

Case Report

Unusual presentation of intranasal myxoma - A case report

Roshni Kuniyil¹, Shruti Ogra¹, Ameen Sumais A. P. Achaparambil¹, Meesala Bhageeratha¹¹Department of Otorhinolaryngology, Northern Railway Central Hospital, Delhi, New Delhi, India.**ABSTRACT**

Myxomas are benign and locally invasive neoplasms of primitive mesenchymal origin. They are most commonly found in the atria of the heart. They have also been reported in bones, muscles, and other connective tissues. Myxomas are rarely found in the nose and paranasal sinuses. Myxomas are notorious for their recurrence after excision and local invasion. We report a case of intranasal myxoma in a 30-year-old male who presented with blocked ear sensation and progressive nasal block. With this report, we intend to increase awareness about the clinical presentation, histologic characteristics, and management options of myxoma.

Keywords: Myxoma, Nose, Paranasal sinuses**INTRODUCTION**

Myxomas are benign and locally invasive tumors of primitive mesenchymal origin^[1] of unknown etiology. They have been most frequently reported in the atria of the heart. They are rare in the head-and-neck region but have been reported in sites such as mandible, maxilla paranasal sinuses, temporal bone, pharynx, larynx, and soft tissues of the face and neck. A high recurrence rate has been reported mainly due to incomplete excision of the tumor. We report a case of intranasal myxoma arising from the maxillary sinus and extending to the nasopharynx. Endoscopic surgery was performed to remove the tumor in toto.

CASE REPORT

A 30-year-old male presented with complaints of bilateral ear blocked sensation for 3 months. He gave a history of progressive nasal obstruction more on the right side than the left side for 2 years. There was no associated rhinitis, headache, epistaxis, puffiness of face, and reduced sense of smell or visual disturbances. He gave the history of nasal surgery (septoplasty) in 2010. His medical history was unremarkable.

On otoscopic examination, his right tympanic membrane showed Grade 1 retraction of pars tensa with normal pars flaccida. His left tympanic membrane was normal. Anterior rhinoscopy revealed a large septal perforation. A diagnostic nasal endoscopy was done under surface anesthesia and proper decongestion. It revealed a non-pulsatile pearly white mass with vascular markings almost completely filling the nasopharynx, blocking both eustachian tube orifices [Figure 1]. Its stalk was traced and found to be emerging from the natural

Ostia of the right maxillary sinus. He was advised radiographic evaluation. His plain computed tomography (CT) of paranasal sinuses showed a lobulated hypodense fluid attenuating lesion of 23 × 22 × 16 mm in the posterior nasopharynx abutting the adenoids posteriorly, right inferior turbinate anteriorly, right fossa of rosenmuller, and right eustachian tube opening. Complete opacification of the right maxillary sinus by mucosal thickening was seen. The bony walls of bilateral paranasal sinuses were normal. Contrast-enhanced magnetic resonance imaging was done and confirmed the CT findings.

Patient was taken up for endoscopic excision of the nasal mass under general anesthesia. Wide right middle metal antrostomy was performed. A Polypoidal stalk was found to be attached to the medial wall. Stalk was dissected from the medial wall using different angled endoscopes and forceps. Both the antral and nasopharyngeal part of the mass was removed and delivered in toto through the nasal cavity. Histopathological examination showed tissues lined by ciliated columnar pseudostratified epithelium with squamous metaplasia. The subepithelium is loose edematous with spindle cells, stellate cells, and interspersed blood vessels consistent with intranasal myxoma. Immunohistochemical staining was performed and was positive for vimentin but negative for s100.

DISCUSSION

Myxomas are benign and locally invasive neoplasms of primitive mesenchymal origin. The term myxoma was first coined by Virchow in 1871.^[2] He named them so due to their resemblance to umbilical cord. The true mesenchymal nature of myxoma was

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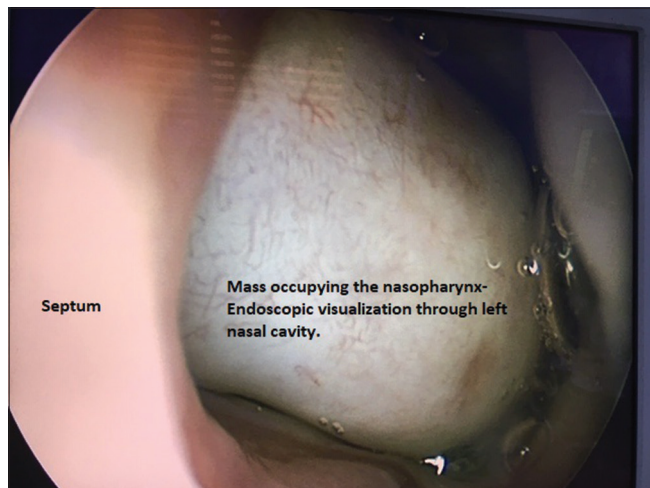


Figure 1: Mass occupying the nasopharynx.

first described by Stout. The incidence of myxomas of the head and neck is 0.1%.^[3] Peak incidence has been described between the ages of 25 and 35 years with slight female predominance.^[4]

Myxomas present as a slow growing painless mass. Patients usually present with obstructive symptoms. Rhinorrhea, headache, and epistaxis are other symptoms. Due to their locally invasive nature myxomas can erode the orbital floor and dural plates resulting in proptosis, infraorbital paresthesia, CSF leak, and cranial nerve palsies.

Myxomas appear as well circumscribed greyish mass with a false capsule. Microscopically myxoma is composed of stellate-shaped cells containing small bland spindle shaped nuclei and elongated cytoplasmic tails. They have a matrix rich in acid mucopolysaccharides, composing 80% of hyaluronic acid and 20% of chondroitin sulfate.^[3]

The differential diagnosis for myxomas include rhabdomyosarcoma (embryonal or botryoid), neurofibroma, rhabdomyoma, fibro myoma, fibrous histiocytoma, myxoid chondrosarcoma, mucoid liposarcoma, and neurogenic sarcoma. Moshiri *et al.*^[5] demonstrated the characteristic light microscopy finding of the presence of spindle-shaped cells surrounded by amorphous material and that the tumor cells were positive for vimentin, a pan mesenchymal marker, and also for actin.

Myxomas can be odontogenic (associated with teeth) or non-odontogenic/osteogenic (arising in bones away from tooth bearing areas) in origin. Thoma and Goldman^[6] observed that the osteogenic type of myxomas are more aggressive than those of the odontogenic type. Myxomas of the head and neck are extremely rare tumors. Myxomas of bone primarily arise from facial bones.^[7] Perzin *et al.*^[8] reported that out of 256 non-epithelial tumors involving the nasal cavity, paranasal sinuses, and nasopharynx, only six were myxomas.

Complete extensive surgical resection with wide margins is the treatment of choice due to the high potential of

recurrence. It has a poor response to chemotherapy and radiotherapy.^[9] No metastatic potential has been reported. In our case, we had done a complete endoscopic resection with negative margins. Patient has been followed up for 1-year postoperatively with nasal endoscopic examination on every visit. No signs of recurrence have been noted.

CONCLUSION

Myxomas are benign and locally invasive neoplasms with varied presentations. As they are rare in head and neck region, thorough patient evaluation is required for differentiating a myxoma from other paranasal sinus tumors. Complete surgical resection with wide margins will prevent their recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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