Dumbbell posterior mediastinal schwannoma invading trachea: Multidisciplinary management – weight off the chest

Sir,

Neurogenic tumors represent approximately 35% of all pediatric and 20% of all adult tumors.[1,2] They are the most common cause of a posterior mediastinal mass. Schwannoma is the most common benign mediastinal neurogenic tumor originating from the neuroectodermal embryonic cells and is characterized by the proliferation of neural sheath Schwann

cells of the peripheral, cranial, or autonomic nerves. Primary involvement of trachea with schwannoma is rare.[3,4] Dumbbell or hourglass growth pattern is recognized when tumor commonly extends to vertebral canal and spinal cord from posterior mediastinum, but the same growth pattern invading the trachea is more rarely reported.[5-9] We herein present a patient with a mediastinal schwannoma invading the trachea treated successfully by combined bronchoscopic and surgical resection.

A 44-year-old previously healthy male was referred to our hospital with complaints of productive cough and gradually increasing breathlessness for 5 months, followed by worsening of symptoms and an audible monophonic wheeze for the last 1 month. He denied any history of other respiratory complaints, fever or weight loss. He was a nonsmoker with no other pulmonary or cardiac disease. He had been treated along the lines of bronchial asthma with bronchodilators and inhaled steroids during the course of illness with no relief of symptoms. The general as well as physical examination findings were unremarkable except an audible wheeze heard over the trachea. Neurofibromatosis was excluded as there was no evidence of café au lait spots, multiple neurofibromas, or hyperpigmented skin lesions on dermatological examination. Laboratory investigations were also within normal limit. Spirometry revealed evidence of obstruction in expiratory loop of flow-volume curve. Computed tomography (CT) of the thorax showed posterior mediastinal mass invading trachea with intraluminal extension [Figure 1]. He underwent diagnostic fiberoptic bronchoscopy (BF-1TQ190, Olympus Corp., Japan) that revealed a polypoid growth with broad base and overlying vessels on right posterolateral wall in the middle third of the trachea located 3 cm proximal to vocal cords at level of fourth tracheal cartilage ring occluding nearly 50% of tracheal lumen and involvement of three cartilage rings [Figure 2]. Polypoid growth was excised with combination of electrosurgical snaring (Olympus™ reusable electrocautery snare, Germany) and cryodebridement therapy (ERBE™ contact cryotherapy console; flexible cryoprobe with 1.9 mm tip, Germany) with use of rigid bronchoscope (Size 14 tracheoscope, Karl Storz Endoskope, Germany) under general anesthesia, and patency of tracheal lumen was achieved. Hemostasis was achieved with argon plasma coagulation (APC) (ERBE™ electrocautery and APC unit, Germany), injection adrenaline (1 ml of 1 in 10,000), and tranexamic acid. The patient had symptomatic relief after procedure.

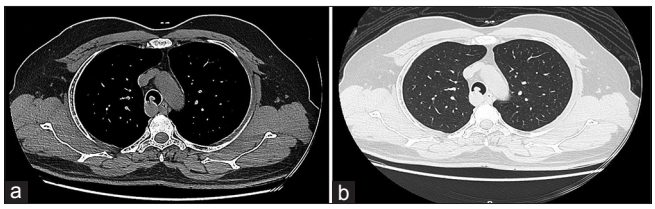

Figure 1: Axial cuts of computed tomography scan thorax mediastinal (a) and lung (b) window demonstrating posterior mediastinal mass invading trachea with intraluminal extension

Esophagoscopy was also done to rule out any mucosal abnormality suggestive of tracheoesophageal fistula. Histopathological analysis of the biopsy specimens was suggestive of benign neurogenic tumor likely schwannoma (Antoni A consisting of spindled cells and elongated nuclei arranged in a palisading pattern surrounding an ovoid mass of eosinophilic cytoplasm with Verocay bodies and extremely scanty mitotic figures) [Figure 3]. Immunohistochemistry further supported the diagnosis (S-100 positive). Subsequently, he underwent right posterolateral thoracotomy for excision of mediastinal mass along with involved segment of trachea and end-to-end anastomosis after consultation with thoracic surgeon. However, extensive resection could be avoided as tumour could be shelled out easily and the tracheal dehiscence was closed with primary sutures. The resected specimen was large encapsulated mass [Figure 3f] originating in posterior mediastinum adhered posterolaterally in the middle third of the trachea. Histopathological and IHC examination findings of resected specimen demonstrated same histological pattern as observed for bronchoscopic specimen. Regional mediastinal lymph node sampling was also performed, and findings were unremarkable. He became asymptomatic postprocedure with flow-volume loop reverted to normal when repeat spirometry was performed. He is under regular follow-up since last 1 year with no evidence of recurrence on periodic bronchoscopic surveillance.

Schwannomas and neurofibromas are the two most common neurogenic tumors of the posterior mediastinum. These tumors are more common in adults and do not show a predilection for sex.[2] They are mostly benign, slow-growing tumors originating primarily from a spinal nerve root but may involve any thoracic nerve commonly involving intercostal nerves and autonomic nerves, especially sympathetic chain. Schwannoma is also termed as neurilemmoma, neurinoma, or perineural fibroblastoma.

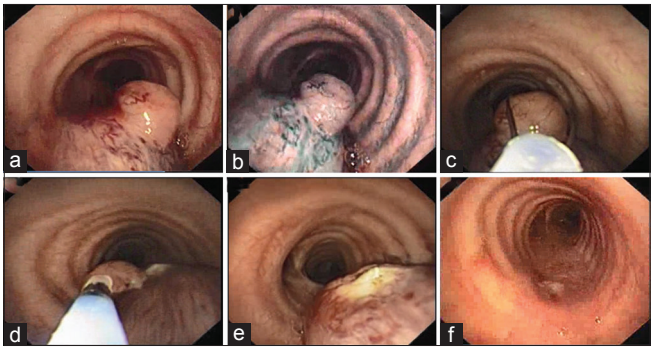

Figure 2: (a) Bronchoscopic view showing polypoid growth with broad base and overlying vessels on right posterolateral wall of the trachea occluding nearly 50% of tracheal lumen (b) narrow band imaging showing unbranched vessels with loss of tortuosity overlying tumor growth (c) electrosurgical snare placement around polypoid tracheal tumor pedicle. (d) Tumor removal (cryodebridement) with cryoprobe. (e) Residual anatomy after tumor removal. (f) Posterior wall of trachea showing no recurrence post resection during bronchoscopic surveillance after 1 year

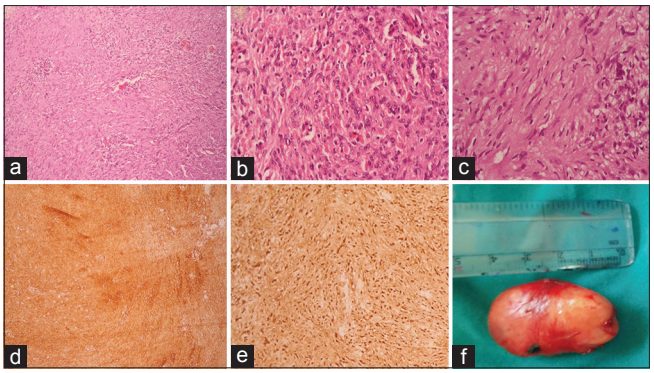

Figure 3: Histopathological slides of bronchoscopic tumor specimen: (a and b) Antoni A pattern showing spindled cells and elongated nuclei arranged in a palisading pattern surrounding an ovoid mass of eosinophilic cytoplasm (H and E, ×40 and ×100 respectively) with (c) Verocay body; (d and e) tumor showing S-100+ on IHC staining (×40 and ×100 respectively); (f) resected specimen showing large encapsulated posterior mediastinal mass with size 4 cm × 2.5 cm × 2 cm

They arise either from neural sheath schwann cells or associated fibrous connective tissue causing extrinsic compression to nerve fascicles or bundles. Malignant form of schwannomas is usually rare but occurs in more than 50% of patients with neurofibromatosis.[2] Transformation usually occurs commonly to malignant peripheral nerve sheath tumor and less commonly angiosarcoma or epithelioid malignant change. Gross examination of schwannoma is being described as usually solitary, soft, yellow-tan, encapsulated, and well-demarcated lesions arising from the periphery of the nerve of origin. These tumors can show dumbbell growth pattern with extension to vertebral canal from posterior mediastinum. This similar growth pattern may be shown by either extension into the tracheal lumen from posterior mediastinum through the intercartilaginous membrane presenting as polypoid or pedunculated growth or primary involvement of trachea leading to intraluminal growth as well as extraluminal extension.[10] The growth occurring primarily in trachea arises from intraluminal nerve tissue, especially from Schwann cells of the nerve sheath. Microscopic examination shows Schwann cells in a background of loose reticular tissue arranged in two distinct types of tissue pattern, Antoni A corresponding to the cellular spindle cells arranged in short bundles or interlacing fascicles and Antoni B corresponding to hypocellularity and myxoid changes. These two forms are usually associated with the same tumor with no evidence of mitoses in benign variety. The rare malignant variety has potential to exhibit other cellular components such as epithelial cells, mucinous glands and mesenchymal structures and focal staining with S-100 on immunohistochemistry. The symptoms are determined by the location and size of the tumors. Patients with tumors in trachea are usually asymptomatic in the early stage of disease. The patients will usually present with some nonspecific symptoms such as dyspnea, dry cough, and wheezing when there is more than 50% occlusion of trachea.[11] Other symptoms encountered less frequently are hemoptysis, recurrent respiratory infections, and chest pain. These patients are prone to be misdiagnosed and treated as some respiratory diseases such as asthma as observed in our case. The initial imaging to identify location of mediastinal mass is chest radiography. It may reveal a filling defect in air column if tracheobronchial tree is invaded by tumor. CT thorax is superior imaging modality that can provide accurate localization of mediastinal masses and its relationship to adjacent structures including dumbbell growth invading trachea.[12] The three-dimensional reconstruction CT scan can provide better details, especially distal to growth where bronchoscopic view is not accessible. Magnetic resonance imaging allows the study of the invasion of the vascular structures of the mediastinum and the spinal canal with better precision that can influence surgical management. Fiberoptic bronchoscopy is important in determining the appearance and extent of the lesion. It has been used as definitive treatment by using different modalities such as snaring, electrocautery, APC, cryodebridement therapy, and endoscopic laser therapy for resection of tumor.[3,4,13-17] Bronchoscopic treatment is curative, especially if tumor is pedunculated and completely intraluminal or in patients with poor cardiorespiratory reserve.[3] However, rigid bronchoscopy should be preferred as it provides for use of multiple instruments, better control of specimen after resection, and control of bleeding that could occur after biopsy. The treatment of the benign mediastinum schwannoma is surgical, with resection through open thoracotomy or video-assisted thoracoscopy. Resection of the involved tracheal portion, with end-to-end anastomosis, is required if there is involvement of trachea.[3,18] Bloc surgical resection followed by adjuvant chemoradiotherapy is the desired treatment modality for malignant schwannoma or recurrence after bronchoscopic resection. However, the benefit of adjuvant therapy is questionable.[1,3] Periodic bronchoscopic surveillance is mandatory to detect recurrence. We have treated our case with multimodality approach by resecting intraluminal component of tumor with electrosurgical snaring and cryodebridement therapy combined with APC through rigid bronchoscopy followed by surgical excision of mediastinal component with involved tracheal segment without any recurrence. Gold standard for management is still lacking because of rare occurrence of this tumor. These tumors should be diagnosed early by imaging. Complete surgical resection is preferred if there is malignant transformation. Multidisciplinary approach is required for effective management of such tumors.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given consent for images and other clinical information to be reported in the journal. The patient understands that their names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

Abhijeet Singh, Vallandramam R Pattabhiraman, Arjun Srinivasan, Sivaramakrishnan Mahadevan

Department of Interventional Pulmonology and Sleep Medicine, Royal Care Super Speciality Hospital, Coimbatore, Tamil Nadu, India

E-mail: vr.pattabhiraman@gmail.com

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