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Lymphatic Malformation of the Nasopharynx in a Young Pregnant Female: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCE 1 Yehuda Galili
BEF 1 Meghan Lytle
BD 2 Kataria Amandeep
BCF 2 Jonathan Bartolomei
DE 3 Li Ge
AEF 4 Steve J. Carlan
AEF 1 Mario Madruga

1 Department of Internal Medicine, Orlando Regional Healthcare, Orlando, FL, U.S.A.
2 Department of Internal Medicine, Florida State University College of Medicine, Tallahassee, FL, U.S.A.
3 Department of Pathology, Orlando Regional Healthcare, Orlando, FL, U.S.A.
4 Department of Obstetrics and Gynecology, Division of Academic Affairs and Research, Orlando Regional Healthcare, Orlando, FL, U.S.A.

Corresponding Author: Steve J. Carlan, e-mail: stevecarlan@gmail.com
Conflict of interest: None declared

Patient: Female, 21
Final Diagnosis: Lymphatic malformation of the nasopharynx
Symptoms: Dysphagia
Medication: —
Clinical Procedure: Mass was localized in the nasopharynx and detached from a broad 1 cm stalk through both the oral and nasal approaches using bipolar cautery
Specialty: Critical Care Medicine

Objective: Congenital defects/diseases

Background: Lymphangiomas represent the focal proliferation of benign, well-differentiated lymphatic tissue. They are most likely congenital, thus more commonly diagnosed at birth and before the age of 2 years. When they are found in adults, they favor the head, neck and axillary region. Rarely do they involve the nasopharynx region or occur in a pregnant patient.

Case Report: A 21-year-old primagravida in the third trimester of pregnancy developed difficulty swallowing that progressed into difficulty breathing over a 1-month period. Imaging and examination suggested a benign mass in the nasopharynx and the patient underwent surgical removal of the stalk with bipolar cautery. The pathology report revealed a simple 4.5×1.5×0.8 cm lymphangioma. She had no fetal compromise during anesthesia.

Conclusions: Surgical removal of a nasopharyngeal lymphangioma during the third trimester of pregnancy is indicated if respiratory obstruction may be a complication.

MeSH Keywords: Lymphangioma, Cystic • Nasopharyngeal Diseases • Pregnancy Outcome • Pregnancy Trimester, Third

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Background

Lymphangiomas represent the focal proliferation of benign, well-differentiated lymphatic tissue. They are referred to as lymphatic malformations (LM) and most commonly occur in childhood. The etiology is not understood, but most likely LMs are congenital. When they occur in adults they are generally asymptomatic depending on size and location. They are rarely reported to involve the nasopharynx in adults and treatment requires surgical removal. LMs in pregnancy have been reported and have even been noted to change in appearance and size presumably secondary to the hematologic and hormonal changes that accompany pregnancy. [1]. LMs, however, have never been reported to occur in the pharynx of the pregnant or postpartum woman. We present a case of a pregnant woman at 32 weeks gestation with a symptomatic nasopharyngeal lymphangioma diagnosed secondary to respiratory obstruction that underwent surgical removal during pregnancy.

Case Report

A 21-year-old gravida 1 female at 32 weeks gestation as determined by a seventh week ultrasound, who had no significant past medical history, presented with complaints of increasing throat irritation and discomfort of approximately 1-month duration. She stated that she had a mass she could pull out of her throat and lay on her tongue. She complained of trouble swallowing, and recently, the mass had made her breathing very difficult. She could not lay supine, and felt that she was struggling to breathe. She was admitted for a likely pharyngeal mass resulting in airway obstructive. She denied chest pain, nausea or vomiting, hemoptysis, ear ache, nose bleeds, or sinus congestion. Her family history was negative for congenital anomalies and she did not use tobacco, alcohol, or illegal substances. Her prenatal course had been uneventful. She denied trauma, radiation, or head and neck surgery. On arrival her vital signs and initial laboratory findings were stable and within normal limits. Magnetic resonance imaging of the face, orbits, and neck revealed an elongated mass extending across the midline from the nasopharynx into the pharyngeal region along an approximately 4 cm length from the adenoidal region to the level of the epiglottis, not extending below the larynx (Figure 1). This was contiguous with the posterior wall of the pharynx and measured approximately 1.5 cm in width and approximately 0.9 cm anterior-posterior dimension. This was contiguous with the wall but signal characteristics were separate from the posterior pharyngeal wall suggesting this was a pedunculated mass with attachment in the nasopharyngeal region (Figure 2). Ear, nose, and throat specialist was consulted to evaluate the mass, and recommended immediate resection of the mass. She was taken to the operating room, placed in the supine position with a pelvic tilt, and orotracheally intubated. Continuous external fetal monitoring (CEFM)

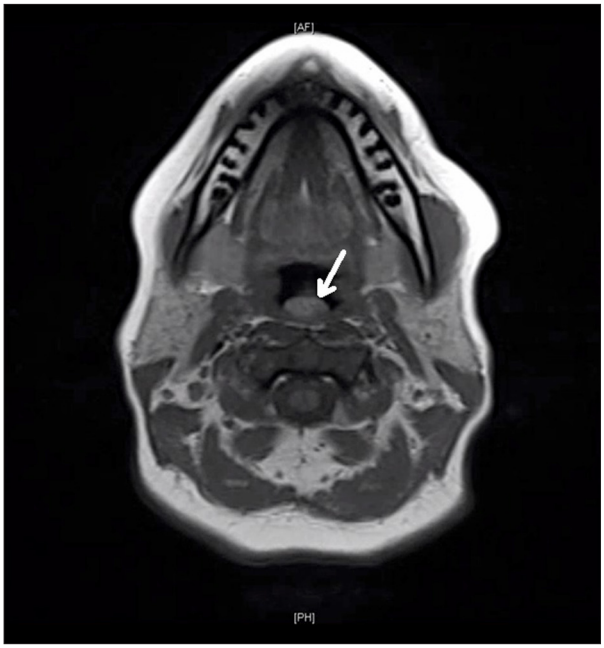

Figure 1. MRI neck axial view showing round mass anterior representing the lymphangiomatous polyp in the nasopharynx (white arrow).

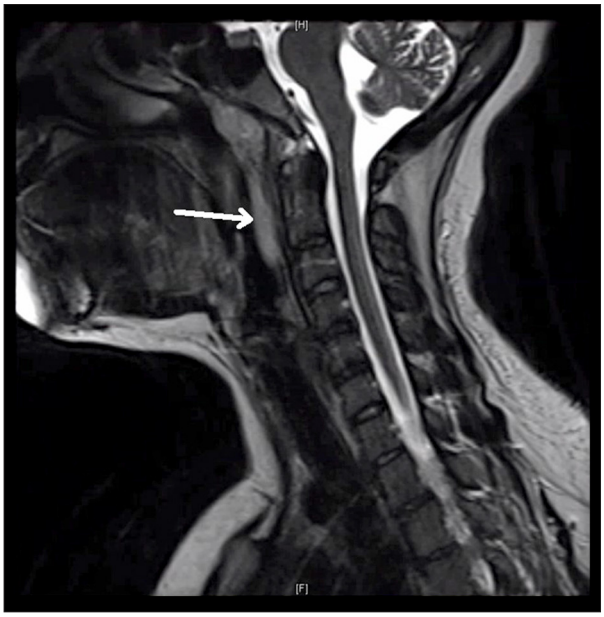

Figure 2. MRI neck sagittal view showing elongated mass extending down the pharynx (white arrow). This mass was the lymphangioma attached to a stalk.

was attached. The mass was localized in the nasopharynx and detached from a broad 1 cm stalk through both the oral and nasal approaches using bipolar cautery. No blood loss was reported and the extracted 4.5×1.5×0.8 cm polypoid mass was sent for analysis. There was no need for packing of the surgical area. Histopathology results showed a polyp composed of squamous

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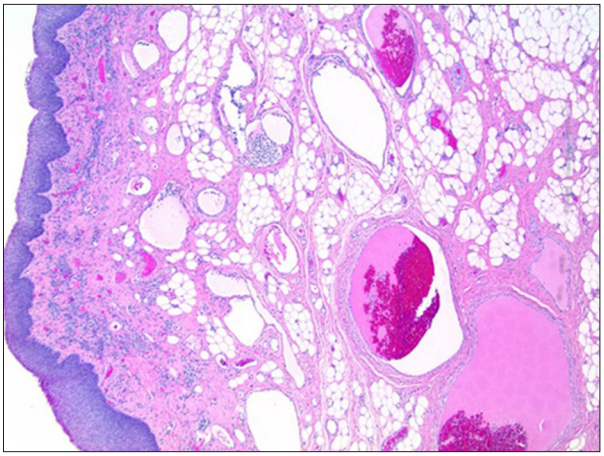

Figure 3. Mass composed of squamous surface epithelium and fibrous stroma with intermixed adipose tissue and open vascular channels.

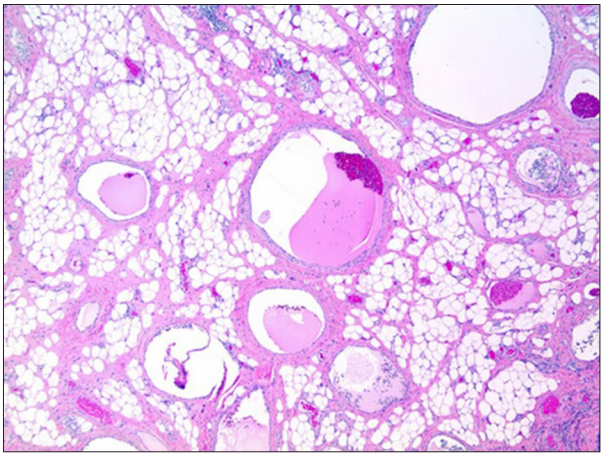

Figure 4. Dilated lymphatic channels filled with lymph fluid and lymphocytes.

epithelium overlying dilated lymphatic channels surrounded by a stroma of fibroadipose tissue (Figure 3). The lymphatic channels were thin-walled, lined by lymphatic endothelium, and contain proteinaceous fluid and mature lymphocytes (Figure 4). The patient was discharged the same day and continued her prenatal course uneventfully. She presented at 41 weeks gestation in early labor and underwent Cesarean delivery for arrest of dilation and nonreassuring fetal surveillance. Her male infant weighed 3427 grams and had a cord pH of 7.13. She was seen 2 years later for an unrelated viral syndrome and there was no physical evidence of regrowth of the lymphangiomatous polyp. She had no complaints of nasopharyngeal obstruction.

Discussion

This case was unique for 3 reasons. First, LMs are very unusual outside of childhood. In fact, 50% are present at birth and 90% are diagnosed within the first 2 years of life [2,3]. They probably represent a congenital malformation, which suggests that even if they were not diagnosed as a child, they would be present and their discovery as an adult would be either coincidental or they changed in size or character possibly resulting in noticeable symptoms or a change in appearance. This is likely, considering the physical character of simple LMs. They are generally painless, soft, spongy, compressible masses with indistinct edges that represent areas of regional dilatation of lymph channels regardless of their location [4]. Nonetheless, they have a significant predilection for the head, neck, and axillary region, most often presenting as asymptomatic posterior triangle masses. Only 5% of LMs are found in the abdomen or mediastinal region [5] and reports of nasopharyngeal LM in adults are rare [6]. Thus, the second unique element in this case was the nasopharyngeal location of the LM. Our patient developed progressive complaints suggesting rapid growth, and when she noticed difficulty breathing she presented for care. The imaging and examination did not reflect findings of a malignancy and there was no evidence of a connection to the meninges, consequently a surgical removal was offered. The procedure necessarily required the patient be placed in a favorable position for surgical access but she was 32 weeks pregnant and extensive uninterrupted time in the supine position can pose a transient fetal threat even with a pelvic tilt. This dilemma emphasizes the third unique component of this case. We performed a MEDLINE search of the English language literature from January 1, 1966, to December 31, 2018, using the keywords “Lymphangioma, Lymphatic malformations, Pregnant, Pregnancy, Pharynx/pharyngeal, Oropharynx/oropharyngeal, and Tonsil”. We could find, however, no cases of pharyngeal lymphangioma reported in a pregnant woman. Several immediate concerns were evident with this combination that reflected the mass effect of the LM. First was the nutritional demands of pregnancy with a pharyngeal mass that could influence swallowing. Second was the regurgitation and aspiration risk with a large dangling intraoral mass. Third was the potential respiratory effect of a large potentially obstructing nasopharyngeal growth. Finally, if an obstetric emergency occurred during the third trimester and general endotracheal anesthesia was needed, the large mass may be an impediment in obtaining the necessary anesthesia for delivery. Infection and hemorrhage are considered potential complications, as well, but are not unique to pregnancy [6]. After considering the risks versus benefits, the patient requested the mass be surgically removed under concomitant CEFM. CEFM can detect both evolving and abrupt fetal hypoxemia and maternal position change and intervention can be implemented quickly. In all likelihood this LM grew rapidly during her gestation. In fact, it has been reported that growth in size and vascularity during pregnancy may be due to excess production of cytokines and vascular endothelial growth factor [7]. Whatever the reason, when the nasopharynx is involved during pregnancy,

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the potential consequences and the prospect of continued rapid growth of the LM should be considered while developing a treatment strategy. Since these lesions do not usually spontaneously regress and are progressive, treatment is mandatory. In the pregnant woman with potential airway obstruction treatment is urgent. Consequently, our patient was admitted for surgery, which is the main treatment option for most LMs. Sclerosing agents and radiation are probably ineffective [8,9] but laser debulking has been reported to be a reasonable choice, however, it has not been used in the pharynx in the third trimester [8].

Conclusions

In summary, this case describes a rapidly growing nasopharyngeal LM that was surgically removed at 32 weeks' gestation when the patient presented with symptoms. The examination and imaging were consistent with a benign nasopharyngeal mass and surgical removal confirmed the diagnosis of a LM. Surgical intervention in this particular pregnant case was safe with co-management among the anesthesia, obstetrics, and throat teams.

Conflict of interest

None.

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