

Primary lymphangioma of palatine tonsil

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Abstract

Lymphangiomas are benign congenital tumors of the lymphatic system. Tonsillar lymphangiomas are extremely rare. Histopathological confirmation is essential to make the correct diagnosis. We report a case of primary lymphangioma of the left tonsil in a 17-year-old male patient who presented with difficulty in swallowing and was clinically diagnosed as a tonsillar cyst.

Keywords: Benign, lymphangioma, tonsil, tumor

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INTRODUCTION

Benign tumors occur infrequently in the tonsils. The most commonly reported benign tumors of the tonsils are papillomas, angiomas, fibromas, myxomas, lipomas, chondromas, inclusion cysts and teratogenous cysts.^[1] Tumors of the lymphatic system have very few documented cases in the world literature. Lymphangiomas are rare benign congenital tumors of the lymphatic system with an incidence of 1.2–2.8/1000 in children.^[2] Although they are usually present at or around the time of birth, they 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 thin-walled capillary sized lymphatic channels; (2) cavernous lymphangioma, which in almost half of the cases occurs in the tongue; (3) cystic hygroma.^[3] They present as unilateral or bilateral tonsillar outgrowths and cause a large spectrum of symptoms related to local irritation and airway obstruction.^[4] Their pathogenesis has not been clarified and multiple theories have been proposed.

Histopathological confirmation is essential for its correct diagnosis.

CASE REPORT

A 17-year old male patient presented to the ENT OPD with a 2-month history of recurrent fever and difficulty in swallowing. He had received several courses of oral antibiotic therapy but had a persistent left-sided pharyngeal discomfort. No other significant clinical findings were noted. Examination of the oropharynx revealed left-sided tonsillar enlargement with a large, pale polypoidal growth. It was non-tender, non-friable and did not bleed on touch. CT scan revealed a poorly circumscribed, multiloculated hypodense mass with fluid accumulation. The mass was clinically diagnosed as a tonsillar cyst and polyps, parapharyngeal masses were considered as close possible differential diagnoses. It was attached to the lower pole of the left tonsil by a narrow elongated stalk. The right tonsil appeared to be clinically normal. The left tonsil and the pedunculated mass were removed by surgery and sent for histopathological study.

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Gross examination showed an exophytic polypoid nodule measuring by 3.7 cm × 2.6 cm × 0.8 cm. Cut section shows grayish-white area with tiny cystic spaces [Figure 1]. Microscopy showed tonsillar tissue with underlying multiple dilated lymphatic spaces [Figure 2a]. The cystic spaces lined by endothelial cells were filled with lymph and lymphocytes [Figure 2b]. The surrounding stroma was infiltrated by lymphocytic aggregates, fibrous tissue and mature adipocytes [Figure 2c].

DISCUSSION

Benign vascular tumors of the tonsils are rare. Lymphangioma of the tonsil is a rare benign tumor of the lymphatic system. Al Samarrae *et al.*^[5] reported 2 cases of this disease in 1985, and in a review of the literature, they found only 6 well-documented cases previously reported. Then, a few more cases in adults,^[5,6] and occasionally in children, were published.^[7,8] Chen *et al.*^[4] reported bilateral lymphangiomatous polyps of the palatine tonsils in a 4-year-old girl. The clinical behavior of the tumor is largely unknown, as most of these lesions are diagnosed histologically after surgical excision of the tonsils. In our patient, the tumor was large oval, approaching the size of the tonsil, and although protruding into the oral cavity, the patient reported recent manifestation of symptoms, such as dysphagia and foreign-body sensation.

Lymphangiomas occur mostly during the first two decades of life and can sometimes be large. The mean age of the patient was 21.1-year-old with a male-to-female ratio of 6:2. Many authors believe that the true incidence may be higher than reported.^[4,5,8] The most common presenting symptoms are dysphagia and sore throat.^[1,3] Lymphangiomas may be asymptomatic and can be very often an incidental finding. Most of the time, the tumor appears as a painless mass.

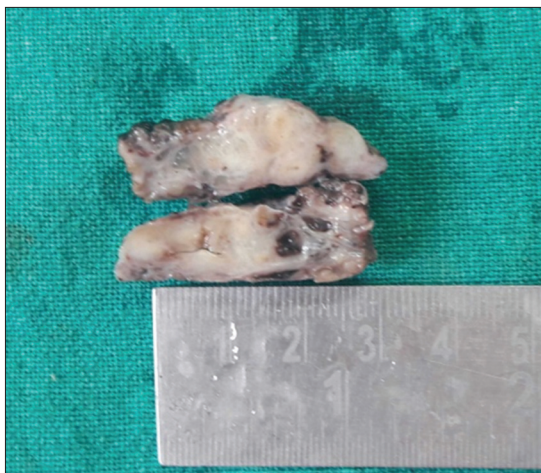


Figure 1: Gross: Greyish polypoidal mass, cut section showing greyish-white areas with cystic spaces

If it is large, it can affect the surrounding vital structures to produce rhinolalia clausa, respiratory difficulty, stridor and difficulty in swallowing. The history and the clinical examination are important, but histological examination is needed to establish the diagnosis. The pathogenesis of tonsillar lymphangioma is uncertain and three theories have been proposed to explain it.^[9]

Failure of the primordial lymphatic sacs to drain into the veins

According to this theory, the failure of the lymphatic sacs to drain into the veins leads to dilated lymphatic channels.^[10]

Abnormal sequestration of lymphatic tissue

It has been hypothesized that abnormal sequestration of lymphatic tissue occurs early in embryogenesis. This theory helps explain the morphology of the more peripheral lesions, such as capillary and cavernous lymphangiomas.^[11]

Abnormal budding of the lymphatics

According to this theory, these aberrant buds lose their connections with the lymphatic primordial and eventually canalize to form lymph-filled cysts. These cysts maintain their ability to branch and grow and do so in an uncontrolled, disorderly manner.^[11,12]

Differential diagnosis of lymphangioma includes other benign lesions like squamous papilloma, epidermal inclusion cyst, juvenile angiofibroma, hemangioma, fibroepithelial polyp, fibroma, fibroxanthoma, lipoma, adenoma and chondroma,^[13] acute or chronic tonsillitis, parapharyngeal masses which may give rise to apparent

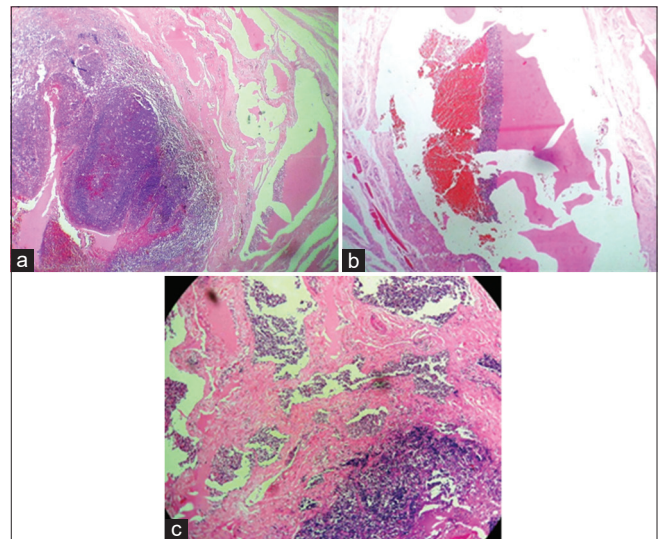


Figure 2: (a) Microscopy showing tonsillar lymphoid follicles with underlying tissue showing multiple dilated lymphatic spaces (x100). (b) Microscopy showing cystic spaces filled with lymph and lymphocytes (x400). (c) Microscopy showing surrounding stroma infiltrated

tonsillar enlargement due to medial displacement of the tonsil. Of these benign entities, the most common is squamous papilloma, which is a proliferation of the surface epithelium without the involvement of the underlying stroma and with the absence of lymphatic and lymphocytic components.^[1] Surgical excision is the choice of treatment in tonsillar lymphangiomas. Recurrence after surgical excision is a rare phenomenon.

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. In addition, the walls of the dilated lymphatic vessels expressed smooth muscle actin. In the lymphoid population of cells, the leukocyte markers CD3, CD20 and CD45 (leukocyte common antigen) revealed a distribution of lymphocyte expression. 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.

CONCLUSION

This case demonstrates a case of tonsillar lymphangioma occurring in a 17-year-old boy. Though it is rare, we should consider this entity in the differential diagnosis of tonsillar masses in childhood. Histopathology is the gold standard to arrive at the correct diagnosis. As wide surgical excision with tonsillectomy facilitates excellent prognosis, so, it is a great challenge to the clinician and pathologists to diagnose this lesion at an early stage.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal.

The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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