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CASE REPORT

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Lymphatic malformation in larynx masquerading as respiratory papillomatosis

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ABSTRACT

Introduction: Lymphatic malformations are benign congenital lesions of abnormal lymphatic channel common in head and neck region. Extensive disease involving the larynx and presenting as a laryngeal pathology is a rare occurrence and can cause diagnostic enigma.

Case report: We report an unusual case of an extensive mixed cystic type of lymphatic malformation in the larynx without any external neck or oral swelling presenting in stridor and clinically mimicking as juvenile respiratory papillomatosis. Endoscopic debulking and tracheostomy were done and subsequent MRI showed lymphatic malformation. After the failure to inject doxycycline sclerotherapy properly, definite surgery was planned. However, the patient was lost to follow up.

Discussion: Lymphatic malformation extending into the larynx is a rare cause of airway problem. History and clinical examination alone is not always adequate in every case. In the case of an unusual presentation, the final diagnosis should include appropriate imaging and should be confirmed by histopathology.

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Lymphatic malformation; stridor; larynx; child

Introduction

Lymphatic malformations are benign congenital lesions with abnormal lymphatic development and commonly presents as a cystic lesion in the head and neck region [1].

ISSVA classifies lymphatic malformations (LM) into various subtypes among which Common (cystic) LM is further classified as macrocystic, microcystic, and mixed cystic [2]. The diagnosis of lymphatic malformations is made by clinical examination finding of mobile, cystic, fluctuant, doughy mass with positive transillumination test but difficult sites, secondary hemorrhage, and unusual presentation can create a diagnostic confusion [3]. Lymphatic malformation spreading to the larynx and causing obstructive symptoms is a rare presentation with very few reported literatures [4] and can mimic other causes of airway obstruction and stridor in children.

Various theories have been postulated regarding the formation of lymphatic malformation without any reliable evidence. Some hypotheses are failure of primordial lymphatic sacs to connect to the venous system, deposition of lymph tissues in an incorrect area during embryogenesis failing to connect with the normal lymph system, and tumor-like proliferating lymphatics forming a new network of lymph channel [5].

There are various manifestations of lymphatic malformations depending upon the anatomical location, size, and local compressive symptoms. In the head and neck region, the problem ranges from cosmetic issues, change in voice, swallowing problem to life-threatening airway problem due to compressive symptoms and can mimic other benign causes of the larynx [4].

Case report

A four-year-old boy from the remote village who presented to the outpatient clinic with the chief complaint of noisy breathing since seven months of age which was insidious in onset and continuous and harsh in quality. There was aggravation in noisy breathing on exertion, URTI, and got relieved by rest and medication. There was an increase in the severity of noisy breathing for the last eight days with

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shortness of breath. Parents reported a history of aspiration of brownish colored fluid from left lateral neck swelling at the age of seven months for the swelling. There were no written documents available.

He was admitted two weeks back in a local hospital in the intensive care unit with the diagnosis of Croup and was under mechanical ventilation for three days. He had no history of Asthma, Pulmonary tuberculosis, or any other chronic illness in the past. Flexible Nasopharyngolarygnoscopy in the clinic showed multiple papillomatous growths in the bilateral vallecula, the laryngeal surface of the epiglottis, bilateral aryepiglottic fold, and inter arytenoid area. We could not access the endolarynx properly in the flexible nasopharyngolarygoscopy in an outpatient clinic (Figures 1 and 2). We planned for emergency debulking under general anesthesia with a provisional diagnosis of Juvenile respiratory papillomatosis. Direct laryngoscopy under general anesthesia showed multiple cystic mass presents over bilateral vallecula, lingual and laryngeal surfaces of epiglottis (more on the left side), bilateral arytenoids and aryepiglottic folds, bilateral false vocal cords, lateral pharyngeal walls (more on the left side). But bilateral true vocal cords, subglottic area, and trachea were free of disease.

We planned for endoscopic removal of the mass. The tissue was too tough to debulk, so only partial debulking was done followed by a tracheostomy to avoid further respiratory compromise before final definite treatment.

Contrast-Enhanced MRI was suggestive of the mixed cystic type of common (cystic) LM which demonstrated an ill-defined cystic mass in the region of the posterior wall of the hypopharynx measuring 3.8×2.5 cm. Mass was infiltrating the bilateral vallecula, epiglottis, and bilateral aryepiglottic folds. Bilateral pyriform fossa sinus was obliterated with severe narrowing of the supraglottic airway. The cystic mass was extending along the left lateral pharyngeal wall of the oropharynx, hypopharynx, left submandibular spaces displacing and encasing the gland. Inferiorly, it was extending along with the left anterior strap muscles into the thoracic inlet. Another 2.6×1.4 cm ill-defined cystic area was also noted in the left paratracheal region with mass effect and right lateral displacement of the trachea (Figures 3 and 4).

The histopathology report showed multiple fragments of tissue lined by hyperplastic squamous epithelium with dense inflammatory infiltration. Focally, few fragments show dilated vascular channels lined by endothelial cells in the stroma and epithelium consistent with a lymphatic malformation (Figure 5).

Discussion

Laryngeal lymphatic malformation is rare with a few literature mentioning larynx as an isolated site [4,6,7]. Lymphatic malformation from the cervicofacial region has a possibility of extension to the larynx. The largest series of 160 patients found laryngeal involvement in ten patients [6]. The diagnosis of lymphatic malformation is based on history, clinical findings, and imaging. In our case, the diagnosis initially was based on history and clinical assessment alone, so, the misdiagnosis of Juvenile respiratory Papillomatosis was made. The uncommon sites and presentations can be

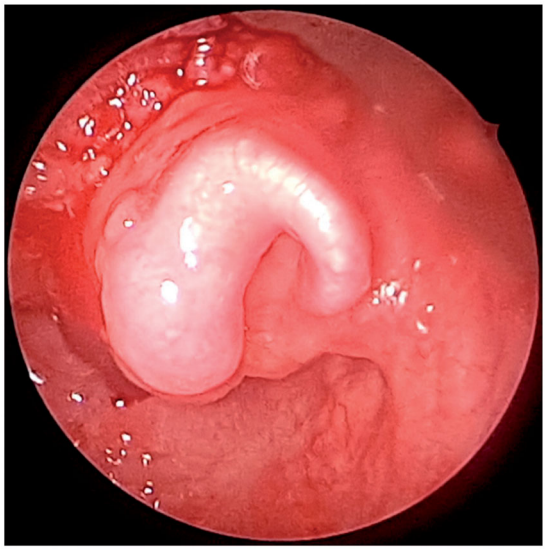

Figure 1. Endoscopic finding of papillomatous lesion in epiglottis, base of tongue, pyriform sinus but endolarynx could not be seen.

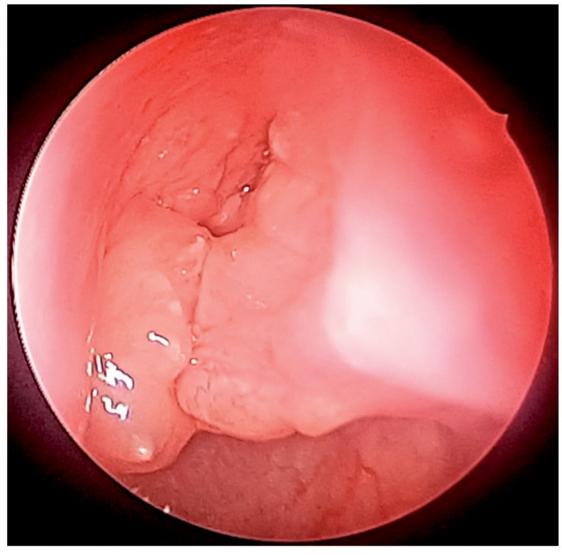

Figure 2. Endoscopic image of the lesion with irregular and papillomatous appearance.

Figure 3. Sagittal imaging of MRI showing enhancing lesion around oropharynx, hypopharynx, trachea and oesophagus.

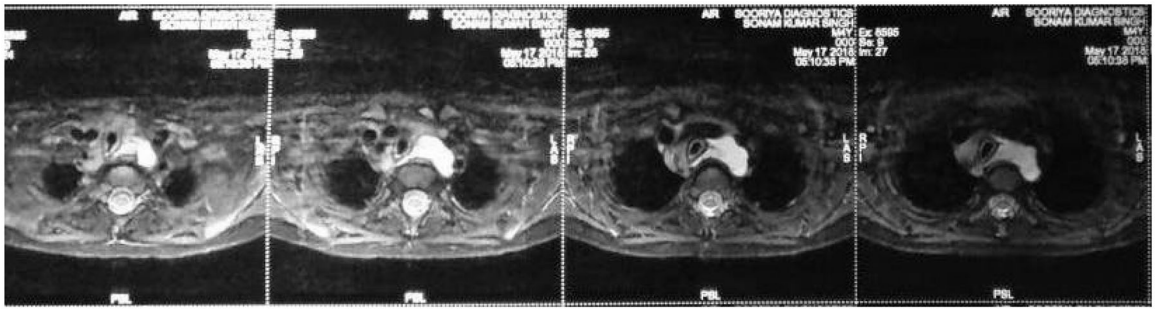

Figure 4. Axial imaging of MRI showing enhancing lesion around trachea and oesophagus.

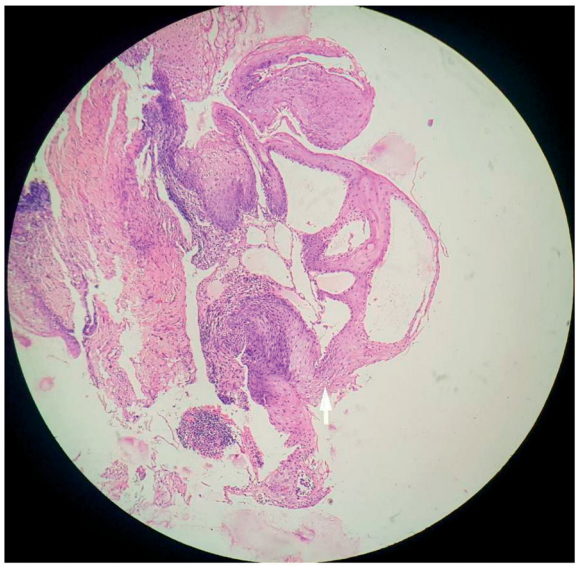

Figure 5. Histopathology section showing dilated vascular channels lined by endothelial cells in the stroma and epithelium.

usually missed if the diagnosis is based on one modality. The presentation of stridor after an upper respiratory tract infection which was constant and showed no alteration during sleep or with activity-associated difficulty feeding was missed as laryngomalacia [7]. Some studies in adults even reported having symptoms suggestive of acute epiglottitis [8].

The problem arises more especially in syndromic children where history is unreliable and clinical examination is not always possible. A syndromic child with progressive dyspnoea for lymphatic malformation involving the epiglottis has been reported by Claros et al. [9].

Regarding the role of imaging, in clinically evident cases, sonography and Contrast-enhanced CT scan can give detailed information of the lesion. MRI is the investigation of choice and delineates the extent of the lesion from the normal tissues along with the content, size of lymphatic malformation in the head and neck region.

The treatment of lymphatic malformation can be through surgical excision methods such as conventional methods, laser, and non-surgical methods using sclerosing agents like OK432, doxycycline, steroids, etc. [10]. The management depends on the age of presentation, comorbidities present, cyst type, extent, location of the lesion with studies showing varying results for each option [5]. Sclerotherapy and surgical excision are preferred in macrocystic type over microcystic type. Sirolimus, a serine/threonine kinase, has been used recently in complex lymphatic malformation due to its antiangiogenic and antimitotic properties with good efficacy and tolerable side effect [11]. This could be a better non-surgical modality in this case but the overall cost of treatment and the lack of easy availability of the drug was the limiting factor. The primary treatment of sclerotherapy with doxycycline was attempted but could not inject properly as the lesion was very firm and had more microcystic components on MRI.

Since complete surgical excision in the laryngeal lesion was not possible in our case, we planned to do endoscopic excision after securing the airway by tracheostomy and after further assessment by MRI for the extension. However, the patient lost to follow up because of the covid pandemic and definite management is yet to be performed. Our further plan is to do an endoscopy and re-evaluate the airway and perform endoscopic laser excision/marsupialization and do regular follow up to check the recurrence.

The management of the lymphatic malformation is challenging with a high recurrence rate even after complete excision. There is a high recurrence rate (100%) in cases with aspiration as primary modality compared to 40% recurrence rate with partial debulking in laser surgery and only 17% overall (33% in head and neck) with complete excision [12].

We reported this case because of the unusual site, presentation. Moreover, the case was initially missed as respiratory papillomatosis in history and clinical examination and there was a challenge in management. Although rare in occurrence, clinicians should always keep in mind the diagnosis of laryngeal lymphatic malformation as an alternative diagnosis of a common lesion such as laryngomalacia or respiratory papillomatosis with atypical features.

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Informed consent

The written informed consent for publication of images and other information in the case report has been provided by the patient party.

Author contributions

Bijaya Kharel: Conception and design of the work, provision, and collection of study materials, literature review, design of methodology, writing the initial draft. Yogesh Neupane: Literature review, Design of methodology,

collection of operative notes and images, revision of content, final draft preparation.

Disclosure statement

The authors declare no conflict of interest regarding the publication of this case report.

References

[1] Grasso DL, Pelizzo G, Zocconi E, et al. Lymphangiomas of the head and neck in children. Acta Otorhinolaryngol Ital. 2008;28:17–20.

[2] International Society for the Study of Vascular Anomalies. ISSVA classification for vascular anomalies [EB/OL]. Available from: http://www.issva.org/classification., 2018-05-18

[3] Kennedy TL. Cystic hygroma: lymphangioma: a rare and still unclear entity. Laryngoscope. 1989; 99:1–10.

[4] Gupta N, Goyal A, Singh PP, et al. Isolated laryngeal lymphangioma: a rarity. Indian J Otolaryngol Head Neck Surg. 2011;63:90–92.

[5] Kenton A, Duncan N, Bhakta K, et al. Laryngeal lymphatic malformation in a newborn. J Perinatol. 2003;23:567–571.

[6] Cohen SR, Thompson JW. Lymphangiomas of the larynx in infants and children: a survey of pediatric lymphangioma. Ann Otol Rhinol Laryngol. 1986;95:1–20.

[7] Papsin BC, Evans JN. Isolated laryngeal lymphangioma: a rare cause of airway obstruction in infants. J Laryngol Otol. 1996;110:969–972.

[8] Seven H, Topuz E, Turgut S. Isolated laryngeal lymphangioma showing the symptoms of acute epiglottitis. Eur Arch Otorhinolaryngol. 2004;261:548–550.

[9] Claros P, Viscasillas S, Claros Sr A, et al. Lymphangioma of the larynx as a cause of progressive dyspnea. Int J Pediatr Otorhinolaryngol. 1985;9:263–268.

[10] Orvidas LJ, Kasperbauer JL. Pediatric lymphangiomas of the head and neck. Ann Otol Rhinol Laryngol. 2000;109:411–421.

[11] Tian R, Liang Y, Zhang W, et al. Effectiveness of sirolimus in the treatment of complex lymphatic malformations: Single center report of 56 cases. J Pediatr Surg. 2020;55:2454–2458.

[12] Alqahtani A, Nguyen LT, Flageole H, et al. 25 years experience with lymphangiomas in children. J Pediatr Surg. 1999;34:1164–1168.