

Lymphangioma Circumscriptum of the Tongue

A 7-year-old girl presented with diffuse enlargement of the tongue since birth. She had developed complaints of recurrent bleeding from the tongue associated with fissuring since one year. The swelling was asymptomatic except for difficulty in eating and pain, which occurred only during episodes of severe bleeding. She did not have any other cutaneous and/or systemic symptoms. She was born to healthy nonconsanguineous parents following an uneventful full-term pregnancy.

Clinical examination revealed diffuse enlargement of the tongue, which was soft and non-tender. Close inspection revealed



Fig.1. Numerous pinhead sized clear and few hemorrhagic vesicles on the dorsal surface of tongue.

numerous, pinhead-sized, clear vesicles, few of which were hemorrhagic on the dorsal surface of the tongue along with a few longitudinal fissures (*Fig. 1*). Systemic examination did not reveal any abnormalities. Tongue biopsy revealed features of lymphangioma.

Lymphangioma circumscriptum is a congenital malformation of superficial lymphatics present at birth or soon after. It presents as a group of vesicles containing clear fluid (resembling frog's spawn), the size of which can be up to 5 mm in diameter. Sometimes the vesicles can be hemorrhagic. Hemorrhage can occur spontaneously or secondary to trivial trauma. The proximal part of the extremities is the commonest site involved. It can also occur over the abdomen and mouth, particularly the tongue. In the typical form the superficial lymphatics communicate through deep vessels with large closed lymphatic cistern in the sub-cutis or the deep dermis. This will explain the tendency of lesions recurring after superficial excision. Magnetic resonance imaging and lymphangiography are useful in demonstrating the full extent of the disease. Histopathologically it shows dilated lymphatics (either solitary or grouped) containing lymph or blood. The epidermis overlying the vesicle appears to be atrophic with elongation of rete ridges since the vesicles may appear intraepidermal. It is an important cause of macroglossia in children and the clinical differential diagnosis include vascular malformation, hemangioma, neurofibroma, idiopathic muscle hypertrophy, Down's syndrome, mucopolysaccharidosis, hypothyroidism, and diabetes. It can easily be differentiated by the presence of superficial, tiny vesicles with or without hemorrhage.

There is no satisfactory treatment for this condition. Surgical excision, Carbon dioxide (CO₂) laser, liquid nitrogen and injectable sclerosants have been used with variable results. The recognition of this entity is important as it may cause speech disturbances, difficulty in eating and swallowing, episodes of choking or dyspnea and severe hemorrhage.

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