

TRACHEAL SCHWANNOMA: A MISLEADING ENTITY

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ABSTRACT

Primary tracheal tumors are rare with the majority being malignant. Benign lesions are less frequent with primary tracheal schwannomas accounting for less than 0.5% of tracheal tumors. They are more common in females and their clinical presentation is non-specific. Chronic cough, progressive respiratory distress and even asthma-like conditions prevail as presenting symptoms and signs. Laryngotracheal endoscopy reveals a solitary, well encapsulated mass arising most often from the posterior tracheal wall. The diagnosis of tracheal schwannomas is primarily pathological. Endoscopic excision, sleeve excision or tracheal resection, are all commonly accepted treatment modalities. Proper awareness of these lesions is crucial in the pre-operative work-up of patients presenting with stridor.

Key words: Trachea, shwannoma, resection.

Introduction

Primary tracheal tumors are rare with the majority being malignant. Benign lesions are less frequently encountered and embrace a wide heterogeneous group comprising adenomas, papillomas, fibromas, lipomas, hemangiomas and neurogenic tumors¹. Though benign as indolent lesions, their clinical presentation might be malignant. The lack of localizing symptoms often misleads the primary physician and results in a delayed diagnosis. Pre-operative work-up and proper mode of anesthesia delivery is important and may avoid a tracheostomy on these patients unless severe obstruction is present. A case report of a rare, primary tracheal schwannoma is described with emphasis on the pathologic findings and modalities of treatment.

Case Report

A 68 year old lady diabetic, hypertensive and cardiac presented six weeks status open heart surgery with history of respiratory failure following multiple attempts of extubation. Her history dates back to the time of surgery when difficult intubation was encountered upon induction of anesthesia. Direct laryngoscopy and bronchoscopy done by an outside physician revealed a tracheal mass at the level of the second tracheal ring obstructing the airway. A tracheotomy was done and the patient was sent to us for further evaluation and treatment. Further questioning

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of the family members revealed history of asthma with noisy breathing, exertional dyspnea and decrease in exercise tolerance for which the patient was treated to no avail. A fiberoptic nasopharyngeal laryngoscopy revealed normal vocal cord mobility with the evidence of a high tracheal mass. Magnetic resonance imaging of the neck revealed a 1-2 cm tracheal lesion arising from the posterior tracheal wall with a high signal intensity on a T2 weighted images (Fig. 1). There was no evidence of cartilage invasion or extratracheal spread. Bronchoscopy was done under general anesthesia and showed a soft smooth well rounded 2x2 cm mass located around 3cm below the true vocal cords. Incisional biopsy showed an expanding, well circumscribed submucosal mesenchymal neoplasm arising from the respiratory mucosa and compressing submucosal glands. High power view of the tumor showed neural spindled cells with bland nuclei that focally showed a palisaded arrangement forming verocay bodies. Immunohistochemical staining for S-100 protein and vimentin were positive (Fig. 2). The diagnosis was consistent with tracheal shwannoma.

Fig. 1

T2 weighted images, coronal cuts taken at level of tracheal narrowing, showing obliteration of the lumen by this high signal mass.

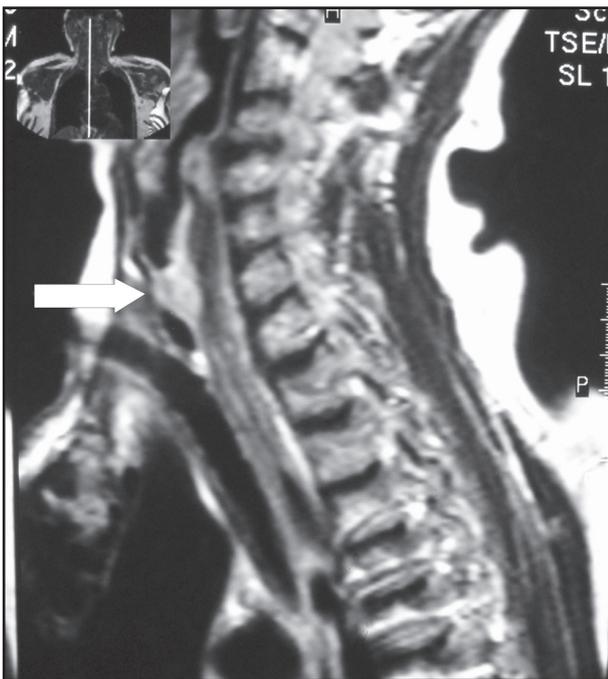
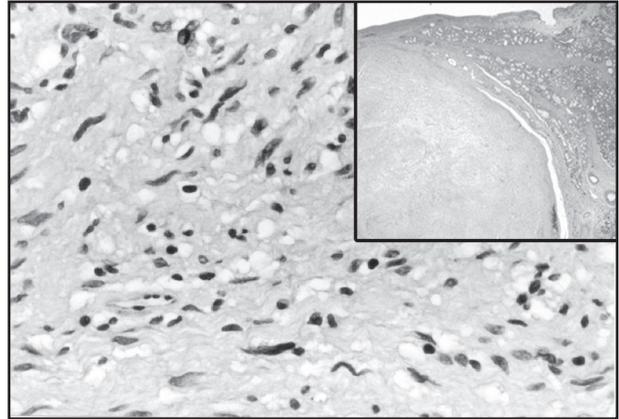


Fig. 2

Well circumscribed submucosal mesenchymal neoplasm

arising from the respiratory mucosa and compressing submucosal glands. High power view of the tumor showed neural spindled cells with bland nuclei that focally showed a palisaded arrangement forming verocay bodies.



Patient underwent surgical excision of the tumor with resection of two tracheal rings and primary end to end anastomosis. Patient was extubated directly post-operatively with no complications. Routine fiberoptic endoscopy at 6 months interval for the first year showed no recurrences.

Discussion

Primary tracheal schwannomas are rare lesions of the trachea, accounting for less than 0.5% of tracheal tumors¹. They are more common in females with no age predilection². Their clinical presentation varies and is usually non-specific. Chronic cough, progressive respiratory distress and even asthma-like conditions prevail as presenting symptoms and signs^{1,2,3}. Poor response to conventional medical treatment prompts the physician to seek further investigation. Unfortunately, an average delay of 10-15 months between onset of symptoms and diagnosis is reported³.

Radiological studies can be non specific. Computerized Tomography scan usually shows a well circumscribed enhancing mass within the tracheal lumen^{1,4} with no specific features. Magnetic Resonance Imaging of a tracheal shwannoma, similar to other peripheral schwannomas, displays isointensity or hyperintensity on T1-weighted images and hyperintensity on T2-weighted images with heterogeneous enhancement as seen in our case⁵. Laryngotracheal endoscopy reveals a solitary, well encapsulated mass arising most often from the posterior

tracheal wall; the lower third of the trachea is the most common site followed by the upper and then the middle thirds^{1,3}. Proper awareness of these lesions is crucial in the pre-operative work-up of patients presenting with stridor. The anesthesiology team should be ready to secure the airway by means other than the conventional ones such as endotracheal intubation. Laryngeal mask anesthesia or jet ventilation, delivering oxygenation proximal to the site of the lesion and avoiding direct manipulation of the tracheal lumen are important issues to keep in mind.

The diagnosis of tracheal schwannomas is primarily pathological. Grossly, they are well circumscribed and encapsulated; microscopically, neural spindle cells in compact fashion (Antoni A areas) as well as in loose myxoid fashion (Antoni B areas) are demonstrated. Verocay bodies, which are areas of palisading nuclei, are present in Antoni A areas³. No capsular invasion, nuclear pleomorphism or mitoses is present. Immunohistochemical stains for S-100 protein and vimentin are positive⁴. These were the findings in our case. The differential diagnosis of spindle cell lesions of the larynx and trachea includes spindle cell carcinoma, as well as a variety of benign and malignant mesenchymal lesions. Spindle cell carcinoma can be easily identified by demonstrating

a direct relationship of the tumor with the adjacent epithelium and by immunohistochemical staining for cytokeratin. Among the benign mesenchymal lesions is nodular fasciitis which may display mitotic figures, but the latter are not atypical. Malignant lesions include a number of spindle cell sarcomas such as fibrosarcoma, synovial sarcoma, and leiomyosarcoma; however, these are extremely rare in this location and do not arise from the surface epithelium.

The rarity of these lesions has led surgeons to practice different surgical techniques but the primary treatment is surgical excision. No standard procedure has been adopted. Endoscopic excision, sleeve excision or tracheal resection, are all commonly accepted treatment modalities. Endoscopic excision using either the cold steel instruments or laser is better suited for pedunculated lesions with no extra-tracheal component¹. Tracheal resection with reconstruction has been more advocated for sessile lesions with or without an extra-tracheal component^{1,2}, which was the procedure of choice in our case.

The duration of follow up after surgical intervention has not been consistent throughout the literature, mainly because of the rarity of the lesion.

References

1. RUSCH VW, SCHMIDT RA: Tracheal schwannoma: management by endoscopic laser resection. *Thorax*; 1994, 49:85-86.
2. HOROVITZ AG, KHALIL KG, VERANI RR, ET AL: Primary intratracheal neurilemoma. *J Thorac Cardiovasc Surg*; 1983, 85:313-320.
3. PANG LC: Primary neurilemoma of the trachea. *South Med J*; 1989, 82:785-787.
4. DORFMAN J, JAMISON BM, MORIN JE: Primary tracheal schwannoma. *Ann Thorac Surg*; 2000, 69:280-281.
5. BABA Y, OHKUBO K, SEINO N, CHUREI H, SHIRAHAMA H, NAKAJO M: MR imaging appearances of schwannoma: correlation with pathological findings. *Nippon-Igaku-Hoshasen-Gakkai-Zasshi*; 1997 Jul, 57(8):499-504.

