Original Research Article

LYMPHANGIOMA OF NASOPHARYNX : A RARE CASE WITH AN UNUSUAL PRESENTATION.

ABSTRACT: Lymphangiomas are rare benign congenital tumors of the lymphatics, most often encountered during childhood. They are generally found in the head and neck region, isolated involvement of the nasopharynx is very rare and only two cases have been reported since 1969. Lymphangioma of the nasopharynx in an adult presenting with chronic suppurative otitis media as in our patient has not been reported in English literature.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 6 months the patient is free of recurrence.

KEYWORDS: Lymphangioma; Nasopharynx;IHC-immunohistochemistry.

INTRODUCTION: Lymphangiomas are congenital malformations of the lymphatic system. They commonly present in the head and neck region, with over 90% of lesions manifesting before 2 years of age. The onset of the lesion after 2 years of age is rare.

Lymphangioms are classified as microcystic (capillary hemangiomas), macrocystic (cavernous hemangiomas), and cystic hygromas according to the size of the lymphatic cavities incorporated.[1]

Isolated lymphangioma of nasopharynx is very rare,only 2 cases have been reported in last 40 years,in the year 2013.[2]

Nasopharyngeal lymphangioma causing eustachian tube obstruction and precipitating chronic otitis media has not been reported till date as per our literature search.

CASE REPORT:

A 34 year old male presented with left chronic suppurative otitis media since 10 to 12 years; patient had noticed a small mass in the oral cavity on the left side since last 4 to 5 years.

Left tympanic membrane showed large central perforations with mucoid discharge in the middle ear. Patient gave history of intermittent mucopurulent ear discharge which used to subside on antibiotic medications temporarily.

Oral examination revealed a 3x3 cm pinkish cystic mass in the left supratonsillar fossa. On probing it was found to be attached to the posterior tonsillar pillar and to have an extension up into the nasopharynx.(figure1&2)

On zero degree nasal endoscopy a polyp-like mass was visualized , obliterating the eustachian tube(ET) opening on the left side. It was attached to the supero-lateral wall of the nasopharynx,the posterior lip of the ET opening(figure3) and in continuity with the small cystic mass seen in the oropharynx, through the posterior pillar of the tonsil.

A computed tomography(CT) contrast neck reported a minimally enhancing(27 HU on plain and 31 HU on postcontrast) soft tissue mass measuring 22 x 21 mms.There was minimal obliteration of the left fossa of Rosenmuller and the mass extended into the oropharynx involving the left posterior tonsillar pillar and supratonsillar fossa. (figure4 A&B)

A magnetic resonance imaging (MRI) showed no neuro-vascular extension and a submucosal soft tissue mass similar to a polyp.(figure4 C)

After meticulous blood investigations, pre-anaesthetic checkup and informed written consent, patient was taken up for surgery. A combined approach ie. transnasal and transoral was used. A simple rubber catheter was inserted through the right nostril and used to retract the soft palate for better visualization. A 4mm zero degree endoscope along with a insulated sickle knife on which unipolar cautery was applied was used to gradually separate the nasopharyngeal part. Care was taken to avoid cauterization of the eustachian tube opening. Once the nasopharyngeal part was separated from the lateral wall , the oropharyngeal part was excised in a manner similar to tonsillectomy giving the patient Rose's position. The oropharyngeal extension was removed along with the left posterior pillar of the tonsil(figure5) using bipolar cautery.Hemostasis was achieved,rest of the post-operative period was insignificant. Histopathological examination of the specimen revealed multiple cystically dilated channels lined by a flattened endothelium . Immunohistochemically(IHC),it was positive for lymphatic lineage markers D 2–40(figure6).The histopathological diagnosis was lymphangioma.

DISCUSSION Lymphangiomas are uncommon congenital benign malformations of the lymphatic system. They are slowly progressive vascular hamartomas.
Recently Catalfamo et al.[3] reported nine tongue lymphangiomas in adults.
Naidu and Mc Calla[4] 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.
The 34 year old case reported above is probably the first case of nasopharyngeal lymphangioma causing Chronic otitis media(COM).

Lymphangiomas are diagnosed on the basis of history, examination findings and radiological investigations.
Lymphangiomas at oral cavity,tongue,and larynx can present with symptoms of dysphagia, dyspnea, foreign body sensation, sore throat, tonsillitis, and tonsillar mass. When located at the nasopharynx they may cause nasal obstruction but our case had no nasal complaints and presented with left ear discharge.

Though our patient had a visible mass in the oropharynx,smaller sized nasopharyngeal pathology can go undetected if nasal endoscopy is not performed.Hence from our experience, it is essential that every patient with COM be screened with nasal endoscopy to rule out any nasal/nasopharyngeal pathology.

MRI can demarcate the mass from the surrounding tissues and can give the extent of the lesion. Contrast enhanced CT scan can also be helpful in investigating these lesions.

Antrochoanal polyps, nasopharyngeal carcinoma, angiofibroma, cystic lesions, and other benign masses should be included in the differentials.

Lymphangioma can get infected or can cause hemorrhage and the mass effect associated with rapid growth of the lesion.Also in some patients,lymphocytopenia,a precipitating cause of infection,can be documented.[5]

Lymphangiomas can be managed by namely surgical excision; laser debulking, sclerotherapy and corticosteroids. [6] It is suggested that the desire for complete excision must be weighed against the need to keep vital structures intact.
If the larynx is involved tracheotomy is almost always required [7].

Though in our case we used cautery, surgical laser is considered particularly suitable because it allows precise ablation and recontouring of the tissue so that the functions can be preserved [8]. Coablation too would be similar to laser or perhaps even better,but there is no literature available currently regarding its use for such a case.

Sclerosing agents are under investigation but since they act by causing inflammatory reaction, their use can result in damage to adjacent structures. Radiotherapy is not effective in treatment of lymphangiomas[9].

In our case we were able to excise the lesion without causing any damage to surrounding vital anatomical structures using an transoral and transnasal endoscopic approach.

We reported this case because of the unusual site of involvement; in isolation from rest of the head and neck and unusual age of presentation.Also it is probably the first case of nasopharyngeal lymphangioma presenting as active chronic otitis media due to Eustachian tube obstruction.

CONCLUSION:Although lymphangioma of the nasopharynx is very rare, it can cause ET obstruction and present as active COM. Complete surgical excision followed by management of the otological pathology would be the ideal way to go about.Since there is a risk of recurrence there should be sufficient gap between the surgeries for lymphangioma and the affected ear,during which a regular follow up would be essential.

Informed consent was obtained from the patient and his relative before the procedure.No funds/grants were obtained for this surgery.

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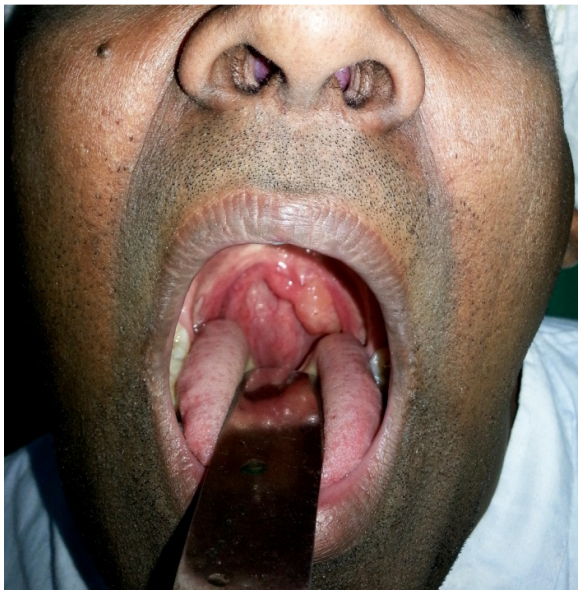


Figure 1:Cystic mass in the oropharynx

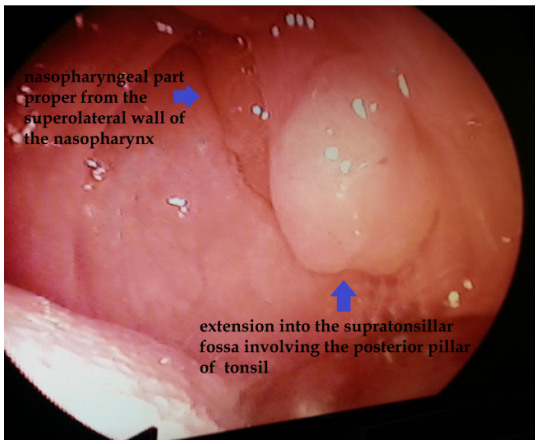


Figure 2:Zero degree view of the mass in the oropharynx

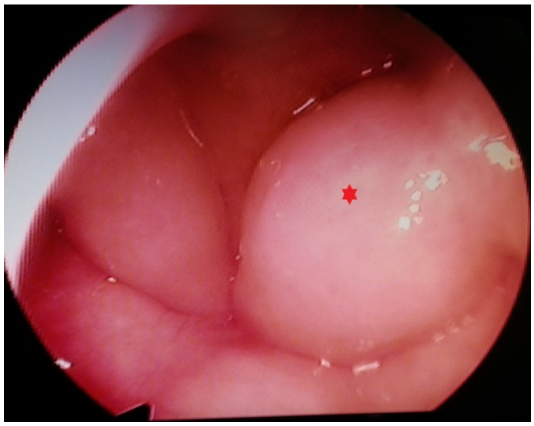
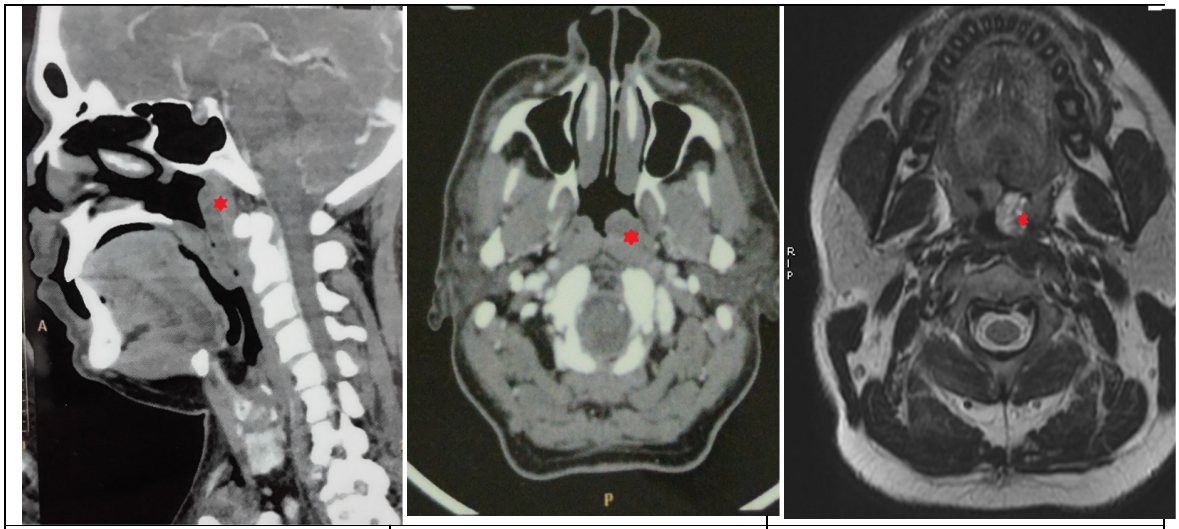


Figure 3:Zero degree view through the left nostril showing the nasopharyngeal mass (*)



a	b	c

Figure 4:Radiological imaging

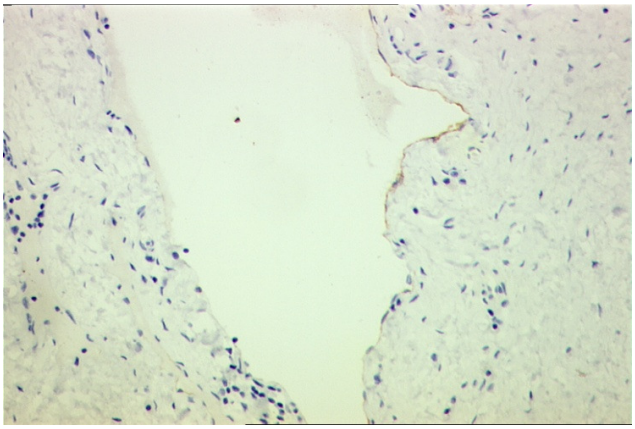


Figure 5:Excisional biopsy specimen

Figure 6:IHC with D2-40 marker for lymphangioma