Case Report

Nasopharyngeal Lymphangioma in an Adult: A Rarity

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Abstract: Lymphangiomas are rare congenital tumors of the lymphatic system, most often encountered during childhood. Although the most frequent locations for lymphangioma are the head and neck region, isolated involvement of the nasopharynx is very rare and only two cases have been reported since 1969. We report a case of a 60-year-old male with a nasopharyngeal mass that presented with nasal obstruction. The surgical excision of mass was performed by a combined transoral and transnasal endoscopic approach. Histopathological evaluation revealed the diagnosis as lymphangioma. After a follow-up of 18 months the patient is free of recurrence.

Key Words: Lymphangioma; nasopharynx; nasal obstruction.

INTRODUCTION

Lymphangiomas are rare congenital tumors of the lymphatic system. They consist of abnormal development of the lymphatic system in localized centers. Lymphangiomas are classified as microcystic (capillary hemangiomas), macrocystic (cavernous hemangiomas), and cystic hygromas according to the size of the lymphatic cavities incorporated. They are usually diagnosed in infancy (50% of all lesions are noted at birth) and develop by 2 years of age and early childhood. They have a marked predilection for the head, neck, oral cavity, axilla, and abdomen. But isolated involvement of the nasopharynx with lymphangioma is very rare. Our literature search for nasopharyngeal lymphangioma displayed no new cases in the last 40 years. Two unique case had been reported in Russian in 1966 and 1969 (no abstract available). We report the case of a 60-year-old male with the diagnosis of nasopharyngeal lymphangioma.

CASE REPORT

A 60-year-old nonsmoker male was referred to an outpatient clinic with complaints of difficulty in nasal breathing. Endoscopic nasal examination revealed a polypoid mass arising from the nasopharynx that narrows the nasal airway. The mass had a smooth globular surface, originating from the right posterolateral nasopharyngeal wall and reaching to the upper pole of the right tonsil (Fig. 1), which appeared by direct oropharyngeal inspection. There was no cervical lymphadenopathy or otologic and blood abnormality.

A magnetic resonance imaging (MRI) of the nasopharynx was carried out. It revealed a soft tissue density at the nasopharyngeal space. The mass was constricting the airway asymmetrically and reaching to the base of tongue. The dimensions of the mass were measured 18 × 21 millimeters (Fig. 2) and the histopathologic analysis was suggested in the assessment of MR.

The physical examination and imaging study of the mass indicated that it could be a benign lesion. A total excision of the mass was performed with combined transoral and transnasal approaches under endoscopic view. The operation was carried out with general anesthesia, and a degree 4 mm telescope was used to visualize the mass transnasally. The mass was excised from the posterior nasopharyngeal wall with the use of bipolar cautery without significant bleeding. A transoral approach, as in tonsillectomy (rose position) was used to excise the elongated component, which reached the upper pole of right tonsil. A rubber catheter was passed through one of the nares and brought out into the pharynx. The catheter was used to retract the soft palate; a clamp on the rubber catheter at the level of the tip of the nose helped accomplish this. The total excision of the mass was completed without any complications. Hemostasis was ensured, and there was no need for packing of the surgical area. Patient was discharged from the hospital at the postoperative second day. The patient was followed up for 18 months. He healed well and reported significant relief of symptoms postoperatively.

Histopathological examination of the specimen revealed multiple cystically dilated channels lined by a flattened endothelium. Scattering lymphocyte was...
present in the stroma (Fig. 3A). Immunohistochemically, it was positive for lymphatic lineage markers D2–40 (Fig. 3B) and was negative for endothelial lineage marker CD31 (Fig. 3C). The histopathological diagnosis was lymphangioma. Warthin’s tumor was encountered in a differential diagnosis. It was ruled out by the lack of oncocytic cells and dense lymphocyte aggregate with germinal centers in histologic specimens.

**DISCUSSION**

Lymphangiomas are uncommon congenital slowly progressive vascular hamartomas. They are usually diagnosed at birth in the majority of cases and only rarely occur after 2 years of age.5 Lymphangiomas present in the head and neck in 48% to 75% of cases, most commonly presenting as asymptomatic posterior triangle masses.

There are limited numbers of articles on the lymphangioma affecting the oral cavity (tongue, palate, gingiva and oral mucosa, and alveolar ridge of the mandible) in pediatric populations in the literature. The occurrence of lymphangioma in an adult is seen infrequently.6 Naidu and McCalla7 reported a comprehensive review of studies from 1828 to 2000 years on lymphatic malformation in adults. They found 91 adult cases. The lymphangiomas were located on the neck in all of these patients. Recently Catalfamo et al.6 reported nine tongue lymphangiomas in adults. Our literature search revealed several sporadic lymphangioma case reports located at the parotid gland, thyroid gland, larynx, and facial region.8–11 But only two cases of lymphangioma located at the nasopharynx had been reported in the Russian language in 1966 and 1969.3,4 The presented case was 60 years old with the diagnosis of nasopharyngeal lymphangioma. As far as we know, this is the first reported case in the literature for this unusual location.
Lymphatic malformations have a growing trend with the age of the child and rarely regress; clinically their location determines the symptoms. Cystic hygromas of the neck may at times be so extensive as to lead to symptoms of airway obstruction.\textsuperscript{10} Common presenting symptoms of lymphangiomas at oral cavity, tongue, and larynx include dysphagia, dyspnea, foreign body sensation, sore throat, tonsillitis, and tonsillar mass. While the nasopharynx is a crucial structure for nasal breathing, lymphangioma at this passage can give nasal obstruction symptoms, as it did in the present case. It may be possible to observe the mass by a simple nasal endoscopic examination. Radiological studies help diagnose the lymphangioma. A MRI can demarcate the mass from the surrounding tissues and give the extent of the lesion (Fig. 2).\textsuperscript{10} The differential diagnosis includes antrochoanal polyps, nasopharyngeal carcinoma, angiofibroma, cystic lesions, and benign other masses. The patient's medical history and examination findings are enough to diagnose the lymphangioma clinically in most of the cases. When it is in an unusual location, the definitive diagnosis can be provided by histopathological examination of the specimen postoperatively.

The potential complications of lymphangioma are infection, hemorrhage, and the mass effect (obstruction of vital structures) associated with rapid growth of the lesion. Also in some patients, lymphocytopenia, a precipitating cause of infection, can be documented.\textsuperscript{12}

Many approaches have been proposed in the treatment of lymphangioma. Unlike hemangioma, lymphangioma rarely regresses spontaneously.\textsuperscript{12} Various treatment options, mainly surgical excision, LASER debulking, sclerotherapy, or corticosteroids have been proposed.\textsuperscript{10} Neodymium: yttrium-aluminum-garnet (Nd-YAG) LASER surgery has become a widely preferred method because of its advantages, less bleeding and edema versus conventional surgical resection.\textsuperscript{12} Sclerosing agents cause fibrosis and are currently under investigation as a possible modality of treatment for laryngeal lymphangiomas. Brennan et al.\textsuperscript{13} reported that sclerosing agents are ineffective, probably as a result of the discontinuous basement membrane of the lymphatic vessels.\textsuperscript{14} In addition, this form of therapy has a risk of damaging the adjacent vital soft-tissue structures. Radiation therapy is not effective in the treatment of these lesions and is associated with secondary malignancies.\textsuperscript{10}

Surgery has been the main treatment option for the most lymphangiomas. Since lymphangiomas are progressive, the treatment is mandatory. On the other hand, some clinicians do not recommend the surgery for non-enlarging lymphangiomas because of the difficulties in removal and the high recurrence rate.\textsuperscript{12} Surgery should be so extensive as to provide complete excision and minimize the risk of local recurrence, but it should also be so conservative as to preserve the adjacent vital structures. Surgeon must have enough knowledge about the anatomy and extensiveness of the lesion.

In the reported case, we were able to excise the lesion without causing any damage to the surrounding structures of the nasopharynx by a combined transoral and transnasal approach under endoscopic view.

We report this case because of the unusual involvement site and the presentation age of patient. Although nasopharyngeal lymphangioma is very rare, otolaryngologists should keep in mind the lymphangioma in the differential diagnosis of nasopharyngeal lesions.

**CONCLUSION**

Although lymphangioma is very rare in the nasopharynx, it should be considered for the patient with nasal obstruction symptoms and for the differential

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Fig. 3. Histopathological examination of the specimen. (A) Dilated lymphatic vessels of different size with dense fibrous wall, which are lined by a flattened endothelium H&E x100. (B) Immunohistochemistry, positive for lymphatic lineage markers D2–40 \( \times 100 \). (C) Immunohistochemistry, negative for endothelial lineage markers \( \text{CD}31 \times 100 \). [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]
diagnosis of nasopharyngeal lesions. Complete surgical excision with the preserving of the adjacent structures is the treatment of choice. Since the lymphangioma have a risk of recurrence, the patient should be followed up regularly.

BIBLIOGRAPHY