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Case Report

Unveiling the rarity: a laryngeal myxoma arising from the epiglottis

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ABSTRACT

Myxomas are benign mesenchymal neoplasms which are rare in the head and neck region. Here, we present a case of a myxoma arising from the epiglottis, an exceedingly rare location in the head and neck. Our patient presented with a solid lesion arising from the lingual surface of the epiglottis. Histopathological examination and immunohistochemistry confirmed the diagnosis of myxoma. A myxoma arising from the epiglottis has not been reported in the literature during the last three decades. Myxomas should be considered in the differential diagnosis of benign-looking lesions of the larynx. The slow-growing nature of the tumor often delays symptom onset and diagnosis. Although recurrence is rarely reported in laryngeal myxomas, close follow-up is recommended.

Keywords: Epiglottis, Myxoma, Supraglottis

INTRODUCTION

Myxomas are slow-growing benign mesenchymal neoplasms that can occur anywhere in the body, but most commonly arise from the heart. The term "myxoma" was first described by Virchow in 1871 to denote a tumor resembling the mucinous part of the umbilical cord.[1] They are true mesenchymal neoplasms consisting of undifferentiated stellate cells in a loose myxoid stroma and have propensity for local infiltration and recurrence.[2]

Myxomas in the head and neck region are rare, with the most common locations being the maxilla and mandible, accounting for 3-6% of all myxomas.[3] Laryngeal localization of myxomas is exceedingly rare, with most reported cases involving the vocal folds.[1] Chen et al reported a case of myxoma arising from the lingual surface of the epiglottis in the American journal of otorhinolaryngology in 1986.[1] To our knowledge, no cases of myxoma arising from the epiglottis have been reported in the last three decades.

Here, we present a case of myxoma arising from the lingual surface of the suprahyoid epiglottis in a 37-year-old male and provide a review of the literature on this subject.

CASE REPORT

A 37-year-old male presented to the Otorhinolaryngology outpatient clinic at our tertiary care teaching institution, with a one-year history of progressive dysphagia and a change in voice. He had no significant past medical or surgical history. Video-laryngoscopic examination revealed a large mass over the base of the tongue, obscuring the view of the valleculae and epiglottis (Figure 1). Plain and contrast-enhanced computed tomography of the neck, with axial and coronal sections, identified a well-defined fluid-density lesion on the lingual surface of the suprahyoid epiglottis measuring 3×3×1.7 cm (Figure 2). The lesion also bulged into the valleculae, with no significant post-contrast enhancement.

With a provisional diagnosis of a vallecular cyst, we planned excision of the lesion. However, as a precautionary measure, an elective tracheostomy was performed to safeguard the airway during surgery. We

proceeded with complete excision of the lesion using coblator under general anesthesia (Figure 3).

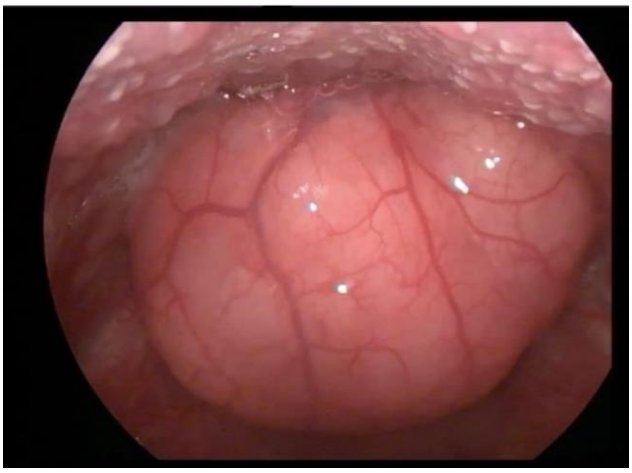

Figure 1: Video-laryngoscopic photograph showing a large mass below the base of the tongue.

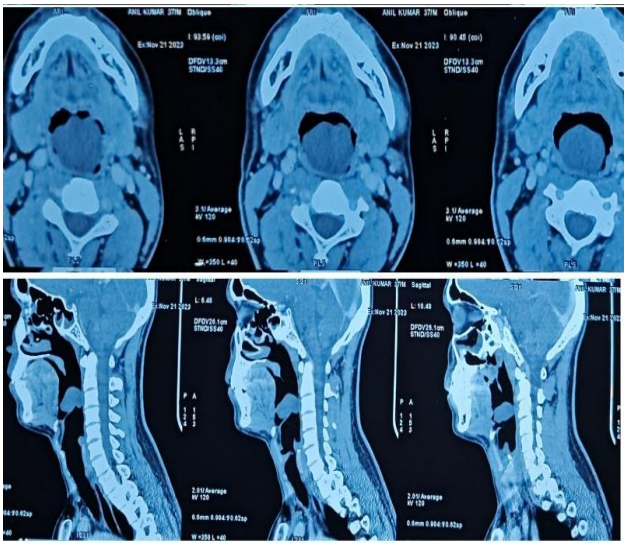

Figure 2: CT image of the neck showing a well-defined fluid-density lesion on the lingual surface of the suprahyoid epiglottis.

The gross specimen appeared well-circumscribed, smooth, and greyish-white in appearance. After performing a videolaryngoscopic examination to assess for post-operative laryngeal edema, we decannulated the patient one week later. Histopathological examination revealed a spindle cell lesion with myxoid stroma. The spindle and stellate fibroblasts were arranged randomly within a basophilic myxoid matrix. Immunohistochemistry showed tumor cells positive for CD34 and negative for S100 and SMA, with no histological signs of malignancy found. These findings were consistent with a diagnosis of myxoma.

Our patient is currently under regular follow-up and has shown no evidence of recurrence to date.

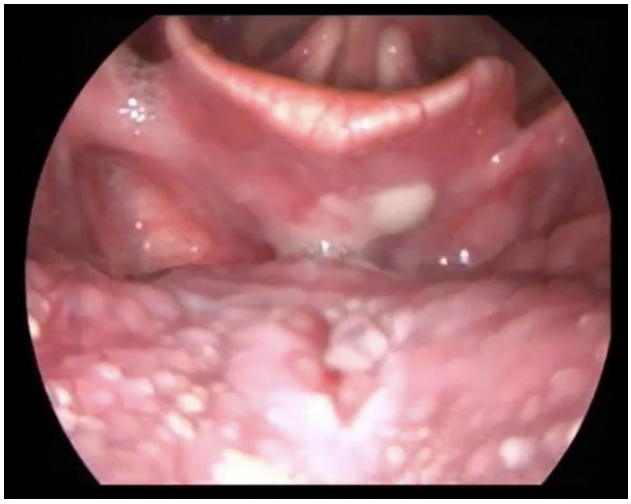

Figure 3: Video-laryngoscopic image following complete excision of the lesion.

DISCUSSION

Myxomas are slow-growing, myxoid benign neoplasms of mesenchymal origin.[2] They may arise from any anatomical site in the body, but they are classically seen arising from the heart.[4] They are the most common primary cardiac tumor, with the left atrium being the common site. Myxomas of the head and neck are rare and commonly arise from the maxilla and mandible.[5] Myxomas of the maxilla or mandible often contain odontogenic epithelium and are believed to arise from odontogenic primordial mesenchyme.[3]

Myxomas of the larynx are exceedingly rare. Among them, tumours are commonly found in the glottis (79.17%), and hoarseness is the common symptom.[5] Most of them are small lesions resembling vocal fold polyps.[1] The small space in the glottis restricts tumour growth and hence glottic myxomas are detected early and are usually small. In contrast to glottic tumours, the tumour in our patient was huge and arose from the lingual surface of the suprahyoid epiglottis, presenting with dysphonia and dysphagia. Due to the spacious areas in the supraglottis and the slow-growing nature of the tumour, supraglottic tumours are detected late and tend to be larger in size. Initial clinical presentations can include foreign body sensation in the throat, dysphagia, dysphonia, and snoring.[5]

The etiopathogenesis of laryngeal myxoma is unknown. Immature fibroblasts are not able to polymerize collagen and produce a large amount of glycosaminoglycans, giving myxoma a gelatinous appearance on macroscopic examination.[2] Histopathologically, myxomas show loosely arranged stellate, spindle, and round cells in abundant loose myxoid stroma.[1] Immunohistochemistry, with S-100 protein indicating lipoblasts and chondroblasts (which was negative in our case) and CD34 commonly used as a marker for myxomas (which was positive here), aids in diagnosis.[5] Histopathological

differentials for myxoma include myxoid liposarcoma, myxoid leiomyosarcoma, rhabdomyosarcoma, chondrosarcoma, and other soft tissue tumours with myxoid degeneration.[1,5]

Although myxomas are well-circumscribed, they lack a fibrous capsule, leading to infiltration of surrounding tissue and a known risk of recurrence if not excised with adequate margins.[2] Recurrence is not widely reported in laryngeal myxomas compared to extra-laryngeal myxomas. Removing the tumour with a margin of normal tissue minimizes the risk of recurrence.[2] Frozen section analysis should be used in cases with questionable margins to ensure adequate resection. Malignant transformation has not been reported in the literature.

CONCLUSION

Myxomas of the supraglottis, especially the epiglottis, are extremely rare. Their slow-growing nature added with the spacious anatomical areas in the supraglottis contribute to delayed diagnosis. Although benign, they have a propensity for local recurrence, which underscores the importance of performing adequate excision.

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