

## Case Report

**Primary Lymphangioma of Palatine Tonsil-A Rare Case Report**

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**Abstract:** Lymphangiomas are uncommon benign tumours of tonsils. Lymphangiomas are benign lymphatic tumours typically composed of dilated lymphatic channels which often occur in the head and neck region. Primary lymphangioma of tonsil have been reported rarely in the literature which presents as a mass lesion. Histopathological confirmation is necessary for diagnosis. We reported a case of primary lymphangioma of the tonsil in an 15 year old female patient who presents with difficulty in swallowing and clinically diagnosed as tonsillar cyst.

**Keywords:** Lymphangiomas, benign tumour, tonsils

**INTRODUCTION**

Benign tumors of the tonsils occur infrequently. The most frequently reported benign tumors of the tonsil are papillomas, angiomas, fibromas, myxomas, lipomas, chondromas, inclusion cysts and teratogenous cysts [1]. Histopathological confirmation is essential for diagnosis. Lymphangiomas are rare congenital tumors of the lymphatic system. Although they are usually present at or around the time of birth, they usually manifest within the first two decades of life.

Three types of lymphangiomas in the head and neck region may be distinguished : (1) Lymphangioma simplex, which is composed of thinwalled capillary-sized lymphatic channels; (2) cavernous lymphangioma, which in almost half of the cases occurs in the tongue; (3) cystic hygroma.

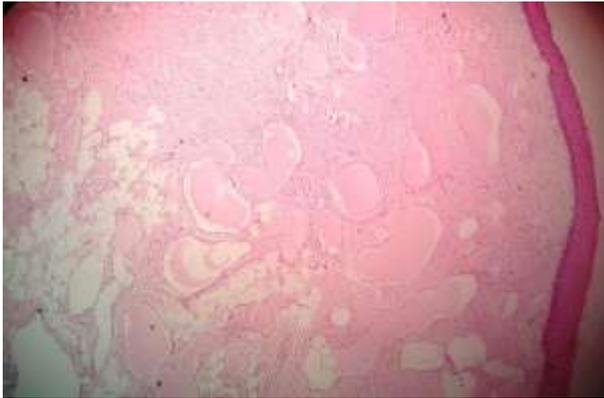
**CASE REPORT**

A 15-year old female patient presented with a 2-month history of recurrent fever, discomfort in oropharynx .She had been treated with several courses of oral antibiotics. She was having constant right sided pharyngeal discomfort. She was otherwise normal with no other significant clinical findings. Examination of oropharynx revealed right sided tonsillar enlargement with polypoidal growth. The left tonsil appears to be clinically normal. Patient underwent right side tonsillectomy under general anesthesia.

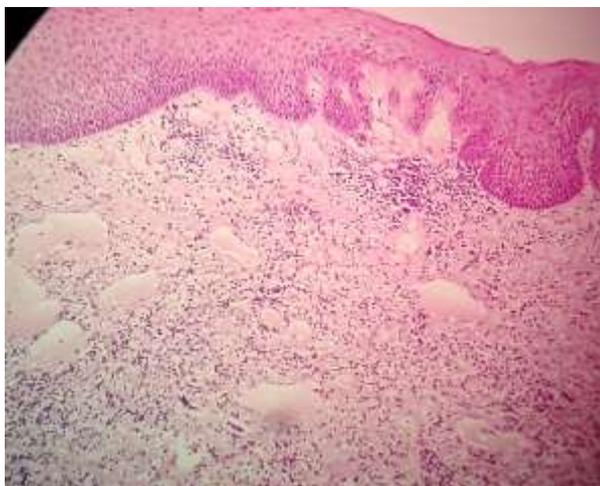
Gross examination revealed a single pearly white polypoidal tonsillar mass measuring  $2.0 \times 1.5 \times 0.5$  cm (Gross Fig-1). Cut section shows homogenous white area with tiny cystic spaces and yellowish areas. Microscopy showed stratified squamous epithelial lining with underlying tissue shows multiple dilated lymphatic spaces lined by endothelial cells and filled with lymph (Fig-2). The surrounding stroma was infiltrated by lymphocytic aggregates, fibrous tissue stroma and mature adipocytes (Fig-3).



**Fig-1: Single, pearly white, polypoidal soft tissue mass measuring about 2x1.5x0.5 cms Cut section – homogenous white appearance with tiny cystic spaces and yellowish areas**



**Fig-2: (High power, 40X) shows numerous dilated lymphatic channels with fibrous stroma & mature adipocytes**



**Fig-3: (Low power, 10X) Stratified squamous epithelium with underlying stroma infiltrated by extensive lymphocytic aggregates**

## DISCUSSION

Lymphangiomas are benign tumours which have been reported to occur rarely in the tonsil. Lymphangiomas occur in the head and neck region in 90 % of cases, being located primarily in the skin or subcutaneous tissue. They classically occur in association with congenital malformations of the lymphatics, presenting in childhood as cystic masses (cystic hygroma). They may also occur in adults in various locations including the oral cavity, tongue, larynx and parotid gland. The majority of previously reported cases involving the palatine tonsils were pedunculated or polypoidal lesions with histological features consistent with those noted in this case.

Al Samarrae *et al* [2] reported 2 cases of this disease in 1985, and in a review of the literature, they found only 6 well-documented cases previously reported. Since then, a few more cases in adults [3, 4], and occasionally in children, were published [5, 6]. Recently, Chen *et al* [7] reported bilateral

lymphangiomatous polyps of the palatine tonsils in a 4-year-old girl.

Surgical excision (tonsillectomy) is definitive management for these lesions, with patients undergoing an uncomplicated post-operative recovery. There have been no reported instances of disease recurrence after complete excision. The pathogenesis of tonsillar lymphangioma is controversial. Chronic inflammation and associated obstruction of lymphatic channels have previously been suggested as a possible mechanism, with congestion eventually leading to mucosal prolapse and the appearance of a polypoidal swelling [8].

However, evidence against this explanation is that chronic tonsillitis occurs much more commonly than tonsillar lymphangiomas, and many patients presenting with tonsillar lymphangioma lack a history of tonsillitis. The recurrent sore throats some patients experience could be a consequence, rather than a cause, of a tonsillar lymphangioma [9]. Furthermore, a study of 14 cases of tonsillar lymphangioma found no gross evidence of lymphatic obstruction in the lesions or associated resected normal tonsils, even after serial sectioning of the tissue specimens [10]. This suggests that another mechanism is likely responsible for the pathogenesis of these tumors.

The differential diagnosis of a benign polypoid tonsillar lesion includes squamous papilloma, lymphangioma, epidermal inclusion cyst, juvenile angiofibroma, hemangioma, fibroepithelial polyp, fibroma, fibroxanthoma, lipoma, adenoma, and chondroma [10]. Of these benign entities, the most common is squamous papilloma, which is a proliferation of the surface epithelium without involvement of the underlying stroma and with absence of lymphatic and lymphocytic components [1].

Hemangioma, fibroma, lipoma, adenoma, and chondroma refer to benign proliferations of blood vessels, fibrous tissue, mature fat cells, glandular elements, and hyaline cartilage, respectively [10]. Routine hematoxylin-eosin sections are sufficient for making the diagnosis of tonsillar lymphangioma, but Kardon *et al* [1] used immunohistochemical staining to determine the immunoprofile of these lesions. In the 15 cases examined, the endothelium and subendothelium of the lymphatic channels stained positive for factor VIII-related antigen, and the majority of channels stained positive for either anti-CD31 or anti-CD34.

Additionally, the walls of the dilated lymphatic vessels expressed smooth muscle actin. As far as the lymphoid population of cells, the leukocyte markers CD3, CD20, and CD45 (leukocyte common antigen) revealed a distribution of lymphocyte expression consistent with tonsillar tissue. Intraluminal and

intraepithelial lymphocytes were predominantly CD3 immunoreactive. Thus, Kardon *et al* [1] confirmed the endothelial origin of the vascular proliferation and a mixed lymphoid population.

In summary, lymphangioma of the palatine tonsil presents as a mass lesion constituting a benign proliferation of dilated lymphatic spaces amid stroma with fibrous, lymphoid, and adipose tissue components, covered by stratified squamous epithelium. The histologic features, which suggest that the lesion represents a hamartomatous proliferation, allow differentiation from other possible entities included in the differential diagnosis of a tonsillar mass.

### CONCLUSION

A rare case of Primary Lymphangioma of palatine tonsil has been reported as an incidental finding. Because of the low incidence of this lesion, which accounts for only 1.9%, it is the greatest diagnostic challenge for operating surgeon and pathologist. However, wide surgical excision with Tonsillectomy facilitate excellent prognosis, thus the identification of this lesion at an early stage becomes mandatory.

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