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Unusual laryngeal mass: when it's not malignancy – a case of supraglottic Schwannoma

Sudeep KC[1], Manisha Paneru[2], Deepak Yadav[3], Manisha Shrestha[4]

[1]Lecturer, Dept. of Radiology & Imaging, [2]Lecturer, [3]Assoc. Prof., Dept. of ENT & HN Surgery, [4]Asst. Prof., Dept. of Pathology, Patan Hospital, Patan Academy of Health Sciences, Lalitpur, Nepal

Abstract

Neurogenic tumors of the larynx are very rare, with laryngeal schwannomas making up about 0.1-0.3% of cases. Laryngeal schwannoma is a slow-growing, benign encapsulated, neurogenic tumor typically found in the supraglottic larynx, rarely in the glottis. This report discusses a 44-year-old female with a hoarseness of voice and dyspnea on exertion. Imaging findings were suggestive of benign lesion. Patient underwent Microlaryngeal Surgery (MLS) excision and was confirmed histologically as a Schwannoma. This report highlights this rare condition and reviews diagnostic methods, differential diagnoses, and treatment options.

Keywords: Antoni A and B; Peripheral Nerve Sheath Tumour; MRI; Schwannomas

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Correspondence: Dr. Sudeep KC, Dept. of Radiology & Imaging, Patan Hospital, Patan Academy of Health Sciences, Lalitpur, Nepal. **Email:** sudeepkc@pahs.edu.np

Introduction

Neurogenic tumors in the larynx are uncommon, making up less than 1.5% of benign tumors.[1] These tumors typically arise from the aryepiglottic fold (80%) or false vocal cords and extend into the supraglottic area. Rare in the larynx, schwannomas often involve the internal branch of the superior laryngeal nerve, although this origin is often not visible during surgery due to the small nerve fibers in the laryngeal submucosa.[2]

They can occur at any age, typically in the 4th-5th decades, and show a female predominance (3:1). Patients typically experience hoarseness of voice, globus sensation, odynophagia, dyspnea during exertion, often with inspiratory or biphasic stridor. Imaging studies cannot provide a definitive diagnosis but can rule out other differential diagnosis.[3] Histopathological examination is the mainstay for diagnosis, and immunohistochemistry is required for differentiating it from neurofibroma. Complete surgical resection is the only curative treatment.[2] We are presenting a case of laryngeal schwannoma due to its rare prevalence as well as the slow growth of the lesion without symptoms.

Case Report

A 44-year-old woman presented to the Department of Otorhinolaryngology with a 2 3-year history of hoarseness of voice and dyspnea on exertion. She had a 15-pack-year smoking history. Further medical history was unremarkable, and physical examination was normal. Fiberoptic laryngoscopy revealed a submucosal mass within the right supraglottic larynx obstructing the view of the true vocal cord, Figure 1c,d. The mobility of the right vocal cord was normal. In view of supraglottic mass, patient was advised to undergo Contrast-enhanced Computed Tomography (CECT), which showed a well-defined homogeneously enhancing soft tissue density lesion measuring 12.5x17.2x19.3 mm (APxTRxCC) in right supraglottic larynx, Figure 1a,b. No internal calcifications, cystic or hemorrhagic components were noted. No definite erosion or sclerosis was noted in the surrounding bones and cartilage. Imaging findings were suggestive of a benign lesion.

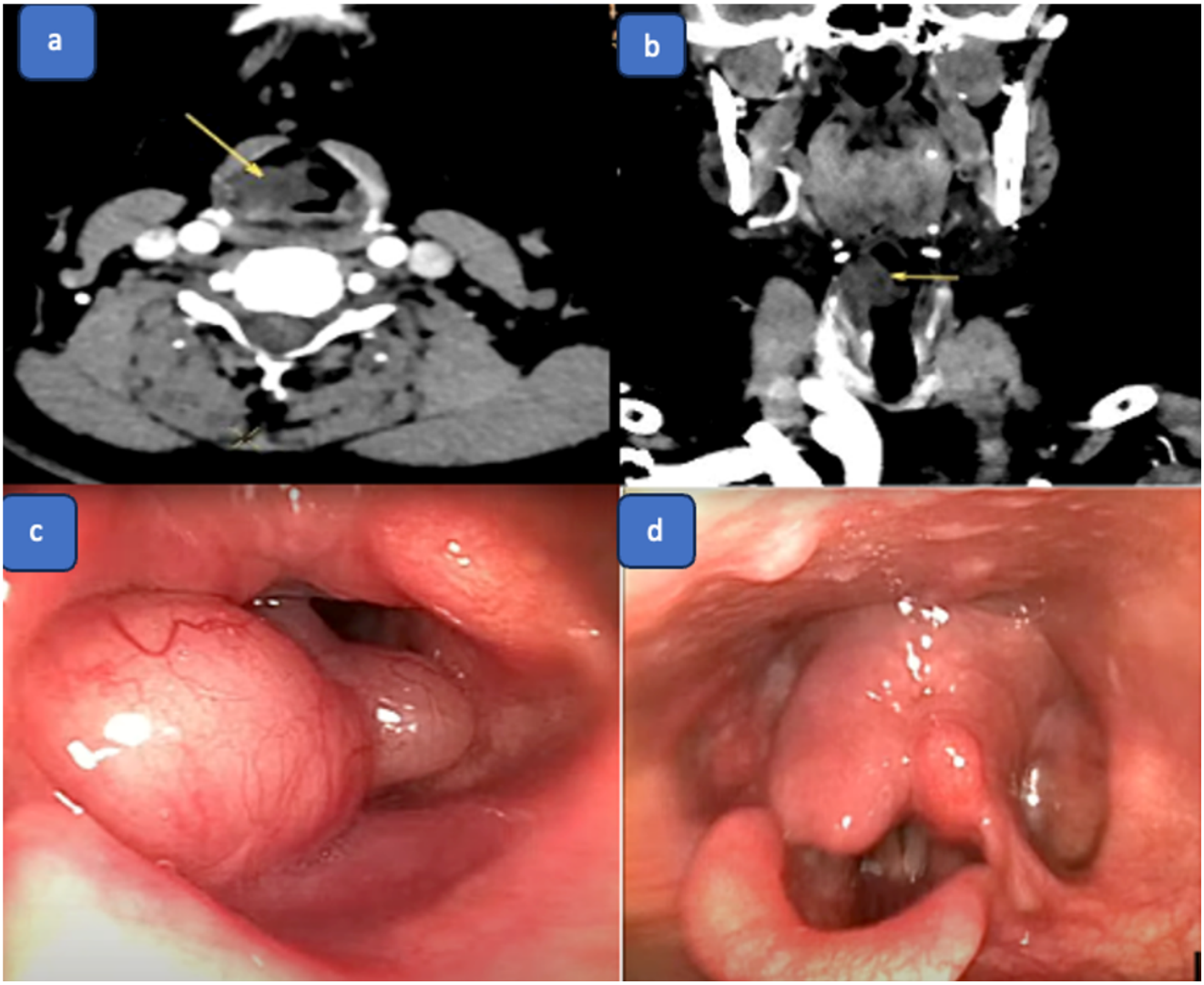

Figure 1: Post contrast CT neck showing well-defined mildy enhancing soft tissue density lesion(arrow) in right supraglottic larynx(a,b). Mass presenting as a submucosal bulge obstructing view of vocal cords(figure c).Post operative changes showing minimal edematous changes only at the post operative supraglottic larynx(d)

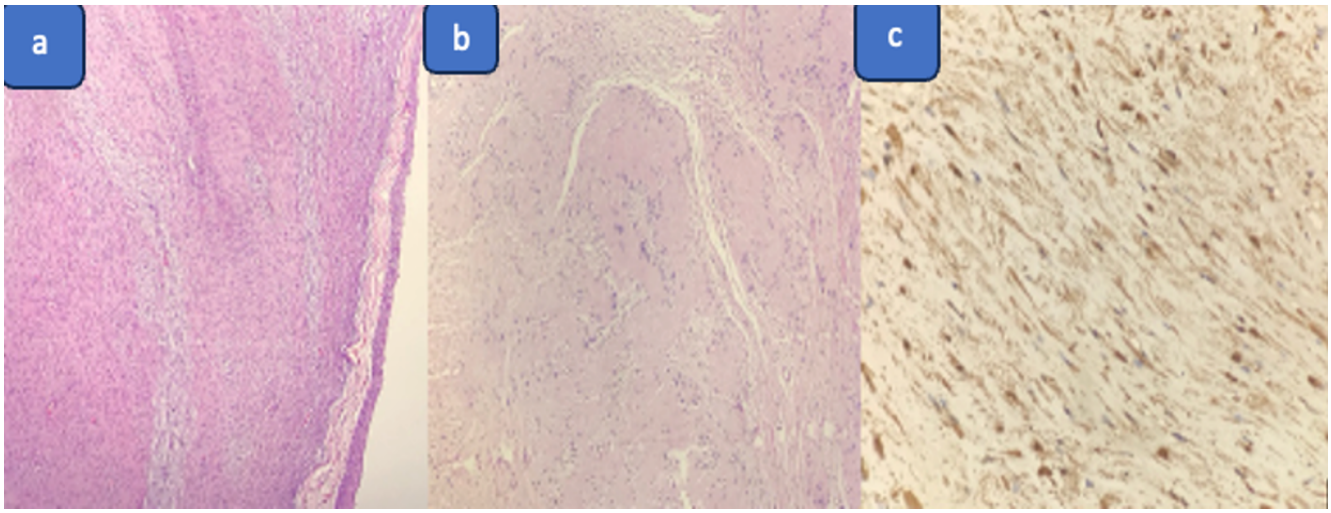

Figure 2: section(a) shows larynx lined by stratified squamous epithelium. The underlying stroma shows schwannoma composed of hypercellular "Antoni A" and hypocellular "Antoni B" areas. (H and E, 10x). Figure b: section shows palisading of tumor cell nuclei forming verocay bodies (H and E, 10x). Figure c: Tumor cells are positive for S-100 (immunohistochemistry, 40x)

She underwent Microlaryngeal Surgery (MLS) Laser Excision. Intra-operative findings showed a well-circumscribed mass occupying the laryngeal surface of epiglottis. Histopathology confirmed the tumour to be a schwannoma. Microscopic examination revealed predominantly cellular Antoni A areas as well as hypo cellular Antoni B areas, Figure 2a,b,c. Hypocellular areas showed hyalinization, edema, sclerosis and focal myxoid change. However, the Verocay bodies, mitosis, and necrosis was not observed. S100 protein staining was positive in our case, confirming the diagnosis of Schwannoma. The postoperative period was uneventful without neurological deficit and the patient was subsequently discharged and was kept under regular follow-up. On follow-up after 1 month, the hoarseness of voice has disappeared. Fiberoptic laryngoscopy showed minimal edematous changes at post operatives, Figure 1d.

Discussion

Benign neurogenic tumors in the larynx, particularly in the aryepiglottic folds, include schwannomas and neurofibromas.[1] Schwannomas arise from perineural Schwann cells, are well-encapsulated, and grow alongside the parent nerve, while neurofibromas originate from perineural fibrocytes, lack encapsulation, and intertwine with the parent nerve.[4]

The disease's symptoms resemble a slowly progressing laryngeal lesion. Some patients may have dyspnea when lying down due to the lesion's position. A case of asphyxiation from a laryngeal schwannoma has been reported.[3] A large supraglottic tumor can obscure the true vocal cord and create a mass effect that mimics cricoarytenoid joint fixation, termed 'pseudo-fixation'. The affected true vocal cord may show immobility or reduced mobility, although some studies indicate normal movement.[5] Fiberoptic laryngoscopy typically reveals a red or blue smooth, lobulated, submucosal mass lesion.

The role of ultrasonography is not well established.[6] CECT shows a well-defined, hypodense lesion without signs of infiltrative growth, showing variable contrast enhancement. These modalities provide valuable insights into the lesion and its relation to surrounding structures, making CECT the preferred initial imaging method.[5,6] Calcification has also been reported in degenerating laryngeal schwannoma. On Magnetic Resonance Imaging (MRI), schwannomas typically present low-isointense signals on T1-weighted images compared to adjacent muscle and hyperintensity on T2-weighted images, often with significant gadolinium enhancement. While MRI can identify schwannomas, it cannot confirm if the mass is fully excisable.[6]

The main goal of imaging in laryngeal mass is to determine its origin and location (mucosal or submucosal). Radiologically, mucosal lesions are often squamous cell carcinomas (SCC) unless proven otherwise.[7] SCCs are more common and have a worse prognosis. Imaging alone cannot definitively distinguish between benign and malignant lesions, so SCC should be included in the differential.[7] Most laryngeal SCCs show clear mucosal abnormalities, which were absent in our case. Lysis or sclerosis of laryngeal cartilage, lymphadenopathy, and multifocal or infiltrative lesions are associated with malignant lesions.[7] Other malignant possibilities include non-Hodgkin's lymphoma, adenoid cystic carcinoma, and sarcoma. Calcification in a mass or laryngeal cartilage involvement may suggest chondrosarcoma or chondroma.

Chondroma, papilloma, laryngeal cyst, paraganglioma, and internal laryngocele are common submucosal differentials. Paragangliomas are typically found in supraglottic or paraglottic areas, showing avid enhancement. A single polypoid mass in a middle-aged patient might indicate a papilloma.[7] In our case, we had kept schwannoma, paraganglioma, and SCC as the differentials.

The role of preoperative biopsy is debated due to the need for airway intervention and deeper biopsies for better diagnostic yield.[6] A definitive diagnosis requires histological examination, with key criteria for Schwannoma including a well-defined capsule, presence of Antoni A and/or B regions, and strong S-100 protein immunoreactivity as seen in our case.[6]

Surgery is the main treatment for laryngeal schwannoma, with tracheostomy sometimes needed for airway security. Smaller tumors may be treated with endoscopic (laser) resection, while larger ones require external surgical approaches like lateral thyrotomy or pharyngotomy.[8] The surgical separation of the tumor from the nerve is theoretically possible in Schwannoma, whilst in neurofibroma it is impossible. Post-surgery, vocal cord mobility is often restored, even if initially immobile.[4]

Recurrence of Schwannoma are rare. Malignant transformation of laryngeal schwannoma is uncommon; there are currently no reports of it in the literature.[5] In the largest published series, no malignant transformation was reported at mid-term follow-up of benign solitary facial nerve Schwannomas.

Conclusion

Laryngeal neurogenic tumors, though rare, require prompt recognition with airway protection as a priority. Imaging, especially CECT/MRI, aids in diagnosis and surgical planning, but definitive differentiation between schwannomas and neurofibromas relies on histopathology. Complete excision is necessary to prevent recurrence.

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Conflict of Interest

None

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Author Contribution

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