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Case Report

A rare case of retropharyngeal fibromatosis presenting with stridor

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ABSTRACT

Stridor is one of the common emergencies in otolaryngology clinics, the usual causes being malignancies of glottis and hypopharynx. We present a rare case of stridor caused by retropharyngeal fibromatosis in an elderly patient. A 62-year-old male patient presented with complaints of breathing difficulty associated with dysphagia for one-month duration. On evaluation, he was found to have moderate stridor and a smooth mucosa-covered bulge in the posterior pharyngeal wall obscuring the endolarynx. With a working diagnosis of retropharyngeal mass causing stridor, the patient underwent an emergency tracheostomy. Initial punch biopsies from the lesion were non-diagnostic. On further evaluation by imaging and histopathological examination of incisional biopsy, we arrived at a diagnosis of giant retropharyngeal fibromatosis extending from skull base to C6. The patient underwent en-bloc trans-cervical excision of the tumor. Fibromatoses of the retropharynx is a group of rare tumor entity, which is benign, locally aggressive and recurrent. Dysphagia is the usual presentation though large tumors can present with stridor due to upper airway obstruction. Imaging and preoperative surgical planning play a crucial role in individualizing the management of these tumors due to their inaccessible location and proximity to major vasculature and nerves in the head and neck. Complete, meticulous pre-operative evaluation with imaging followed by surgical management is of paramount importance in the treatment of retropharyngeal tumors and thereby prevent its recurrence.

Keywords: Dysphagia, Airway Obstruction, Biopsy, Benign tumor, Skull base, Tracheostomy

INTRODUCTION

Stridor is one of the most common emergencies seen in otolaryngology clinics. It is high-pitched, noisy breathing occurring due to turbulent airflow through narrowed airway passages. The common causes of stridor in adults include laryngeal malignancies, post-intubation laryngotracheal stenosis, and deep neck space infections. Benign tumors of the pharynx presenting with stridor in adults is extremely rare.

A pharyngeal obstruction causes a bidirectional, low pitched and fricative sound whereas a typical laryngeal stridor is inspiratory, high pitched sound.[1,2] We report a rare case of stridor caused by benign retropharyngeal fibromatosis in an elderly patient.

CASE REPORT

A 62-year-old male presented to the Otorhinolaryngology department with complaints of difficulty in swallowing for one and half months, which was insidious and progressive in nature. The patient also had change in voice and noisy breathing for fifteen days. There was no history of neck swelling, fever, loss of weight or appetite. He had no other comorbidities. He had a pulse rate of 88/min, blood pressure of 130/80 mm Hg and a saturation of 98% with moderate stridor.

Examination revealed a smooth mucosa covered bulge in the posterior pharyngeal wall with its superior and inferior extent not being visualized. Video laryngoscopic

examination revealed the same finding. There were no palpable nodes in the neck.

With a working diagnosis of retropharyngeal mass for evaluation, he was taken up for emergency tracheostomy. A trans-oral biopsy was taken under local anesthesia and on histopathology revealed fibro-muscular tissue with dense inflammatory infiltrate and bacterial colonies suggestive of actinomyces. The patient was started on injection crystalline penicillin and doxycycline with no improvement for 10 days. Trans-oral biopsy was taken under general anesthesia, which was reported as features consistent with fibromatosis. A T2-weighted magnetic resonance imaging (MRI) neck revealed a large hyperintense lesion of 10×7×4 cm involving retropharyngeal and prevertebral space extending from skull base superiorly up to lower border of C6 vertebra with no bony erosion (Figure 1).

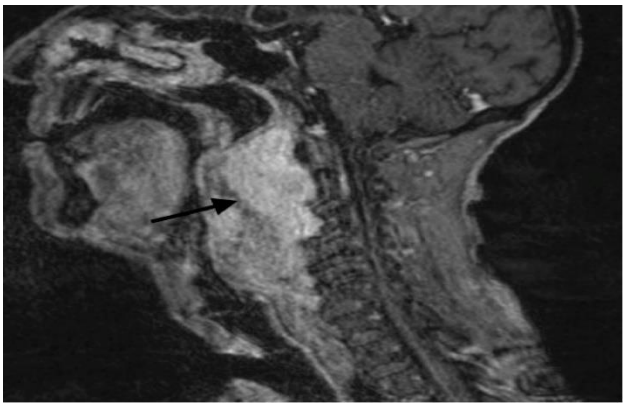

Figure 1: T2-weighted MRI neck showing a large hyperintense lesion (arrow) involving the retropharyngeal and prevertebral space extending from the skull base to the lower border of C6 vertebra.

The patient underwent trans-cervical excision of the tumor under general anesthesia. Intra-operatively, a firm, lobulated mass was seen involving prevertebral and retropharyngeal space displacing carotids and internal jugular vein laterally on the left side. Blunt dissection was carried out all around the tumor, and sharp dissection was done posteriorly, where the tumor mass was involving the prevertebral muscle and ligament and the tumor was removed in toto (Figures 2 and 3).

A 5×0.5 cm defect noted in the posterior wall of the oropharynx was sutured. Post-operatively, the patient was given intravenous antibiotics and Ryles tube feeds. Neck drain was removed on postoperative day 3. Swallowing therapy was initiated. The patient was decannulated after three weeks. The histopathological examination revealed fascicles of spindle-shaped cells in the fibrous stroma with sparse mitotic activity and without necrosis or myxoid changes. On immunohistochemistry, the lesion was strongly positive for smooth muscle actin (SMA) and was consistent with benign fibromatosis.

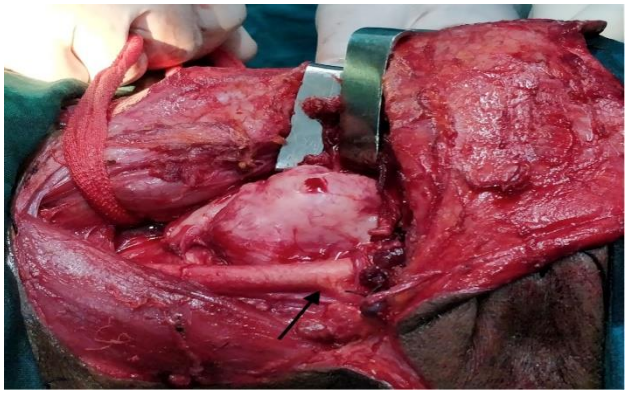

Figure 2: Intra-operative picture showing a mass lesion in the retropharyngeal space displacing the carotid artery (arrow) and internal jugular vein laterally on the left side.

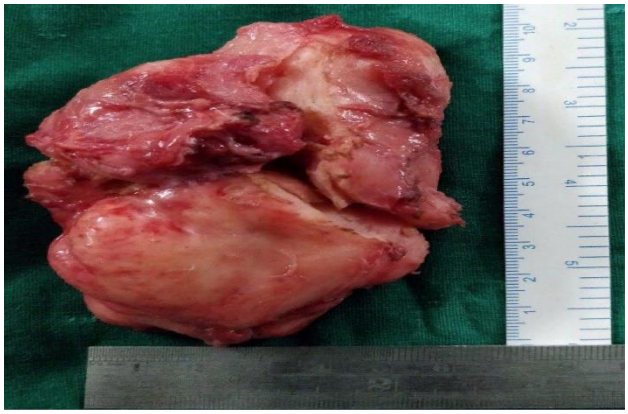

Figure 3: The resected tumor specimen.

DISCUSSION

Primary tumors of the retropharynx are extremely uncommon. The most common primary tumor in the retropharyngeal space is lipoma, as this space is mainly filled with fatty tissues. Other tumors seen in this region are liposarcomas and lymph node masses.[3] Fibromatoses of the retropharyngeal space are even rarer.

Fibromatosis is a rare benign soft tissue neoplasm with locally aggressive behavior. They arise from the fascial or musculoaponeurotic structures and have an extremely high propensity for local invasion. The incidence of fibromatosis is 2.5 to 4.5 per million, out of which 7-25% occur in the head and neck.[4]

Fibromatoses usually remain asymptomatic for a long time and therefore present late with huge masses. Common presenting symptoms include dysphagia, obstructive sleep apnea, dysphonia, globus sensation, and rarely respiratory obstruction. Dysphagia is due to prolonged mechanical obstruction and is more for solids than liquids. Difficulty in phonation occurs due to a loss of resonance due to obstruction of nasopharynx and hypopharynx and restricted tongue movements. Respiratory obstruction is

attributed to the compression of the airway by the growing tumor. In our case, the patient presented with dysphagia, hoarseness of voice, and stridor.

Examination by direct laryngoscopy or endoscopy can reveal a posterior pharyngeal wall bulge with intact mucosa. Large masses may be palpable in the lateral aspect of the neck.[4] Lymph nodal enlargement is not seen in patients with fibromatoses.

Radiological imaging by CT scan and MRI play a crucial role in the diagnosis and treatment. Imaging aids in evaluating the site, size, the extent of the tumor, and its relation to the surrounding vessels and nerves. On a contrast CT scan, a smooth, well-defined soft tissue mass with minimal enhancement is characteristic. In our case, on radiological examination, the tumor was seen displacing the carotids and internal jugular veins laterally.

Fine needle aspiration cytology (FNAC) is usually non-diagnostic. It is usually done to exclude infectious pathology like tuberculosis. It is advisable to perform an FNAC after imaging, as it will give an idea about the characteristics of the lesion. A needle biopsy is of immense value in arriving at the diagnosis. However, the risk of edema formation and worsening of airway obstruction following biopsies should be borne in mind.[5]

Patients with airway obstruction require a tracheostomy before proceeding to definitive treatment, as in our case. Complete surgical excision is the treatment of choice for fibromatoses of the retropharynx. Different surgical approaches like trans-oral with or without soft palate split, trans-cervical, and trans-parotid are reported.[6,7] The trans-oral approach is used for small midline tumors. In our case, a trans-cervical approach was used along the anterior border of the sternocleidomastoid, and the tumor was removed en-bloc. Pharyngeal tears should be identified and repaired immediately. A prophylactic nasogastric feeding tube is placed before surgery. Radiotherapy and chemotherapy are of no role in the treatment of benign retropharyngeal tumors.

Long-term follow-up may be needed as these tumors can recur in case of incomplete removal, and there is also a rare chance of malignant transformation of the tumor into fibrosarcoma.

CONCLUSION

Tumors of the retropharynx are rare, and surgical treatment is challenging due to the proximity to major vessels and nerves. Complete, meticulous pre-operative evaluation with imaging is of paramount importance in the treatment of retropharyngeal tumors. The surgical approach should be individualized depending on the presentation and location of the tumor.

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