Primary Tracheal Schwannoma with extension to the Thyroid Gland: Management

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INTRODUCTION

Primary tracheal tumors are rare. Commonly tracheal neoplasms are either squamous cell carcinoma or adenoid cystic carcinoma.[1]Tracheal Schwannoma are extremely rare and are more often found in the lungs and bronchi.[2] Schwannoma's are tumors that originate from tumorigenic Schwann cells attributed to loss-of-function mutations of NF type2 tumor suppressor gene.[3] Standard treatment has not been established yet. Endoscopic resection has been widely used as a treatment modality but recurrence has always been a possibility. Previous literature has shown that optimal treatment is surgical resection and anastomosis of the involved trachea. Cases such as multiple tracheal schwannoma,[4] transmural tracheal schwannoma,[2,5] transmural schwannoma of trachea compressing the esophagus[6] and plexiform schwannoma involving trachea and left recurrent laryngeal nerve[7]have been reported which were all treated by resection and anastomosis of involved trachea.

We report a similar case of transmural tracheal schwannoma with extraluminal component of tumor extending into the left lobe of thyroid. Initially Nd:YAG laser ablation was performed to relieve the airway obstruction. Patient later underwent surgical resection with end to end anastomosis of trachea with en-bloc resection of left thyroid lobe. This case has been reported in line with the SCARE criteria.[8]

KEY CLINICAL MESSAGE : Tracheal Schwannoma are rare neoplasms and can have spectrum of clinical presentation which causes diagnostic dilemma and delay in diagnosis. So far, tracheal resection and anastomosis has been the optimal management and follow up is essential.

CASE PRESENTATION

A 40 year old male presented to the Out Patient Department on August of 2021 with the complaint of scanty hemoptysis since 4 days. He had recovered from mild COVID pneumonia recently. He denied having fever, shortness of breath or chest pain. Physical examination of chest and neck were within normal limits with no visible neck swelling. No inspiratory or expiratory wheeze.

METHODS

In the background of COVID pneumonia and hemoptysis HRCT was performed where incidentally 29*29*25mm pre-tracheal mass with extension and partial occlusion of the lumen of trachea was noted. For better assessment of the mass, CECT neck and chest was ordered(Figure 1). In addition to the HRCT findings, the pre-tracheal mass was abutting left thyroid lobe of retrosternal extended part of thyroid.

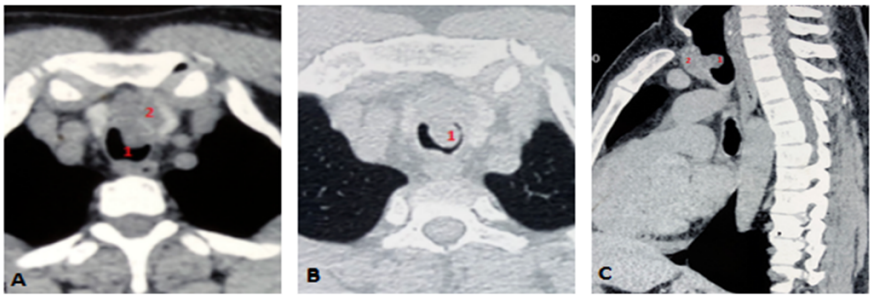

Figure 1. Computed Tomography A) Axial view with 1) intraluminal polypoidal mass 2) extraluminal component of the mass B) Mediastinal Window showing the intraluminal mass occluding 75% of the tracheal lumen C) Sagittal view

Not sure of the origin, either from the thyroid or trachea, an ultrasound neck along with FNAC of the lesion in the neck was performed as it was least invasive with better yield. FNAC revealed it to be Spindle Cell tumor likely a Schwannoma. To better delineate the lesion and as workup for tumor, whole body MRI was performed as PET-CT was not available. MRI was suggestive of single lesion with involvement of thyroid and infiltration into the trachea causing marked obstruction. No lymph nodes were noted in the neck.

With the intent of relieving the airway, biopsy of the tracheal tumor and assessment of the tumor as well as distal bronchial tree, rigid and flexible bronchoscopy with Nd:YAG laser ablation of tumor was performed. Tumor was sessile of size 2*1.5cm in the left posterolateral aspect of the trachea about 2cm below the True Vocal Cords almost occluding 75% of the tracheal lumen(Figure2).

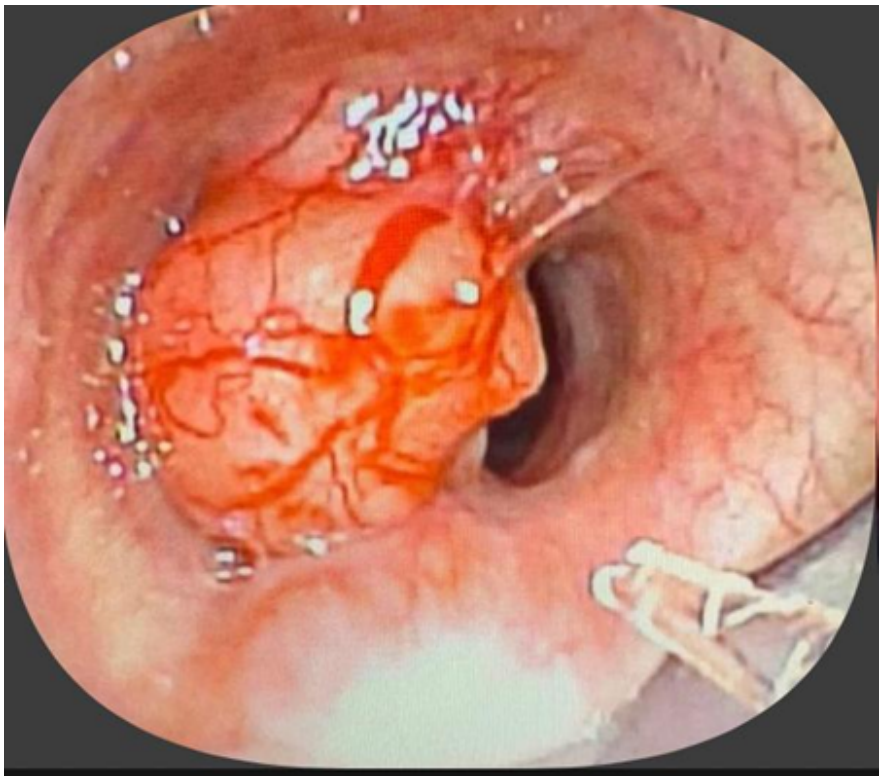

Figure 2. Bronchoscopic view of the tumor showing partial airway obstruction. Superficial blood vessels can

be seen.

Histopathological examination was consistent with tracheal schwannoma. After 4 weeks of laser ablation, he underwent tracheal resection and anastomosis with en- bloc excision of the left thyroid lobe via cervical approach(Figure 3). 5 tracheal rings were excised in the procedure.

RESULTS: Final histopathological examination demonstrated a transmural tracheal mass abutting the left thyroid lobe consistent with tracheal schwannoma. Tracheal resection margins and lymph node were free of tumor. Tracheal tumor was abutting the thyroid lobe but no invasion was seen as thyroid capsule was intact(Figure 4).

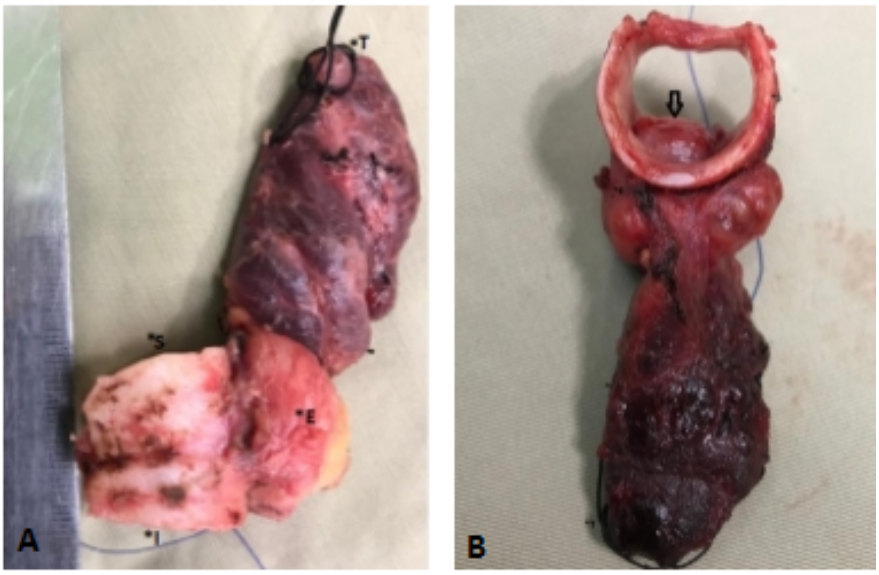

Figure 3. Gross finding A) The excised trachea with en-bloc left thyroid lobe, *T- superior pole of left thyroid lobe, *S- superior margin of the trachea, *I-inferior margin of trachea, *E- extraluminal component of the schwannoma abutting the left thyroid lobe B) Intraluminal component of schwannoma demonstrating growth after laser ablation(shown by the arrow).

Microscopically, hypercellular Antoni A and hypocellular Antoni B areas were noted with foci of nuclear palisading forming verocay bodies(Figure 5). Tumor was S-100 protein and Vimentin positive. Follow up CT scan and bronchoscopy in 6 months showed healed anastomotic site and in 1 year no growth/ recurrence was noted.

CONCLUSION

We experienced extremely rare case of transmural tracheal schwannoma abutting the left thyroid lobe. Subsequently tracheal resection and anastomosis with en-bloc excision of left thyroid lobe was performed. Follow up is necessary for surveillance of local recurrence.

DISCUSSION

Schwannoma and neurofibroma arise from peripheral nerves. Schwannoma usually occurs in head, neck, retroperitoneum, extremities and mediastinum.[9] Tracheal schwannoma is rare and was first reported by Straus et.al in 1951[10] and is believed to originate from nerves inside the tracheal wall.[2] Schwannoma is common in adults with female predilection.[4] It is frequently found in the lower third of the trachea followed by proximal trachea and middle third of trachea.[11]

Symptoms are misleading in these patients as clinicians mistake dyspnea, wheezing or cough caused by tracheal neoplasm for symptoms of COPD which causes delay in diagnosis of 10-15 months in average.[12] Hemoptysis is uncommon symptom for schwannoma and is predominantly found in squamous cell carcinoma.[6]

In spirometry, characteristic flow-volume loop with flattened inspiratory and expiratory limbs is suggestive of fixed upper airway obstruction.[13] CT scan in Schwannoma demonstrates well-demarcated, low density mass prior to contrast and contrast enhancement after injection of the contrast. CT is essential as it helps to define the location of tumor and allows assessment of luminal and extrinsic extent of tumor.[5] Bronchoscopic evaluation is indispensable in the localization of the tumor in relation to cricoid and carina, to determine the length of the luminal disease, assessment of distal bronchial tree and biopsy of the tumor as well. Assessment of vocal cord function is important as thyroid carcinoma associated with tracheal invasion is seen in about 6% of patients presenting for resection and anastomosis.[14] MRI shows isointensity or hypointensity on T_1 weighted images and hyperintensity on T_2 weighted images with heterogenous enhancement.[12]

According to Kashara et.al[15] pulmonary neurilemmomas can be divided into two types a) Central type b) Peripheral type. In terms of relationship between tumor and tracheal luminal space, central type is further classified into two subtypes 1) Intraluminal type- tumor is confined in the intraluminal space 2) Transmural type- tumor occurs in both intraluminal space and extraluminal space(combined type). We encountered a transmural type of tracheal schwannoma with extraluminal component contiguous to the left thyroid lobe.

Treatment of tracheal schwannoma can either be endoscopic resection or surgical resection based on size, location and extent of the tumor. Endoscopic treatment does not always achieve or confirm complete resection and is applicable in patients with pedicled and intraluminal type tumor, patients with advanced age, limited life expectancy, with severe co-morbidity and in whom complete resection cannot be offered.[16] Complete resection of tracheal schwannoma with bronchoscopic technique has been reported[17] but one should always look out for local recurrence. Local destruction of tumor by core out, Nd:YAG laser ablation, cryotherapy and primary radiotherapy by external beam can be used in addition or as as alternative to endoscopic resection as a tool to relieve the airway obstruction caused by tracheal neoplasm.[9,11,15,16]Surgical resection and anastomosis has been the optimal management where tumors are sessile and broad based, transmural type tumor, in patients with preserved cardio-pulmonary function and in recurrent schwannoma.[9,16]

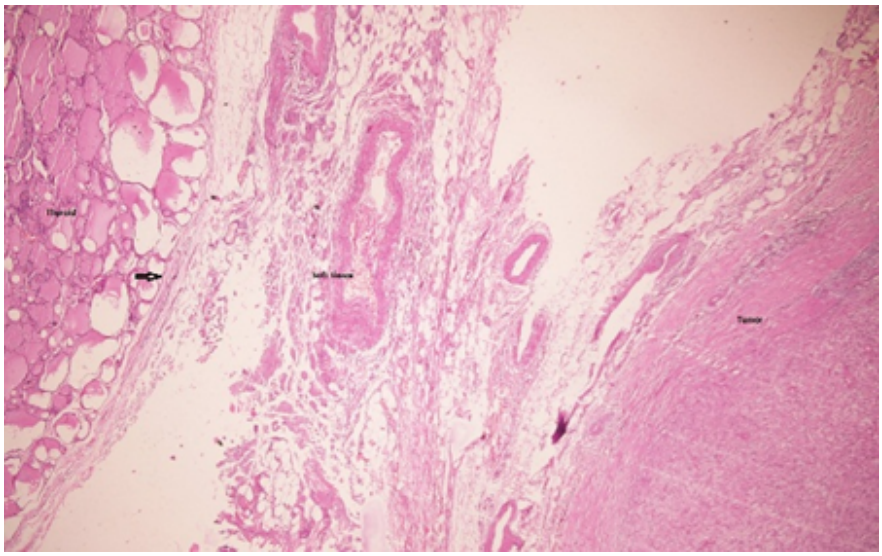

Figure 4. H&E stain shows the intact thyroid capsule (shown by the arrow) and schwannoma separated by soft tissues

In histopathological examination of H&E stain , schwannoma have characteristic hypercellular Antoni A and

hypocellular Antoni B areas. Within Antoni A areas, verocay bodies are seen which are bands of fusiform nuclei alternating with clear zones devoid of nuclei. Schwannoma are usually S-100 and Vimentin positive[18]as seen in our case.

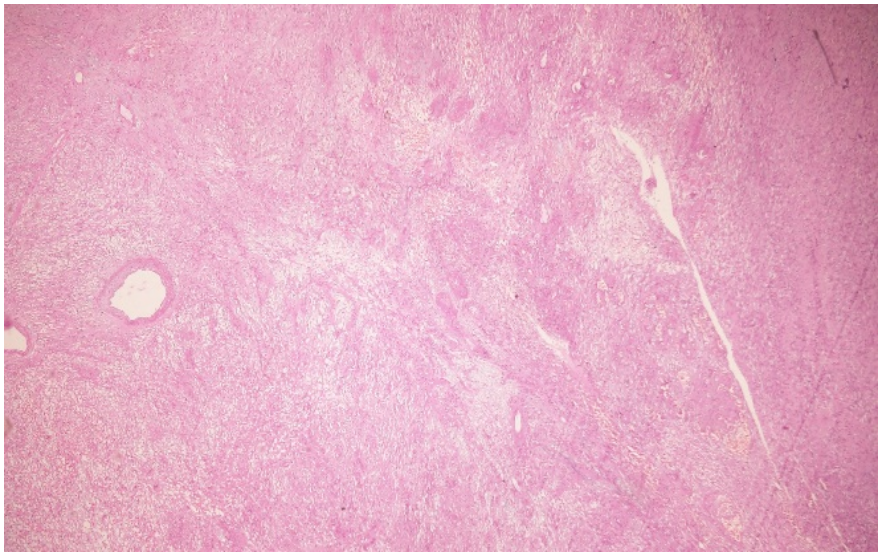

Figure 5 H&E stain showing the Antoni A and Antoni B with nuclear palisading seen in Schwannoma

Abbreviations

CECT- Contrast Enhanced Computed Tomography, FNAC- Fine Needle Aspiration Cytology, HRCT- High Resolution Computed Tomography, MRI- Magnetic Resonance Imaging, Nd:YAG- Neodymium Yttrium Aluminum Garnet, COPD- Chronic Obstructive Pulmonary Disease

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