

Case Report

Massive Hemoptysis: A Rare Presentation of Posterior Mediastinal, Giant, Benign Vagal Schwannoma

Anirban Das MD¹, Sabyasachi Choudhury MD¹, Sumitra Basuthakur MD¹, Anghsuman Mukhopadhyay MD¹, Siddhartha Mukherjee MCH²

Abstract

Schwannomas are rare neurogenic tumor originating from Schwann cells of the nerve sheath, most frequently encountered type of posterior mediastinal tumors. In most cases, schwannomas are benign, malignant and multiple schwannomas are rare. Histopathologically, the tumor is composed of fascicles of spindle cells, which are strongly positive for S-100 proteins. Surgical resection is a treatment of choice, and prognosis is excellent. Here, we report a case of posterior mediastinal schwannoma in a 20-year-old male patient who complained of right-sided back pain and two episodes of massive hemoptysis of recent onset. Contrast enhanced computed tomography (CECT) and magnetic resonance imaging of the chest showed a well circumscribed, heterogeneous mass in the posterior mediastinum, compressing the right lower lobe with widening of intervertebral foramina. CT-guided trucut biopsy revealed spindle cell neoplasm. On immunohistochemistry, tumor cells expressed strong positivity for S-100 protein. Final diagnosis was schwannoma, probably originating from the right vagus nerve. Surgical resection of the encapsulated tumor resulted in the successful recovery, without any recurrence over next one year follow up.

Keywords: Neurogenic tumor, posterior mediastinum, schwannoma, surgical resection, tru-cut biopsy, vagus nerve

Cite this article as: Das A, Choudhury S, Basuthakur S, Mukhopadhyay A, Mukherjee S. Massive hemoptysis: A rare presentation of posterior mediastinal, giant, benign vagal schwannoma. *Arch Iran Med.* 2014; **17(11)**: 779 – 782.

Introduction

Neurogenic tumors account for 20% of all primary mediastinal neoplasms in adults¹ and most of them are located in the posterior mediastinum. On the other hand, neurogenic tumors are the most common type among posterior mediastinal tumors. Neurogenic tumors originate from nerve sheath, ganglion cells, or paraganglion cells. Schwannomas are most frequently encountered type of neurogenic tumors of the mediastinum and originate from Schwann cells of nerve sheath. In most cases, they are benign in nature and occasionally malignant. These tumors are usually asymptomatic and may be detected on a routine chest radiograph. Very few cases of mediastinal schwannomas are reported in the literature, where they are originating from nerve sheaths of vagus or phrenic nerves.² Here, we present a rare case of posterior mediastinal schwannoma probably originating from right vagus nerve in a 23-year-old male patient who complained of right-sided back pain with two episodes of massive hemoptysis of recent onset.

Case Report

A 23-year-old normotensive, non-diabetic, non-smoker male presented with dull aching back pain on right side and dry cough

Authors' affiliations: ¹Department of Pulmonary Medicine, Medical College, Kolkata, West Bengal, India, ²Department of Cardiovascular & Thoracic Surgery, Medical College, Kolkata, West Bengal, India.

Corresponding author and reprints: Anirban Das MD, Department of Pulmonary Medicine, Medical College, Kolkata, West Bengal, India-700 073. Address: C/o Mr. Pradip Das, Peon Para, Bhatchala, Near Vidyarthi Girls' High School, P.O.: Sripalli, P.S.: Burdwan Sadar, Dist.: Burdwan West Bengal, India-713 103. Tel: + 91 9434672168,

E-mail: dranirbandas_chest@rediffmail.com

Accepted for publication: 20 June 2014

with two episodes of massive hemoptysis for last three months. There was no history of chest trauma, shortness of breath, wheeze fever, anorexia, and weight loss. On a general survey, pallor was present, but the absence of clubbing, and palpable lymph nodes were reported. His temperature was 97°F, respiratory rate 16 breaths/minute, pulse rate 96/minute, and blood pressure 110/80 mmHg. Examination of the respiratory system and other systems revealed no abnormality.

Complete hemogram and blood biochemistry were within normal limit, except hemoglobin, which was 9.2 g/dL. Chest radiograph showed a right paracardiac mass with smooth convex margin, (Figure 1). Contrast enhanced computed tomography (CECT) of the thorax revealed a well circumscribed, heterogeneous, soft tissue mass lesion in posterior mediastinum measuring 67 mm x 50 mm, compressing right lower lobe, (Figure 2). No consolidation, pleural effusion or pulmonary nodule in either of the lung fields, hilar, or mediastinal lymphadenopathy, and no evidence of erosion in the adjacent vertebra, or ribs were reported. Magnetic resonance imaging (MRI) of mediastinum showed a well encapsulated, posterior mediastinal heterogeneous mass with intervertebral widening due to its extension (without vertebral erosion) as well as invasion of surrounding vessel and right lower lobe, (Figure 3). CT guided fine needle aspiration cytology (FNAC) showed bundles of spindle cells with plump nuclei and few cells showed nuclear polymorphism and hyperchromasia, suggestive of spindle cell neoplasm. CT guided trucut biopsy revealed a tumor composed of interlacing fascicles of spindle shaped cells with elongated nuclei and very few mitotic figures, suggestive of spindle cell lesion, (Figure 4). Immunohistochemistry showed that tumor cells express S – 100 proteins, but were immunonegative for desmin, SMA, CD 34, HMB 45, CD 21, and CD 35, (Figure 5). Therefore, the diagnosis of benign schwannoma was established.

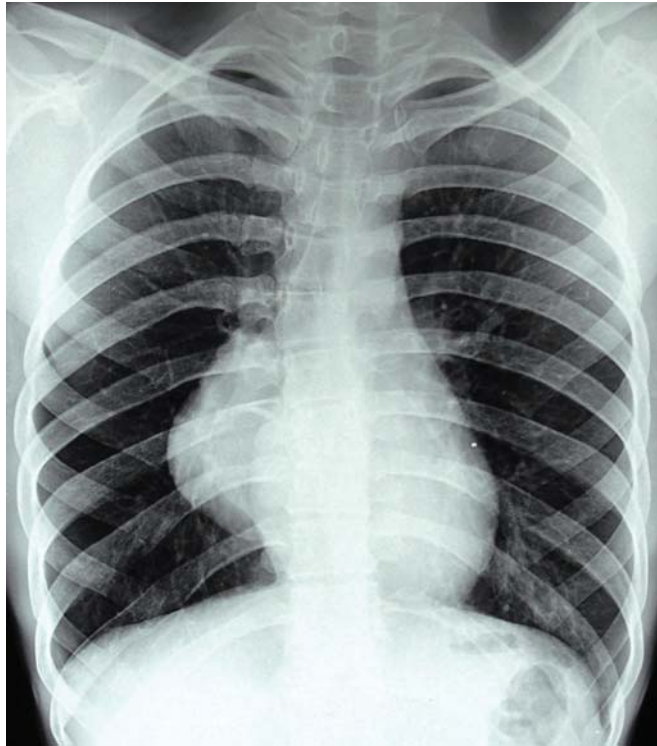


Figure 1. Chest radiograph showing a right paracardiac mass.

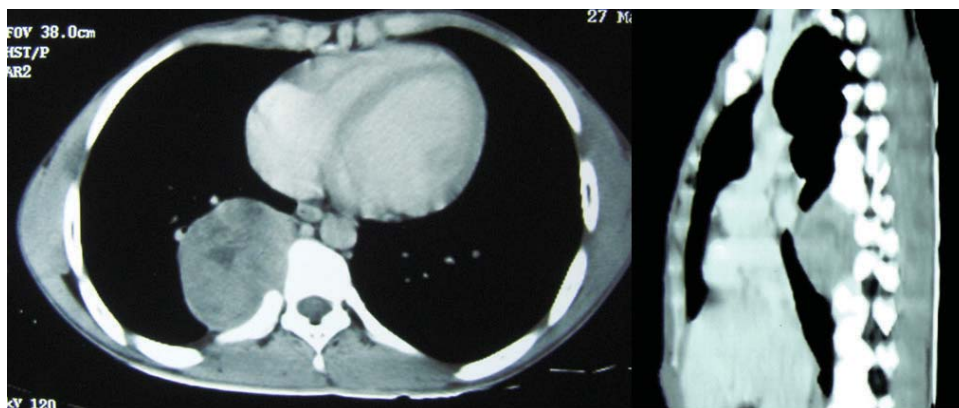


Figure 2. CECT thorax showing a well-circumscribed, and a heterogeneous mass in posterior mediastinum.

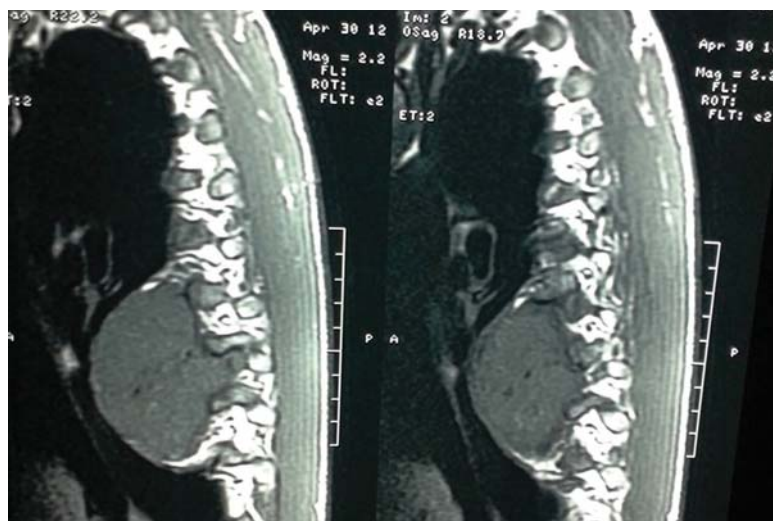


Figure 3. MRI of mediastinum showing a posterior mediastinal, and a heterogeneous encapsulated mass with intervertebral widening.

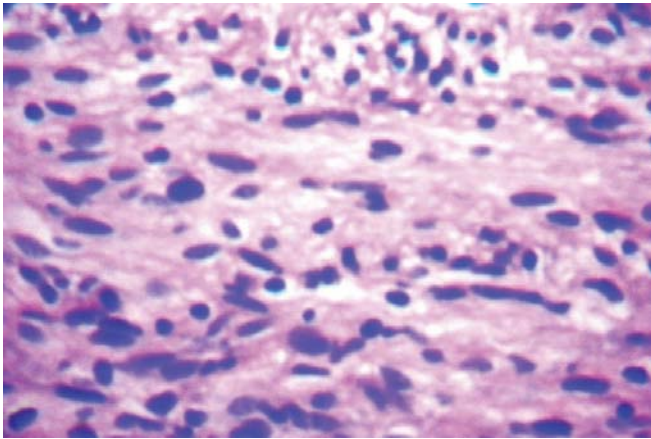


Figure 4. Photomicrograph of CT – guided tru cut biopsy specimen showing spindle cell neoplasm, (H&E stain, x 100).

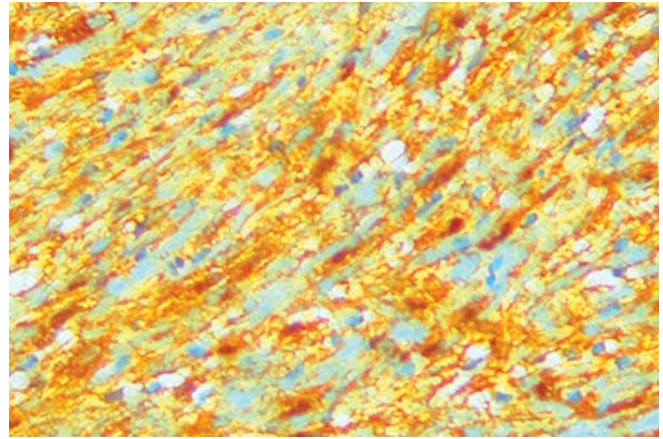


Figure 5. Immunohistochemistry showing S – 100 positivity of tumor cells, (x 200).



Figure 6. Photograph showing schwannoma in-situ

Fibreoptic bronchoscopy revealed narrowing of right lower lobe bronchus without any endobronchial tumor. No biopsy was taken and bronchoalveolar lavage fluid analysis was inconclusive. The patient was successfully treated by complete surgical resection of the tumor, using an open thoracotomy via posterolateral approach, (Figure 6). No recurrence happened during the next one-year follow up. After opening, it was found that right vagus nerve was passing through the enormous paraspinal tumor, which showed parenchymal invasion into the right lower lobe and adjacent small vessels. En bloc resection of encapsulated tumor and surrounding lung parenchyma was done, although preservation of normal lung parenchyma and the right vagus nerve was tried as far as possible. Macroscopically, resected mass was grayish white in color, ovoid, encapsulated. It was heterogamous in appearance with haemorrhagic and yellowish necrotic areas. Consistency of the resected tumour was firm. It was $8 \times 5 \times 4$ cm in size. Weight of the specimen was 1020 g. Findings on histopathological and immunohistochemical examination of the specimen were similar to trucut biopsy specimen, hence, the final diagnosis was posterior mediastinal benign giant schwannoma of the right vagus nerve.

Discussion

According to our findings, 12% – 39% of all mediastinal tumors are neurogenic tumors, 90% occur in the posterior mediastinum, and 95% of these posterior mediastinal neurogenic tumors are schwannomas.³⁻⁵ Mediastinal neurogenic tumors originate from nerve sheath cells (schwannomas or neurofibromas), paraganglion cells (chemodectomas, pheochromocytomas) or from ganglion cells (neuroblastomas, ganglioneuroblastomas, ganglioneuromas).³ Almost all mediastinal neurogenic tumors are benign in character with rare exceptions. The differential diagnoses of the posterior mediastinal mass include neurogenic tumors, meningocele, lymphoma, enteric cysts, bronchogenic cysts, esophageal tumors, aneurysms, parasitic cyst, paraspinal abscess, and etc.

Schwannomas or neurilemmomas originate from schwann cells of nerve sheath and compress the nerve extrinsically. Schwannomas are lobulated, encapsulated spherical benign masses, and affect both male and female equally. These tumors have predominantly seen in third and fourth decades.² These tumors are slow growing and their malignant transformation risk is very low.⁶ Benign schwannomas are asymptomatic in most cases and are de-

tected on routine chest radiograph. Constitutional symptoms like fever, weight loss, and anorexia may be present. Symptomatic lesions raise the suspicion of malignant schwannomas. Features of obstruction or compression may be present due to invasion of mediastinal structures. In this context, superior vena caval obstruction, phrenic nerve palsy, and Horner's syndrome are occasionally associated with schwannomas. Dumb bell tumors may develop when paraspinal schwannomas extend through the intervertebral foramen into the thoracic cavity. In our case, the patient presented with right sided dull aching back pain and massive hemoptysis, which is unusual in benign mediastinal schwannoma and rarely reported in the literature.⁷ Probably invasion of surrounding pulmonary parenchyma and adjacent small vessels might be the source of hemoptysis. Resection of the tumor and infiltrated parenchyma were the reasons for the cessation of the hemoptysis.

They are seen as a well-defined lobulated mass lesion on chest X-ray without any calcification. On CECT thorax, they appear as well demarcated, homogenous, low density (10 – 15 Hounsfield unit) mass lesion with little contrast enhancement (20 – 30 Hounsfield unit), without any calcification, or fat. On magnetic resonance imaging (MRI) of mediastinum, schwannomas have low – to intermediate signal intensity on T1-weighted images and have intermediate – to high – signal intensity on T2-weighted sequence. MRI may be helpful to assess the extension of tumor to adjacent mediastinal structure.⁸

Macroscopically, they are encapsulated, heterogeneous mass with cystic degeneration, attached to epineurium of the nerve.⁵ Microscopically, the tumor is composed of spindle cells in fascicles, which may appear as dense cellular area (Antoni A) or as hypocellular area (Antoni B), and on immunohistochemistry, tumor cells strongly express S – 100 protein.^{9,10} Malignant schwannomas are very rare, and are histopathologically differentiated from benign schwannomas by the presence of atypia, mitoses, nuclear pleomorphism, hyperchromasia, and necrosis.¹¹

Complete surgical resection by open thoracotomy via posterolateral approach or by video assisted thoracoscopic surgery (VATS) is the treatment of choice, and offers an excellent prognosis.^{6,12} In our case, it was found that right vagus nerve was passing through the paraspinal tumor and the neoplasm originated from the vicinity of the nerve sheath of the nerve, hence it was assumed that it was a giant vagal schwannoma which is again rare entity, reported in the literature. Only 4% – 6% of the neurogenic tumors originat-

ing from the nerve sheaths of the vagus or phrenic nerve.² On the other hand, left side vagal schwannoma is more common, right sided tumor is rare, which was noted in our case.¹³

Although rare, prognosis of benign mediastinal lesions like schwannomas is very good among the other intrathoracic neoplasms. Well-circumscribed mass lesion in thorax raise the suspicion of benign lesion, especially in younger individuals, which is subsequently confirmed by histopathology and immunohistochemistry, and definitely the direction of management differs from the malignant one. Hence, the tissue diagnosis and immunohistochemistry are essential to establish the nature of the neoplasms before going to make the treatment strategy, especially in such cases of rarer benign neoplasms of the mediastinum.

References

1. Strollo DC, Rosado-de-Christenson ML, Jett JR. Tumors of the middle and posterior mediastinum. *Chest*. 1997; **112**: 1344 – 1357.
2. Davis CJ, Butchart EG, Gibbs AR. Neurilemmoma of the intrathoracic vagus nerve. *Eur Respir J*. 1991; **4**: 508 – 510.
3. Whooley BP, Urschel JD, Antkowiak JG, Takita H. Primary tumors of the mediastinum. *J Surg Oncol*. 1999; **70**: 95 – 99.
4. Benjamin SP, McCormack LJ, Effler DB, Groves LK. Primary tumors of the mediastinum. *Chest*. 1972; **62**: 297 – 303.
5. Reeder LB. Neurogenic tumors of the mediastinum. *Semin Thorac Cardiovasc Surg*. 2000; **12**: 261 – 67.
6. Fierro N, D'Ermo G, Di Cola G, Gallinaro LS, Galassi G. Posterior mediastinal schwannoma. *Asian Cardiovasc Thorac Ann*. 2003; **11**: 72 – 73.
7. Bandyopadhyay S, Dhua D, Maiti A, Maity PK. Intrathoracic schwannoma presenting with massive haemoptysis. *J Assoc Physicians India*. 2013; **61**: 42.
8. Hayasaka K, Tanaka Y, Soeda S, Huppert P, Claussen CD. MR findings in primary retroperitoneal schwannoma. *Acta Radiol*. 1999; **40**: 78 – 82.
9. Marchevsky AM. Mediastinal tumors of peripheral nervous system origin. *Semin Diagn Pathol*. 1999; **16**: 65 – 78.
10. Knight DM, Birch R, Pringle J. Benign solitary schwannomas: a review of 234 cases. *J Bone Joint Surg Br*. 2007; **89**: 382 – 387.
11. Shoji F, Maruyama R, Okamoto T, Wataya H, Nishiyama K, Ichinose Y. Malignant schwannoma of the upper mediastinum originating from the vagus nerve. *World J Surg Oncol*. 2005; **3**: 65 – 68.
12. Sakumoto N, Inafuku S, Shimoji H, Nomura K, Honma K, Kawabata T, et al. Videothoracoscopic surgery for thoracic neurogenic tumors: a 7-year experience. *Surg Today*. 2000; **30**: 974 – 977.
13. Huang TW, Yang MH, Cheng YL, Tsai WC, Lee SC. Vagus nerve schwannoma in the middle mediastinum. *Thorac Cardiovasc Surg*. 2010; **58**: 312 – 314.