Lymphangioma of the buccal mucosa – A case report and review of literature

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ABSTRACT

Background and Objective: *Lymphangiomas are developmental malformations of lymphatic vessels, considered to be hamartomas rather than true neoplasms. These are usually diagnosed in infancy or early childhood and are uncommon in adults. The objective of this case report is to present an unusual case of lymphangioma of the buccal mucosa.*

Case Summary: *A 17 year old male patient reported with a swelling of the buccal mucosa which was noticed at 4 years of age and gradually increased in size. Histopathological examination of the lesion revealed the presence of dilated lymphatic channels and diagnosed as cavernous lymphangioma.*

Keywords: *Cheek, hamartoma, tumour, lymphatic vessels*

INTRODUCTION

Lymphangioma is a benign hamartomatous tumor of the lymphatic vessels with predilection for the head and the neck region. This was described first by Redenbacher in 1828[(1)]. About 50% of lesions are noted at birth and around 90% develop by 2 years of age. Oral lymphangiomas are most frequent on the anterior two-thirds of the tongue where they often result in macroglossia[(2)]. They are also known to be associated with Turners syndrome, Noonan's syndrome, trisomies, cardiac anomalies, and fetal alcohol syndrome[(3)].

Three theories explain the origin of this abnormality. The first suggests that a blockage of normal growth of the primitive lymph channels occurs during embryogenesis, the second proposes that the primitive lymphatic sac does not reach the venous system and the third theory advances the hypothesis that, during embryogenesis, lymphatic tissue is in a wrong location[(4)]. These lesions are categorized into capillary lymphangioma, cavernous lymphangioma, and cystic lymphangioma.[(5)] Lymphangiomas less than 1 cm in dimension occur bilaterally on the alveolar ridge in about 4% of blackneonates. Lymphangioma simplex involves small vesicles which range from pink to red to black due to secondary hemorrhage. Cavernous lymphangiomas appear as subcutaneous rubbery nodules. They have a clear dimension than lymphangioma simplex. Cystic lymphangiomas are usually larger than cavernous lymphangiomas and commonly occur in the head and neck in the parotid region[(2)].

Specific antibodies for lymphatic endothelium are VEGR3 (vascular endothelial growth factor receptor 3), podoplanin, lymphatic vessel endothelial HA receptor-1 (LYVE-1), Prox1 and D2-40. Podoplanin and D2-40 monoclonal antibody can be used as lymphatic endothelial cell markers for distinguishing blood vessels from lymphatic vessels. Lesions designated as acquired lymphangiectasia may develop as a result of infection or surgery that interferes with regional lymphatic drainage[(6)].

CASE REPORT

A 17 year old male patient reported with a complaint of swelling in the right side of the oral cavity, in the cheek. This swelling was noticed at the age of 4 years and it gradually increased to the present size. On examination, there were small multiple translucent papules on the right commissural region of the lip (Figure-1), which were firm and non tender. Intraorally there was presence of swelling on the right side of the buccal mucosa with multiple small translucent papules(Figure-2).On palpation the swelling was nodular , firm to soft in consistency, non tender, non compressible and without any evidence of bleeding. A provisional diagnosis of intra oral lymphangioma of the buccal mucosa was established. A differential diagnosis of arteriovenous malformation was considered. Occlusion was normal with no alteration in the maxillary and mandibular bone observed on OPG and occlusal radiograph. Incisional biopsy under local anesthesia was performed for histopathological examination.

Microscopic examination revealed the presence of dilated but variably sized lymphatic vessels separated by thin, delicate septae. The parakeratinized stratified squamous epithelium

overlying the lymphatics were thinned out in certain locations. The lymphatic channels were lined with thin layer of bland endothelial cells and filled with lymph. The stroma comprised of loose connective tissue with inflammatory cells (Figure-3 and 4). These pathological features were consistent with the diagnosis of cavernous lymphangioma.

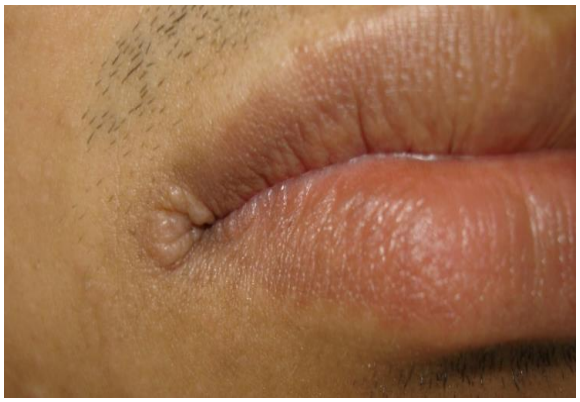

Fig. 1: showing multiple small papules at the right commissure.

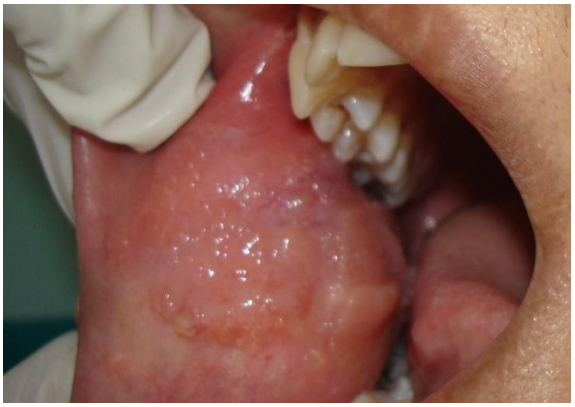

Fig. 2: showing multiple translucent papules on the buccal mucosa.

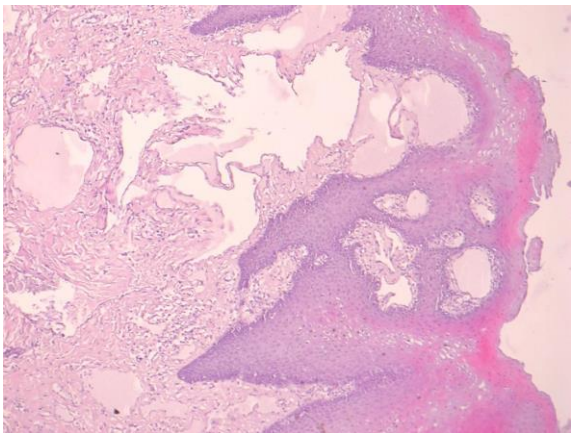

Fig. 3: Section showing irregular, dilated lymphatic channels with lymph, separated by thin septae, below the epithelial surface. Obj 10x, H&E stain.

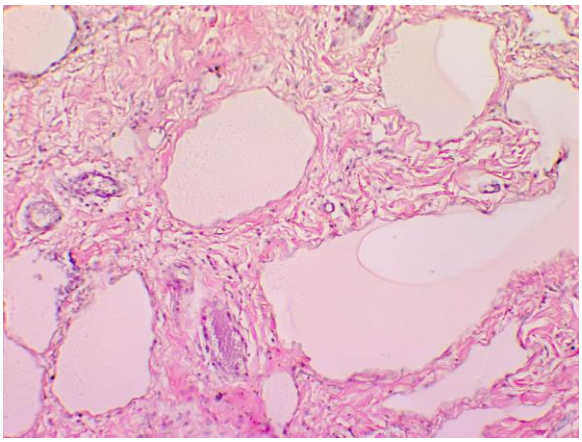

Fig. 4: Thin band of bland endothelial cells lining the lymphatic channels are obvious under higher magnification. The loose connective tissue stroma with minimal inflammatory cell infiltrate is also observed in this section. Obj 20x, H&E stain

DISCUSSION

Lymphangiomatous lesions are rare congenital malformations of the lymphatic system that occur throughout the body with greater frequency in the cervico-facial area. These are benign tumors comprising 5.6 percent of all benign lesions in children [(7)].

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 colour[(8)].

Introduction of prenatal ultrasound allows detection of disease, in approximately 67% cases. Histologically, these lesions are composed of dilated lymphatic channels with one or two endothelial layers, with or without an adventitial layer. These dilated lymphatics can vary in size, depending upon the location and is the basis for their classification. Cystic hygromas, on the other hand, arise from lymphatic tissue in areas where expansion can occur and large multiloculated cystic spaces can develop[(4)]. The incidence of lymphangiomas has been reported to range from 1.2 to 2.8 per 1000 newborns.[(9)]

Clinically, lymphangiomas are slow growing and painless soft tissue masses. These frequently present without a clear anatomic outline, dissect tissue planes and can be more extensive than

anticipated. Intraosseous lymphangioma have been reported. Occasionally, they may undergo a rapid increase in size secondary to inflammation from an infection or hemorrhage from trauma. Large lymphangiomas may become life threatening if they compromise the airway or vital blood vessels. [(10)] Accurate delineation of lesion extension is important for diagnosis, surgical planning and assessing recurrence. Plain radiography and barium study show masses displacing adjacent organs. Ultrasound examination shows uni- or multilocular cystic masses with smooth, thin or irregular, thick walls. The CT density of the fluid ranges from -4 to 34 HU depending on the lipid content and the presence of hemorrhage. The cysts are isointense to muscle on T1-weighted and hyperintense to fat on T2-weighted MR images. The MR imaging delineates the tumour lesion extension more clearly than ultrasound and CT scans [(11)]. Diagnostic workup includes biopsy for histopathologic examinations, which will confirm the diagnosis, ultrasonography will detect the cystic nature and fluid component of a lymphangioma and angiography will rule out vascular lesions [(12)].

Traditionally management involves surgical removal of involved tissue without sacrificing vital structures. Due to the anatomical relationship in the head and neck, extirpation can be cosmetically and/or functionally morbid. Nonsurgical therapies are diathermy, cryotherapy, radiation therapy, fibrin glue, and percutaneous sclerosants. Sclerosants that have been used include sodium morrhuate, dextrose, hypertonic saline, tetracycline, doxycyline, acetic acid, ethanol, boiling water, and OK-432.[(13)]Partial excision or marsupialization is associated with a risk of hemorrhage, infection or lymphatic fistula. Nevertheless, surgical excision is associated with high recurrence rates after complete (~15%) and incomplete (~50%) excision. However, there has been some success with factor OK432, a protein derived from a culture of *S. pyogenes.* The OK432 leads to increased endothelial permeability and lymph drainage through white blood cell activation and cytokine production. The optimal treatment is complete surgical excision with microscopically-free margins, but this may change in the near future as additional work with novel therapies continue[(14)].

CONCLUSION

Lymphangioma should be considered under differential diagnosis when accosted with a soft tissue swelling of the buccal mucosa. Earlier diagnosis helps in providing better treatment options to the patient.

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