Oral lymphangioma – Case reports and review of literature

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Abstract

Lymphangioma is a benign hamartomatous hyperplasia of lymphatic vessels. Majority of them are superficial, but a few may extend deeply into the connective tissue. Intraoral lymphangiomas occur more frequently on the dorsum of tongue, followed by palate, buccal mucosa, gingiva, and lips. Surgical excision is the treatment of choice. The prognosis is good for most patients, although large tumors of neck/tongue may result in airway obstruction and death. This case report series discusses the clinical features, histopathology, and treatment of lymphangioma.

Keywords: Lymphangioma, lymph vessels, papule

Introduction

Lymphangiomas are benign, hamartomatous malformations arising from the sequestrations of lymphatic tissue. Lymphangiomas have marked predilection for the head and neck region [50-70%].[1] About half of the lesions are noted at birth and around 90% develop by 2 years of age. Small lymphangiomas, less than 1cm, occur on the alveolar ridge, with 2:1 male to female distribution. Histopathological features consist of multiple intertwining lymph vessels in a loose fibrovascular stroma. Surgical excision is the treatment of choice. The prognosis is good for most patients, although large tumors of neck/tongue may result in airway obstruction and death.[2] We here report three cases of intraoral lymphangiomas, reviewing the literature.

Case Reports

Case 1

A 7-year-old female child reported with multiple papular lesions on the anterior two-thirds of the dorsum and ventral surface of tongue [Figures 1 and 2]. The lesions were present from birth, she had feeding difficulty in early days, and bleeding occurred frequently. There was no family history of the disease. Medical investigations were all within normal limits. An incisional biopsy was taken and histopathological examination revealed numerous endothelial lined small lymphatic channels containing lymph tissue. Extravasated red blood cells were noticed [Figure 3].

Case 2

A 32-year-old female patient reported to our department with a swelling on the right lateral dorsum of the tongue, which she noticed a few months back. There was bleeding occasionally from the swelling. On examination, a 3 cm × 2 cm nodular mass with papillary lesions, with slight change in the surface texture, was noticed on the middle one-third of the right lateral dorsum of the tongue [Figures 4 and 5]. A few blood-filled papules were noticed on the surface of the lesion. No cervical lymphadenopathy was noticed. Incisional biopsy was performed, and the histopathology showed endothelium-lined lymphatic channels filled with lymph, below the epithelium. A few extravasated RBCs were also noticed [Figure 6].

Case 3

An 8-year-old male child reported with bleeding from an enlarged tongue. On examination, multiple papular lesions were noticed on the anterior two-thirds of the dorsum of the tongue. A few blood-filled papules were seen, and the rest of the lesion had the same color as the adjacent mucosa [Figure 7]. Incisional biopsy was done, and histopathological examination showed multiple intertwining lymph vessels in a loose fibrovascular stroma [Figure 8].

Discussion

Lymphangiomas are benign, hamartomatous tumors of lymphatic vessels. They represent developmental malformations that arise from sequestrations of lymphatic tissue that do not communicate normally with the rest of the lymphatic system. Three types, a] lymphangioma simplex (capillary lymphangioma), b] cavernous lymphangioma, and c] cystic lymphangioma (cystic hygroma)], have been described.[1,2,3]

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Figure 1: Clinical photograph showing papular lesions on tongue

Figure 2: Clinical photograph showing papular lesions on tongue

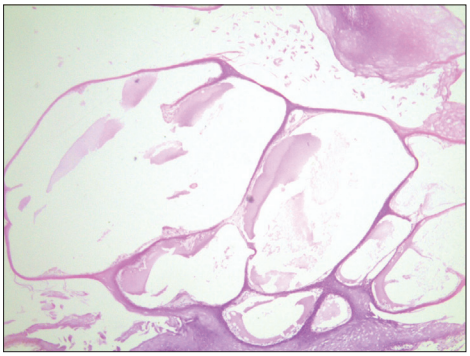

Figure 3: Photomicrograph showing endothelial lined lymphatic channels filled with lymph [H & E stain]

Figure 4: Clinical photograph showing papular lesions on tongue

Figure 5: Clinical photograph showing papular lesions on tongue with macroglossia

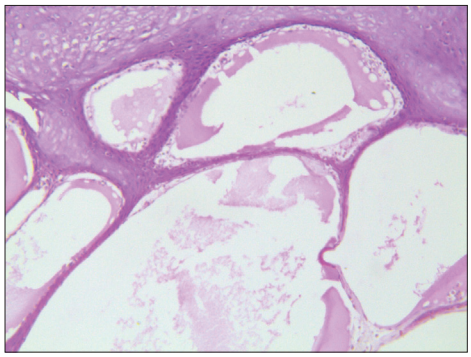

Figure 6: Photomicrograph showing endothelial-lined lymphatic channels filled with lymph [H and E stain]

Figure 7: Clinical photograph showing papular lesions on tongue with macroglossia

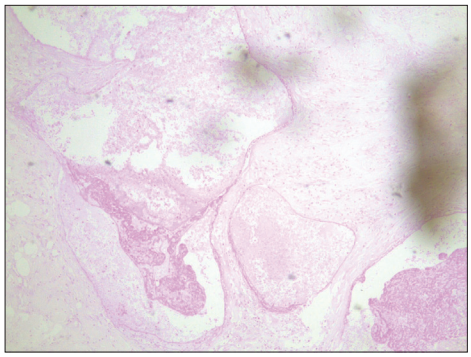

Figure 8: Photomicrograph showing endothelial-lined vessels filled with lymph and extravasated RBC [H and E stain]

Oral lymphangiomas may occur at various sites but are more frequent in the anterior two-thirds of the tongue, where they often result in macroglossia. Usually, the tumor is superficial in location and demonstrates a pebbly surface, resembling a cluster of translucent vesicles. The surface appears like frog eggs or tapioca pudding. Deeper lesions present as soft, ill-defined masses.

The superficial lesions are manifested as papillary lesions, which may be of the same color as the surrounding mucosa or of a slightly reddish hue. The deeper lesions appear as diffuse nodules or masses without any significant change in surface texture or color.

If the tongue is affected, macroglossia occurs. The anterior dorsal part of the tongue is most frequently affected. Irregular nodularity of the surface of the tongue with gray and pink projections and macroglossia is the pathognomonic feature of lymphangioma of the tongue.[4,5,6,7,8]

Small lymphangiomas, less than 1 cm, occur on the alveolar ridge, with 2:1 male to female distribution.

Histopathologic features consist of lymphatic vessels with marked dilatations.[7] The vessels often diffusely infiltrate the adjacent soft tissues and demonstrate lymphoid aggregates in their walls. The lining endothelium is thin and the spaces contain proteinaceous fluid and lymphocytes. Secondary hemorrhage may be noticed in the lymphatic vessels. The lymphatic spaces contain lymphatic fluid, red blood cells, lymphocytes, macrophages, and neutrophils. Surrounding connective tissue stroma consists of loose fibrotic tissue with a number of inflammatory cells.[1,3,5,6]

The capillary lymphangioma consists of small capillary-sized vessels, cavernous lymphangioma consists larger dilated lymph channels, and cystic lymphangioma exhibits large macroscopic cystic spaces. More often, all sizes of vessels may be found within the same lesion.

In the intraoral lymphangiomas, lymphatic vessels are located just beneath the epithelium, often replacing connective tissue papillae.

Surgical excision is the treatment of choice. Because of the non-encapsulated and "infiltrating" nature of the lymphangioma, complete removal is difficult, and hence recurrence is common. Lymphangiomas do not respond to sclerosing agents as do hemangiomas.

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