Case 17
A 12-year-old Thai boy from Saraburi Province
Chief complaint: A 8-year history of progressive mass at tongue


Fig. 17.1

Present illness:
He presented with an 8-year history of multiple erythematous papules on his tongue. In the past four years, the lesion gradually progressed in size without pain nor bleeding. He was performed excisional biopsy one year ago while the size was 1.5 x 0.5 centimeter. The mass recurrent and progressed with no clinical bleeding nor airway compromised. He underwent genetic consultation showed no significant relationship with a genetic disorder, then he was transferred to the dermatologist for further management.

Past history: He was born with gastroschisis, term, normal delivery. His abdominal wall was received closure procedures successfully without any complication.

Family history: There was no family history of cancer or similar lesion.

Physical examination: Surgical scar at abdominal wall. Others were unremarkable.

Dermatological examination:
- Macroglossia with 7x3 centimeter.
- Multiple group of well-defined bright erythematous papules on dorsoanterior to left anterolateral side of tongue, 4x2 centimeter. (Fig.17.1)

Histopathology: (S18-039453, tongue) (Fig 17.2)
- Exophytic nodule composed of proliferation of capillary blood vessels and lymphatic channels with valve formation

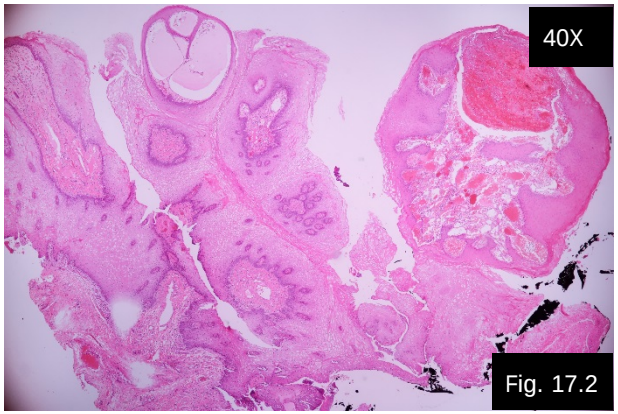

Fig. 17.2

Investigations:
- MRI: No detectable gross lesion at the tongue and no significant cervical lymphadenopathy

Diagnosis: Lymphohemangioma of tongue
Treatment: Long pulsed dye 1064 nm
Presenter: Kallapan Pakornphadungsit, MD
Consultant: Somsak Tanrattanakorn, MD

Discussion:

Vascular anomalies are divided into two primary entities: vascular neoplasms and vascular malformation.[1] The difference between the two types is based on histopathology whether there is increased or decrease endothelial cell turnover. Vascular neoplasms have increased endothelial cell turnover, whereas vascular malformations do not have increased endothelial cell turnover. Alternatively,

vascular malformations are structural abnormalities of the veins, arteries, capillaries and lymphatic vessels.[2]

Infantile hemangioma comprises the majority of vascular anomalies and are considered the predominant vascular tumor type composed of rapidly proliferating endothelial cells. Head and neck are the most common tumors of infancy. Natural course of hemangiomas appears in the first few weeks of life, presents a rapid proliferative phase, which usually lasts 6–10 months, followed by gradual involution. Lymphangiomas frequently manifest at birth or before two years of age. Lymphangioma is considered as a benign hamartomatous tumor of lymphatic vessels. Lymphangioma usually presents at birth or within 2 years of life.[1,3] The most common location for lymphatic malformation is in the neck, the location of the primitive jugular lymphatic sac. Portions of these primitive sacs may sequestrate and form separated remnants that have growth potential.[4] When the resulting proliferation fails to achieve anastomosis with the larger lymphatic or venous channels, a lymphangioma develops. The overlying skin is usually normal or may have a bluish hue.[3] Although vascular anomalies is a benign lesion, it may lead to a complicated case because of local invasion as its infiltrating nature, indefinite boundary, and the involvement of vital structures. Most of the hemangiomas are asymptomatic. Some present like symptomatic lesions including mass effect, dysphagia, dyspnea and hemorrhage. Lymphatic malformations do not commonly regress spontaneously, or involute partially compared to hemangioma which more frequent complete regression.[6]

Malformations may be seen in different combinations of vascular elements including lymphatic and venous endothelium and cannot be identified as purely one or the other entities. These mixed vascular malformations are named as lymphohemangioma or hemanglymphangioma according to the dominant tissue structure. Lymphohemangioma is combined deformities of blood, and lymphatic vessels.[9] To date, lower than 10 cases have been reported in the English-language regarding lymphohemangioma. All of those cases were found on face, especially at oral mucosa. Histologically examination reveals the combination of both lymphatic vessels and capillary vessels filled with red blood cells, which representative of lymphangiomatous and hemangiomatous elements. Hence, the definitive diagnosis is established by histopathology.[1,7-11] However, they can also be thought of as a continuum of the same pathologic process.[11]

Various modalities with various outcomes have been reported for the treatment of hemangiomas and lymphangiomas. Therapeutic procedures such as conservative treatment, radiation therapy, cryotherapy, electrocautery, sclerotherapy, steroid administration, sclerosing agents, embolization, ligation, laser surgery and surgical excision have been reported in treatment of vascular malformations.[4,12-15] Surgical treatment should be performed for the benefit of minimizing the risk of complications. Total excision is often difficult, recurrences are seen.[12] However treatment of choice for vascular malformations should be early, conservative surgical excision with preservation of vital structures. [6,12]

Currently, the standard treatment of lymphohemangioma has not been well-described. Consequently, the appropriate modalities of treatment of lymphohemangioma should be selected wisely considering risks, benefit, and suitability based on treatment as literatures of vascular malformations due to their clinical and histopathological correlation.

References:

1. Yarmand F, Seyyedmajidi M, Shirzad A, Foroughi R, Bakhshian A. Lymphangiohemangioma of buccal mucosa: Report of a rare case. J Oral Maxillofac Surg Med Pathol. 2016;28:358–61.
2. Hashemipoor M. Rare cavernous haemangioma of the oral cavity with numerous phleboliths. J Oral Health Oral Epidemiol. 2012;1:46–51.

3. Marler JJ, Mulliken JB. Current management of hemangiomas and vascular malformations. Clin Plast Surg. 2005;32:99–116.
4. Ikeda H, Fujta M, Nonaka M, et al. Cystic lymphangioma arising in the tip of the tongue in an adult. Int J Oral Maxillofac Surg. 2006;35:274-6
5. Bozkaya S, Ugar D, Karaca I, et al. The treatment of lymphangioma in the buccal mucosa by radiofrequency ablation: a case report. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2006;102:28-31.
6. Tasca RA, Myatt HM, Beckenham EJ. Lymphangioma of the tongue presenting as Ludwig's angina. Int J Pediatr Otorhinolaryngol .1991;51:201-5.
7. Sobhana CR, Beena VT, Soni A, Choudhary K, Sapru D. Hemangiolymphangioama of buccal mucosa: Report of a rare case and review of literature on treatment aspect. Natl J Maxillofac Surg. 2012;3:190–4.
8. Kim SS. Intraosseous hemangiolymphangioma of the mandible: A case report. J Korean Assoc Oral Maxillofac Surg. 2003;29:182–5.
9. Hunchaisri N. Hemangiolymphangioma of the floor of mouth: A case report and literature review. J Med Health Sci. 2013;20:4–9.
10. Jian XC. Surgical management of lymphangiomatous or lymphangiohemangiomatous macroglossia. J Oral Maxillofac Surg. 2005;63:15–9.
11. Selvamani M, Prem S, B Nanda K, Sheethal J. Hemangiolymphangioma of buccal mucosa: A rare case report. J Oral Maxillofac Pathol. 2017;21: 282–5.
12. Lille T, Rand R, Tapper D, et al. The surgical management of giant cervicofacial lymphatic malformations. J Ped Surg. 1996;31:1648-50.
13. Tasca A, Myatt M, Beckenham J. Lymphangioma of the tongue presenting as Ludwig's angina. Int J Pediatr Otorhinolaryngol. 1991;51:201-5.
14. Bonet-Coloma C, Mínguez-Martínez I, Palma-Carrió C, Galán-Gil S, Peñarrocha-Diago M, Mínguez-Sanz JM. Clinical characteristics, treatment and outcome of 28 oral haemangiomas in pediatric patients. Med Oral Patol Oral Cir Bucal. 2011;16:19-22.
15. Bozkaya S, Ugar D, Karaca I, et al. The treatment of lymphangioma in the buccal mucosa by radiofrequency ablation: a case report. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2006;102:28-31.