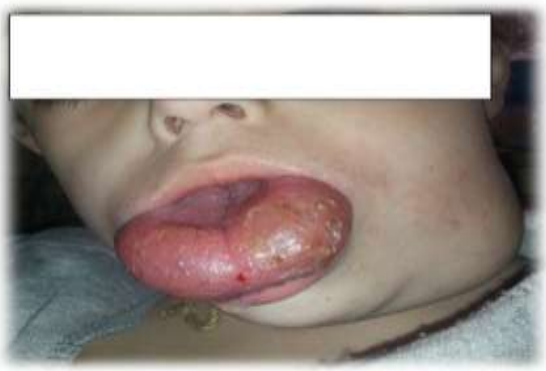
At one year and eleven months of age, patient developed same episode of tongue stages and was readmitted for another cyst decompression surgery this time including the mouth floor. One week later, patient developed bilateral neck swellings that were drained by fine needle aspiration. The drainage contained pus and blood. At two years five months and fifteen days of age, patient developed a left submandibular mass, increasing in size, painful, becomes congested and bluish at night. This swelling was associated with edema and pustular tongue discharge with fever reaching 39℃ associated with chills that resolved three days prior to presentation with end of pustular drainage. MRI of oral cavity revealed: infiltrative process including the tongue with some prominent vessels, there are mildly prominent lingual arteries however the diagnosis is in favor of a low flow vascular malformation including capillary, lymphatic and venous components, the tumor appears to infiltrate the genioglossus, mylohyoid, digastric, and geniohyoid muscles, mandible not involved, the abnormality of the left submandibular region extends into the left parapharyngeal space without involvement of the pterygoid muscles, it measures 5.5cm, containing cystic spaces, the right submandibular mass measures 3 cm containing venous structures on ultrasound.

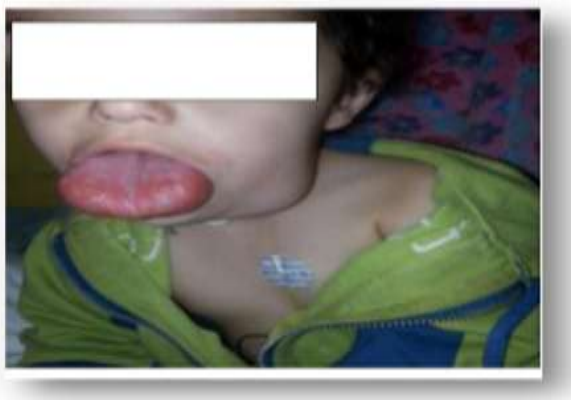
So we are in front of a case of congenital tongue lymphohemangioma S/P multiple excisions of the lesions and S/P prophylactic tracheostomy presented with recurrence of the lymphohemangioma. Given propranolol treatment as by AAP recommendations and had mild decrease in size of the lesion.

Pretreatment



Posttreatment





DISCUSSION

Lymphangioma was first described by Virchow in 1854, and in 1872, Krester hypothesized that hygromas were derived from lymphatic tissue. The origin of lesion is considered to be congenital abnormality of lymphatic system rather than true neoplasm. A portion of the jugular lymphatic sac is thought to sequestrate from the primary sacs during fetal development with failure to establish communications with other lymphatic system.[8]

The fact that most lymphangiomas manifest clinically during early childhood and develop in areas where the primitive lymph sacs occur (neck, axilla) provides presumptive evidence for this hypothesis. On the contrary, it is argued that instead of being a congenital malformation, lymphangioma is a true neoplasm resulting from transformed lymphatic endothelial cells and/or stromal cells.[9]

The pathophysiology of vascular malformation, hemangioma and lymphangioma are interrelated. The classic sequence of events in embryology and development of vasculogenesis falls into three stages: the undifferentiated capillary network stage, the retiform developmental stage and the final developmental stage.[10]

Two major theories have been proposed to explain the origin of lymphangiomas.[11] The first theory is that the lymphatic system develops from five primitive sacs arising from venous system. Concerning the head and neck, endothelial out pouching from the jugular sacs spread centrifugally to form the lymphatic systems. Another theory proposes that the lymphatic system develops from mesenchymal clefts in the venous plexus reticulum and spreads centripetally toward the jugular sacs. Finally, lymphangioma develops from congenital obstruction or sequestration of the primitive lymphatic enlargement.[12]

Several studies have been published regarding possible lymphangiogenic growth factor involvement in the etiology of lymphatic malformations. These factors include vascular endothelial growth factor (VEGF)-C, vascular endothelial growth factor receptor 3 (VEGFR-3), and transcription factor Prox-1. VEGF-C and VEGFR-3 have been shown to be upregulated in lymphatic malformed tissue, and both are involved in lymphatic tissue proliferation.[13]

Oral lesions are most frequently found on tongue. The tumor may be localized in a small area of tongue or floor of mouth or it may diffusely infiltrate these areas. If the tumor is located in a deeper area, it may present as submucosal mass.[14]

The misunderstanding on the nosologic distinction between oral hemangiomas and vascular malformations leads to diagnostic mistakes. Hemangiomas are differentiated from vascular malformations by their clinical appearance, histopathological features, and biologic features. The natural history of hemangiomas involves rapid proliferations for the first several months of life with subsequent spontaneous regression. Vascular malformations are often recognized at birth and grow proportionately with the child, with many becoming more prominent at puberty. Histological, hemangiomas in the proliferating phase show endothelial hyperplasia and large number of mast cells. In contrast, vascular malformations show normal number of mast cells, and consist of mature, often combined, capillary, arterial, venous, and lymphatic elements.[2]

Lymphatic malformations/lymphangiomas are classified microscopically into four categories.[15] Lymphangioma simplex (lymphangioma circumscriptum) composed of small, thin-walled lymphatics; cavernous lymphangioma comprising dilated lymphatic vessels with surrounding adventitia; cystic lymphangioma (cystic hygroma) consisting of huge, macroscopic lymphatic spaces with surrounding fibrovascular tissues and smooth muscle; and benign lymphangioendothelioma (acquired progressive lymphangioma), in which lymphatic channel dissects through dense collagen bundles.

Occasionally, channels may be filled with blood, a mixed hemangiolymphangioma, an uncommon developmental anomaly with a propensity to invade underlying tissues and to recur locally, distinguishing it from the simple lymphangioma or hemangioma.[16]

CONCLUSION

Vascular anomalies like hemangioma and lymphangioma are interlinked in their pathogenesis. The vascular lesions consist of both blood vessels and lymphatic vessels. Whether these can be termed as hemangiolymphangioma, lyphhemangioma or just vascular malformation is still confusing. Thus through the present article we would like to highlight the complexities which can arise from such cases.

REFERENCES

1. Case series on vascular malformation and their review with regard to terminology and categorization. Devi Charan Shetty, Aadithya B. Urs, Harish Chandra Rai, Nitin Ahuja, and Adesh

Manchanda. Contemp Clin Dent., 2010 Oct-Dec; 1(4): 259–262.
2. Buckmiller LM, Richter GT, Suen JY. Diagnosis and management of hemangiomas and vascular malformations of the head and neck. Oral Diseases., 2010; 16: 405–18.
3. Drolet BA, Esterly NB, Frieden IJ. Hemangiomas in children. N Engl J Med., 1999; 341: 173–81.
4. Kohout MP, Hansen M, Pribaz JJ, Mulliken JB. Arteriovenous malformations of the head and neck: natural history and management. Plast Reconstr Surg., 1998; 102: 643–54.
5. Coffin CM, Dehner LP. Vascular tumours in children and adolescents: a clinicopathologic study of 228 tumours in 222 patients. Path Annu., 1993; 28: 97–120.
6. Jeeva Rathan J, Harsha Vardhan BG, Muthu MS, Saraswathy K, Sivakumar N. Venkatachalapathy. Oral lymphangioma: A case report. J Indian Soc Pedod Prev Dent., 2005; 23: 185–9.
7. Dinerman WS, Myers EN. Lymphangiomatous macroglossia. Laryngoscope., 1976; 86: 291–6.
8. Rice JP, Crson SH. A case report of lingual lymphangioma presenting as recurrent massive tongue enlargement. Clin Pediatr Phila., 1985; 24: 47–50.
9. Huang HS, Ho CC, Huang PH, Hsu SM. Co-expression of VEGF-C and its receptors, VEGFR-2 and VEGFR-3, in endothelial cells of lympangioma.Implication in autocrine or paracrine regulation of lymphangioma. Lab Invest., 2001; 81: 1729–34.
10. Shafer, Hine, Levy. In: Textbook of oral pathology. 5th ed. Rajendran, Sivapathasundaram, editors. New Delhi: Elsevier., 2006; 197–9.
11. Patel JN, Sciubba J. Oral lesions in young children. Pediatr Clin North Am., 2003; 50: 469–86.
12. Bill AH, Summer DS. A unified concept of lymphangiomas and cystic hygroma. Surg Gynecol Obstet., 1965; 120: 79–86.
13. Itakura E, Yamamoto H, Oda Y, Furue M, Tsuneyoshi M. VEGF-C and VEGFR-3 in a series of lymphangiomas: is superficial lymphangioma a true lymphangioma? Virchows Arch., 2009; 454: 317–25.
14. Lobitz B, Lang T. Lymphangioma of the tongue. Pediatr Emerg Care. 1995; 11: 183–5.
15. Meher R, Garg A, Raj A, Singh I. Lymphangioma of tongue. Internet J Otorhinolaryngology., 2005; 3: 2.
16. Seong Soo K. Intraosseous haemangiolymphangioma of the mandible: a case report. J Korean Assoc Oral Maxillo Surgeons., 2003; 29: 182–5.
17. Stanescu L, Georgescu EF, Simionescu C, Georgescu I. Lymphangioma of the oral cavity. Romanion J Morph Embryolog., 2006; 47: 373–7.