A Rare Cause of Snoring: Isolated Nasopharyngeal Lymphangioma

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Abstract
Lymphangiomas are rare congenital tumors of the lymphatic system. They are most often seen in childhood and frequently are located in the head and neck region. Nasopharyngeal involvement of an isolated lymphangioma is extremely rare, with very few cases reported in the literature. Together with the relevant information in the literature, we present here a case of a 40-year-old female who presented at our polyclinic with complaints of snoring and obstruction in the right ear and was diagnosed with isolated nasopharyngeal lymphangioma.

Keywords
Lymphangioma; Nasopharynx; Snoring; CD31; CD34

Introduction

Lymphangiomas are rarely seen congenital tumors of the lymphatic system. They are most frequently seen in childhood and generally diagnosed in infancy; approximately 90% of cases are seen by the age of 2 years [1]. According to the size of the cavity, lymphangioma can be classified as microcytic, macrocytic, or cystic hygroma. These tumors are seen most often in the head and neck region and more rarely may be observed in the axilla and abdomen. As isolated nasopharyngeal lymphangiomas are extremely rare, few cases have been reported in the literature since the first case in 1966 [1,2].

In the light of information in literature, we present the case of a 40-year-old patient diagnosed with isolated nasopharyngeal lymphangioma, who underwent surgery and had no complaints during a 1-year follow-up period.

Case Report

A 40-year-old female presented at the polyclinic with complaints of intermittent obstruction of the right ear and snoring which had been ongoing for one year. In the endoscopic nasal examination, a polyposis mass was determined originating from the right posterolateral nasopharynx and extending to the oropharynx, narrowing the nasopharyngeal cavity (Figure 1). The neck examination and other ENT examinations were normal. No additional developmental anomaly was determined in the patient.

On the computed tomography (CT) examination, evident thickening was determined in the torus tubarius in the nasopharynx right hemisphere and obliteration in the Rosenmüller fossa. On magnetic resonance imaging (MRI) of the nasopharynx, a mass was determined, 16 x 12 x 25 mm in size, filling the Rosenmüller fossa in the right nasopharynx and showing extension to the right lateral wall in the inferior oropharynx (Figures 2, 3).

Under local anesthetic and 0° endoscopy guidance, multiple punch biopsies were taken from the mass, which was hard, smooth-surfaced, and originated in the right posterolateral nasopharynx. In the histopathological examination, reactive changes were determined. As the pathology and the clinical findings were not compatible, multiple punch biopsies under sedation were taken for a second time from the hard mass originating from the right Rosenmüller fossa. In the histopathology report, fibroblast proliferation was observed in the lymphatic-rich tissue samples and there was no necrosis or mitotic activity. Later, under general anesthesia and 0° endoscopy guidance, the mass in the right posterolateral nasopharynx was almost completely excised, protecting the surrounding tissues. Bleeding control was achieved with bipolar cauterization. The patient was discharged on postoperative Day 1 and was monitored with endoscopy at monthly intervals.

In the histopathological examination of the excised specimen, an increase was observed in fibrin on the surface and in the lymphatic and vascular structures below the epithelium. The lymphatic structures were observed with CD31 staining and the vascular structures with CD34 (Figures 4, 5, 6). The findings were consistent with lymphangioma.

Sclerotherapy was recommended for the remaining residual tissue, but the patient refused any further treatment as she had...
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no complaints. The patient was monitored postoperatively with monthly endoscopic examinations. No growth was determined in the mass which had been reduced in the right lateral nasopharynx (Figure 7). The patient complaints of fullness in the ear and snoring abated. On the nasopharyngeal MRI taken 6 months after the reduction of the mass, an increase was determined in asymmetrical thickness in the Rosenmüller fossa in the right nasopharynx extending to the right lateral wall. The patient had no complaints during the 1-year clinical follow-up period, and no further growth was observed in the mass.

Discussion

Lymphangioma are benign hamartomatous malformations of the lymphoid vessels. These malformations may be congenital or develop later. However, 90% are seen under the age of 2 years and only extremely rarely in adults. Those that are congenital often accompany chromosomal anomalies such as Turner syndrome and diagnosis is made in the antenatal period with fetal ultrasound. Here we report a case of a 40-year-old with a nasopharyngeal lymphangioma. Lymphangioma that develop later occur as a result of trauma, inflammation, or lymphatic obstruction. There was no trauma story and no chromosomal abnormalities in our present case. Although lymphangioma may occur in any area of the body, they are most frequently seen in the head and neck area. However, isolated lymphangioma with nasopharyngeal location are extremely rare, with two cases published in the Russian literature in 1966 and 1969 [3,4] and two cases in the English literature in 2013 and 2014 [1,4]. To the best of our knowledge, the case presented in this paper is only the 5th reported case in the literature of isolated nasopharyngeal lymphangioma. The previous two most recent cases were males and the current case was a female [1,4].

The symptoms of oral cavity and larynx lymphangioma are dysphagia, dyspnea, the feeling of a foreign body in the throat, throat pain, and frequent attacks of tonsillitis. The cases reported in the literature of isolated nasopharyngeal location presented at ENT polyclinics with complaints of nasal obstruction and irritation in the throat. As in the current case, nasopharyngeal lymphangioma may appear with findings of Eustachian dysfunction. Unlike previous cases in the literature associated with obstruction, the primary complaint on presentation of the current case was snoring.

Endoscopic visualization of the mass is sufficient for advanced testing. Radiological imaging is helpful in the diagnosis of lymphangioma. MR can differentiate the mass from surrounding tissue and can provide information about the size of the mass [5]. The size of the mass in this case was similar to those in previously-reported cases. In the differential diagnosis, nasopharyngeal carcinoma, nasopharyngeal angiofibroma, nasopharyngeal cystic lesions, and other benign masses of the nasopharynx should be considered. In the current case, a polypoid mass was observed filling the right Rosenmüller fossa; this
requires histopathological evaluation for a definitive diagnosis. In the current case, diagnosis could be made only with almost total excision of the mass under general anesthesia, following two previous punch biopsies. Most lymphangiomas represent malformations rather than true neoplasms and are thought to result from failure of the lymphatic system to communicate with the venous system. Microscopically, lymphangioma consist of large lymphatic channels growing in loose connective tissue. A few disorganized bundles of smooth muscle can be present in the wall of the larger channels. Focal areas of papillary endothelial proliferation similar to those described in blood vessels are sometimes found [6]. Large collections of lymphocytes may be present in the stroma and cause mistakes in interpretation. Lymphangioma almost never become malignant and are curable by excision [7]. In the current case, fibrin was observed on the surface along with an increase in lymphatic and vascular structures below the epithelium; the lymphatic structures were observed with CD31 staining and the vascular structures with CD34.

Lymphangioma are progressive and do not spontaneously regress like hemangioma. Therefore, the disease must be treated. Various methods are used in the treatment of lymphangioma such as surgical excision, reducing the mass with lasers, sclero-therapy, and corticosteroids [5]. As bleeding and edema are less in the laser method, it has recently become a preferred method. Various sclerosing agents such as sodium morrhuate, dextrose, tetracycline, doxycycline, bleomycin, ethibloc (alcohol), and OK432 (lyophilized incubation mixture of group A Streptococcus pyogenes of human origin) are effective by inducing fibrosis [8]. Surgery is the basic treatment option for most lymphangioma. However, as lymphangioma do not have a capsule, most clinicians do not recommend surgery for non-growing lymphangioma because of the difficulties in protecting adjacent vital structures in surgical intervention and high recurrence rates. In the current case, surgery was recommended because the mass was narrowing the passage and because of the patient’s complaints. With a transnasal approach with O° endoscopy protecting the surrounding structures, the mass in the nasopharynx was almost totally excised. In the 1-year follow-up period, there were no clinical complaints and no significant mass was observed in the endoscopic view of the nasopharynx. As the longest follow-up period for recurrence in the literature is 18 months, long-term follow-up results have not yet been reported [1].

In conclusion, although lymphangioma in the nasopharynx are rare, they do occur. This must certainly be kept in mind in the differential diagnosis of patients with symptoms of nasal obstruction and patients seen to have a mass lesion in the nasopharynx. In treatment, a surgical method that protects adjacent vital structures must be used. As there is a risk of recurrence of lymphangioma, patients must be regularly monitored with endoscopy and radiology.

Competing interests
The authors declare that they have no competing interests.

References

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