Net Quiz

A 12-year-old girl presented with gradually enlarging painless swellings on the dorsal surface of her tongue. According to her parents, the lesions had been present almost since her birth. Besides slight difficulty in swallowing of foods, the lesions were asymptomatic in nature. There was no history of trauma. Her past medical history was unremarkable. Clinical examination revealed multifocal, pinkish, pebbly, tense vesicular lesions [Figure 1] on the dorsum of the posterior third of the tongue. There was no dental abnormality. Examinations of the skin and other systems were normal. Routine laboratory investigations were noncontributory. An excisional biopsy of the lesion was performed.

Histopathological examination of the serial sections of the excision biopsy specimen showed dilated endothelium-lined channels filled with acellular amorphous eosinophilic materials. The overlying stratified squamous epithelium was hyperplastic. [Figures 2 and 3].

WHAT IS YOUR DIAGNOSIS?



Figure 2: Dilated channels filled with acellular amorphous eosinophilic materials with overlying hyperplastic stratified squamous epithelium (H and E, \times 40)



Figure 1: (a) Multifocal tense cystic lesions on the tongue (b) Excision biopsy specimen showing tense vesicles



Figure 3: Dilated endothelium-lined channels filled with lymph fluid (H and E, $\times 100$)

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Diagnosis: Lymphangioma circumscriptum of the tongue

DISCUSSION

Lymphangiomas are benign hamartomas of the lymphatic system. Although lymphangiomas have a marked predilection for the head and neck region, they can also affect other anatomical areas such as the abdomen and oral cavity, particularly the tongue.^[1] According to the size of the lymphatic vessels involved and the depth of the lesions, lymphangiomas are classified into four types, namely, cavernous lymphangioma lymphangioma, cystic hygroma, circumscriptum, acquired and progressive lymphangioma.^[2]

The lesions of lymphangioma circumscriptum are painless, non-inflammatory, grouped, (LC) tense vesicles containing clear fluid (resembling frog's spawn).^[3] Cavernous lymphangiomas are large, diffuse subepithelial masses that may be fluctuant. Cystic hygromas are circumscribed cystic lesions considered by many as ectatic variants of cavernous lymphangioma.^[1] Acquired progressive lymphangiomas are relatively rare benign tumors that involve middle-aged or elderly adults and evolve slowly over years.^[2] Trauma to, or infection of the lymphangiomas of the tongue may result in sudden increase in the size of the lesions and severe pain.^[4] Lymphangiomas involving the tongue often interfere with speech and swallowing and rarely may cause gross structural deformities of the face. They do not regress spontaneously. The clinical differential diagnoses of lingual LC include^[1,3] hemangioma, neurofibroma, lingual thyroid, dermoid cyst, thyroglossal duct cyst, granular cell tumor, and heterotopic gastric mucosal cyst. The presence of superficial, tiny vesicles with or without hemorrhage is a helpful pointer to the diagnosis of LC. A definitive diagnosis should be made by biopsy and histopathological examination.^[5] Magnetic resonance imaging and lymphangiography are useful in demonstrating the full extent of the disease.^[3] Histopathology of LC shows dilated lymphatics (either solitary or grouped) containing lymph or blood. The epidermis overlying the vesicle may show elongation of rete ridges and the vesicles may appear intraepidermal.^[3] Lymphangiomas are resistant to medical management; surgical excision is the usual treatment of choice.^[1] Radiation therapy, cryotherapy, electrocautery, sclerotherapy, steroid administration, embolization, ligation, and laser surgery are the other treatment options available for lymphangioma of the tongue.^[3,4]

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