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Case Report

Cystic Schwannoma of the Left Phrenic Nerve Presented as Left Paracardiac Cystic Lesion, Case Report and Literature Review

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Abstract

Introduction: Schwannomas are mostly benign tumors in adults and usually an incidental finding. Mediastinum is the second most common site of schwannoma. Mediastinal schwannomas rarely arise in the anterior or middle mediastinum. Less than 10 cases of mediastinal phrenic nerve schwannomas are reported in the English literature and all of them were solid without cystic degeneration. We present a case of cystic schwannoma that originates from the left phrenic nerve in paracardiac location.

Case Presentation: 48 year-old female presented with dry cough. Initial chest x-ray (CXR) showed left cardiophrenic angle mass silhouetting the left cardiac border. A commuted tomography (CT) was performed and revealed multiloculated left paracardiac cyst with few enhancing septations. And for further characterization, an MRI was and confirmed the cystic lesion with better delineation of the enhancing septations. Video assisted thoracoscopic surgery was performed and easily resected. Histological examination revealed two tissue types: Antoni A and Antoni B. Sectioning showed multiloculated cystic lesion containing brown, serous fluid, gelatinous tan material and blood clots. The tumor demonstrated cyst formation, hyalinization, sheets of histiocytic infiltration with no atypia, mitosis or necrosis. Tumor cells composed of bland spindle cells with verocay body formation. S100 and EMA (Epithelial Membrane Antigen) were positive. Pankeratin, demin, HMB45 and CD117 were negative.

Discussion: Most of the mediastinal lesions arise in the posterior mediastinum. 45% of schwannomas occur in the head and neck, and 9% occur in the mediastinum. Most adult's nerve sheath tumors are benign and arise from an intercostal nerve, sympathetic nerves or parasympathetic chains, spinal ganglions, rarely arising from the pericardium, phrenic nerve or vagus nerve. Imaging studies play an important role in diagnosis and localization of the mediastinal lesions. On CXR, these lesions manifest usually as mediastinal lesions or mediastinal contours abnormalities. CT can greatly assists in determining the exact location, its components and relationship to adjacent structures. MRI provides superior soft tissue characterization and can help better delineate the relationship of an identified mediastinal mass to nearby intrathoracic vascular structures.

Conclusions: Cross sectional imaging plays an important role in the diagnosis of cystic mediastinal lesions. Cystic schwannoma of the intrathoracic phrenic nerve, although very rare, might be considered in the differentials diagnosis of cystic mediastinal lesions if the lesion is following the course of phrenic nerve.

Keywords: *Phrenic Nerve Schwannoma; Cystic Schwannoma; Paracardiac Cysts; Mediastinal Schwannoma*

Introduction

Intrathoracic neurogenic tumors are common tumors that generally found in the posterior mediastinum and mostly located in the intercostal nerves or sympathetic chain. Two thirds of these tumors arise from the nerve sheaths (Schwannomas and Neurofibromas) and the rest from the nerve cells (neurocytic tumors). Schwannomas are 90% benign tumors in adults and usually of incidental finding. They are composed of myelinated perineural (Schwann) cells intermixed in a connective tissue stroma [5]. Schwannoma is a benign nerve sheath neoplasm of Schwann cell origin. Mediastinum is the second most common site of schwannoma after the head and neck with the majority of mediastinal schwannomas arising in the posterior mediastinum mainly from the paraspinal nerves. Mediastinal schwannomas rarely arise in the anterior or middle mediastinum. Neurogenic tumors arising from the intrathoracic phrenic nerve are extremely rare; the most common of which is schwannoma. Less than 10 cases of phrenic nerve schwannomas are reported in the English literature most of them were solid without degeneration and most of which are located in the upper mediastinal, paratracheal (middle mediastinum) or parahilar regions [2-7]. We present a case of cystic schwannoma that originates from the left phrenic nerve in a left paracardiac region.

Case Presentation

48-year-old female patient, known case of hypothyroidism and controlled hypertension initially presented with dry cough. Pulmonary function test was normal. Physical examination and blood work up were normal. Chest X-ray (not shown) showed a left cardiophrenic angle mass silhouetting the left cardiac border. To clarify the findings and to rule out lung disease as a cause of the cough and dyspnea, a CT scan of the chest was ordered. The enhanced CT chest showed a well-defined 3 x 2 cm multiloculated left paracardiac cystic lesion that contains few enhancing septations most of which were thin. In some areas the septations were thicker and nodular. The content of the lesion was mostly of fluid attenuation however it was denser in some of the locules reaching up to 40 HU density. Few scattered foci of calcifications noted within the lesion mainly in the septa (Figure 1). At that stage, our differential diagnosis included bronchogenic cyst, infected thymic or pericardial cyst, less likely germ cell tumor (given the patient's age) and hydatid cyst. MRI was performed to further characterize the lesion. MRI confirmed the cystic nature of the lesion and better delineated the enhancing septations, most of which were thin. There were few areas of nodular enhancement and thicker septa measuring up to 6 mm. Multiple fluid-fluid levels noted within the lesion. MR excluded the presence of fat within the lesion (Figures 2a, 2b and 3). The differential diagnosis of the complex cystic lesion based on the MR did not differ from the CT.

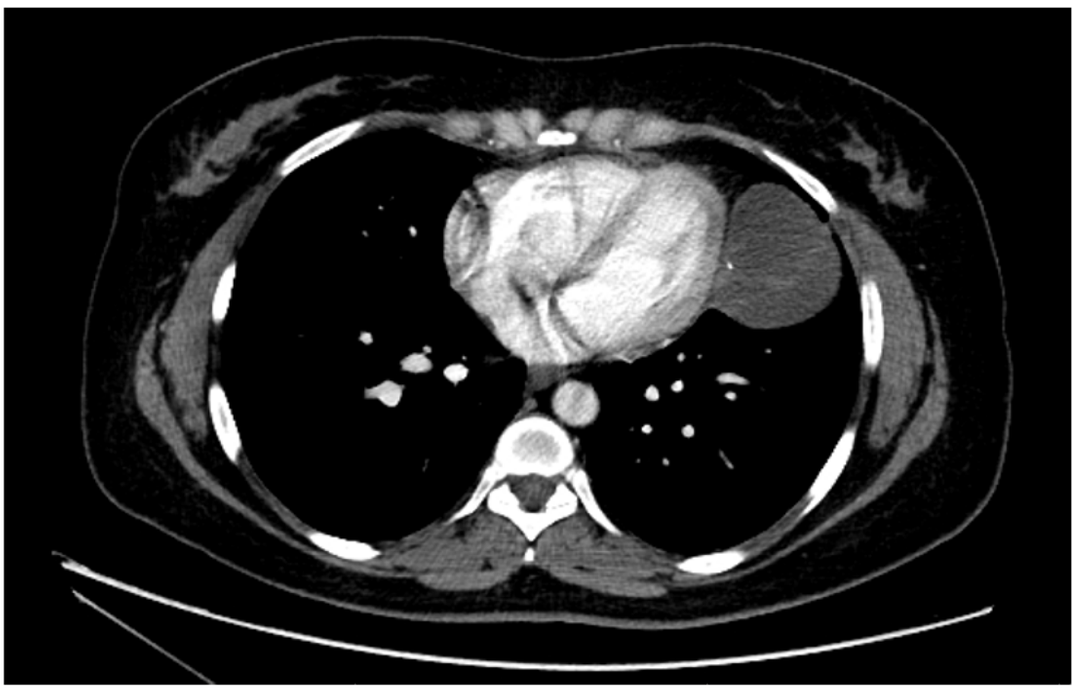

Figure 1: *Axial intravenous contrast enhanced CT scan of the chest demonstrates left paracardial cyst with enhancing septation and tiny calcification originates from the left phrenic nerve.*

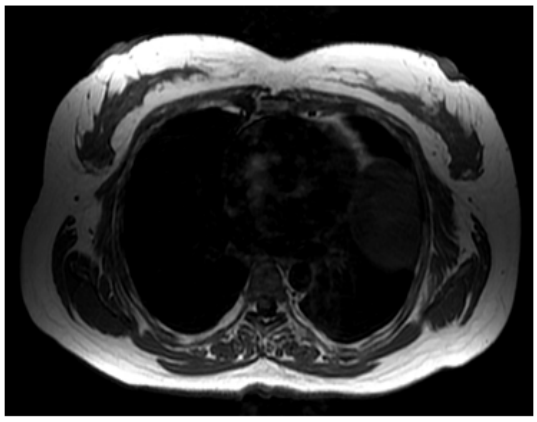

Figure 2a

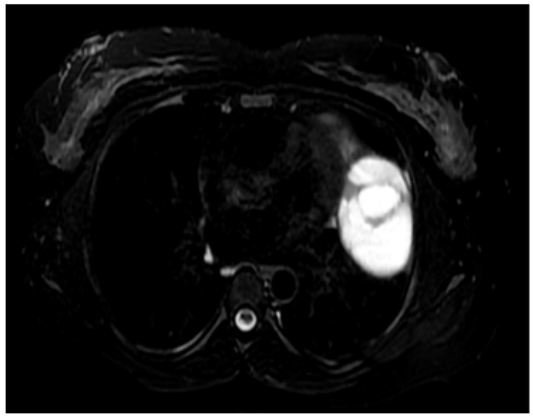

Figure 2b

Figure 2a and 2b: *Left paracardial cyst shows low signal intensity on T1 weighted images (2a) and high signal intensity on T2 fat saturated weighted images with internal low signal intensity septations (2b).*

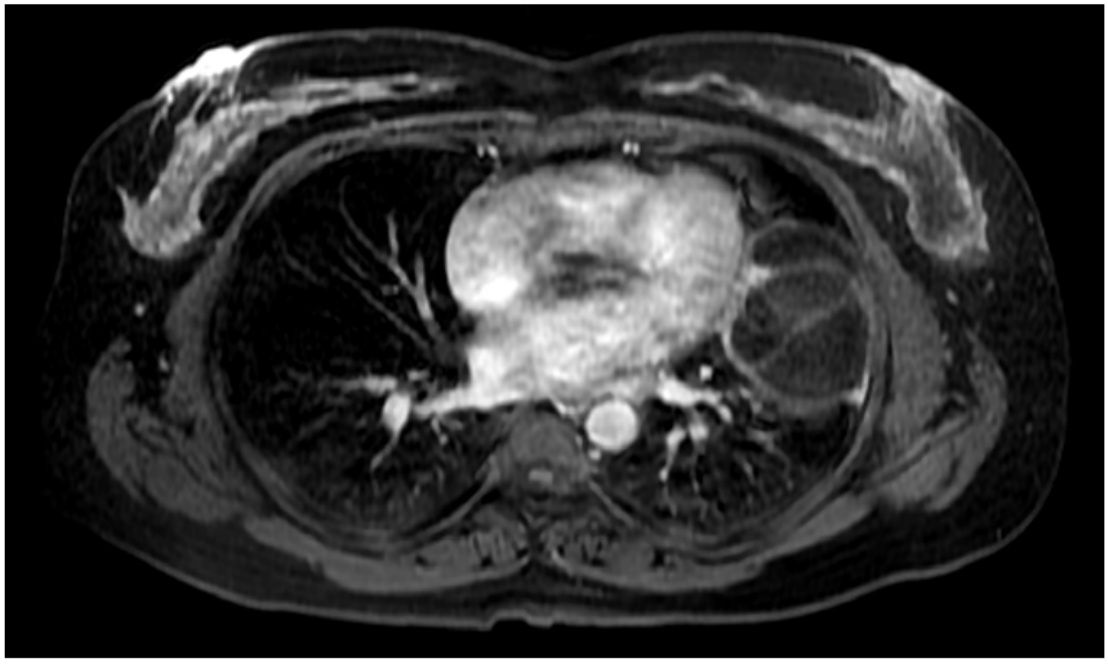

Figure 3: *T1 weighted image post Gadolinium administration demonstrates left paracardial cyst show low signal intensity with enhancing septations.*

Since the patient was symptomatic, complete surgical excision was planned and favored over non-operative management. Additional benefits of complete surgical removal in this case is the decrease in the rate of recurrence, exclusion of malignancy and other complications that could worsen the prognosis if happened. Because the position of the mass was anterior in the left paracardiac location the cyst was easily accessible using the minimally invasive Video-Assisted Thoracoscopic Surgery (VATS) approach and considered safe in this patient since the cyst was away from the major vessels, esophagus and tracheobronchial tree. Additionally, VATS has the advantage of being minimally invasive with better cosmetic and shorter hospital stay. On the other hand, our thoracic surgeon had a good experience on VATS for mediastinal cysts. The tumor was easily resected, and the surgery was tolerated very well. The patient was discharged three days after the surgery. On follow up, the patient had marked improvement of her dyspnea and dry cough. The post-operative X-ray, however, revealed mild left diaphragmatic elevation which was not present in the pre-operative radiological studies. This was likely related to diaphragmatic paralysis due to left phrenic injury during the resection of the cystic mass.

Histological examination revealed that the tumor was made up of two tissue types: Antoni A and Antoni B. Sectioning showed multiloculated cystic lesion containing brown, serous fluid, gelatinous tan material and blood clots. Maximum wall thickness is 0.4 cm. There are yellow to orange, soft-to-firm solid areas within the wall, with a maximum thickness of 1.5 cm. The tumor demonstrated extensive degenerative changes in the form of cyst formation, hyalinization, sheets of histiocytic infiltration with no atypia. Tumor cells composed of bland spindle cells with verocay body formation. No atypia, mitosis or necrosis was seen. Immunohistochemistry for S100 and for EMA (Epithelial Membrane Antigen) were positive. The tumor cells are negative for Pankeratin, desmin, HMB45 and CD117.

Based on the histopathologic findings, a diagnosis of cystic schwannoma was made with degenerative changes (ancient). Postoperative chest X-ray and follow up CT scan showed complete resection of the tumor with mild eventration of left diaphragm (Figures 4a and 4b) as the only complication. She was discharged at the 3rd postoperative day. On follow up the patient had mild dyspnea. The follow up X rays confirmed the elevation of left hemidiaphragm which was related to phrenic nerve injury during the resection of mediastinal cyst. Plication of left diaphragm was offered to the patient but she refused the procedure, since her mild dyspnea was well tolerated and didn't interfere with her daily activities.

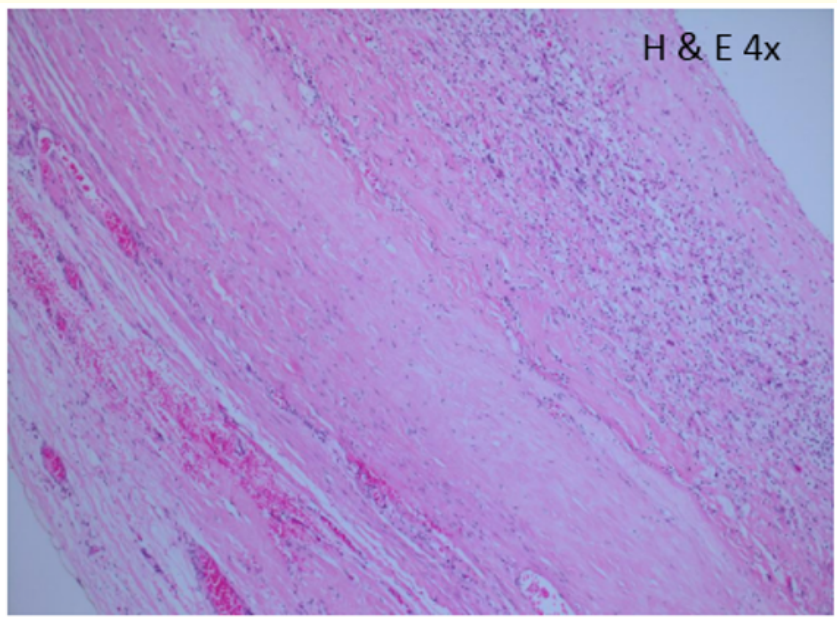

Figure 4a

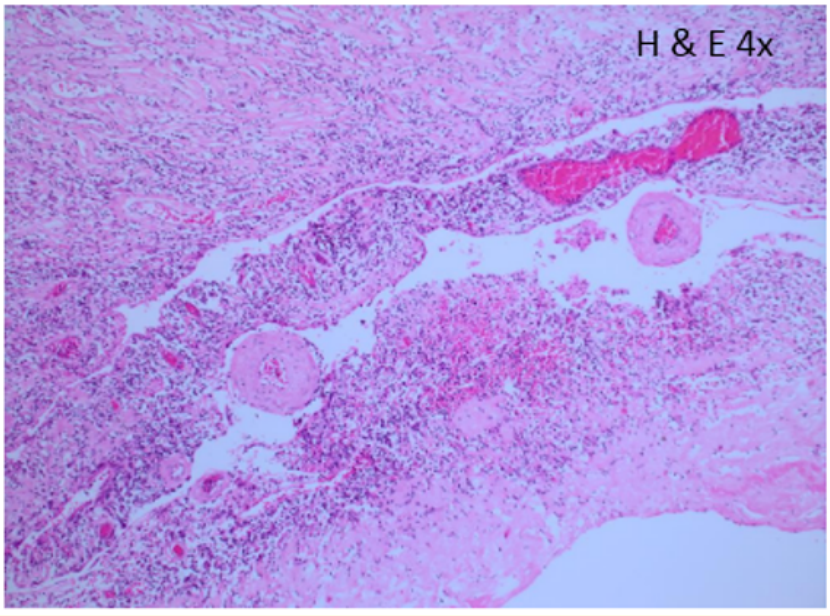

Figure 4b

Figure 4a and 4b: *Fibro-vascularized cyst wall with histiocytic infiltration.*

Discussion

Approximately 20% of mediastinal-based tumors are neurogenic in etiology. Most of these lesions arise in the posterior mediastinum [1,10,11]. Neurogenic tumors are broadly divided into two categories based on their origin: (A) Those that arise from the nerve sheath and (B) those that arise from the nerve cells. Most adult nerve sheath tumors are benign and arise from an intercostal nerve, sympathetic nerves or parasympathetic chains, spinal ganglions, rarely arising from the pericardium, phrenic nerve or vagus nerve. These tumors very rarely arise from nerve sheaths located intrabronchial or intra-parenchymal [12]. There is a single reported case of schwannoma originating from the pulmonary artery [11]. Intrathoracic schwannoma arising from the phrenic nerve is extremely rare, with only 10 cases reported in the literature. 10% of schwannomas contain calcifications [14,18]. Forty-five percent of schwannomas occur in the head and neck, and 9% occurring in the mediastinum. Mediastinal cystic lesions represent 15% - 20% of all mediastinal masses [16].

The differential diagnoses of cystic mediastinal lesions include a foregut cyst (bronchogenic, esophageal duplication, neuroenteric), pericardial cyst, thymic cyst, and lymphovascular malformations. Additional mediastinal lesions that may have cystic appearance includes cystic schwannoma, hydatid cyst, pancreatic pseudocyst, mature teratoma, abscess, and low attenuation lymph node. In our case there was no history of primary malignancy elsewhere, nor she had a prior history of pancreatitis nor exposure to a source of parasitic infection. Furthermore, the characteristic findings of echinococcal cysts (e.g. floating membranes, daughter cysts, and vesicles) were not present in our case. Pericardial cyst and pericardial diverticulum were excluded in view of the presence of septations, septal enhancement and septal calcifications. Cystic teratoma was felt to be less likely given the age of the patient, despite the fact that multiple components including calcium and fluid are seen within the lesion in cross sectional images. Duplication cysts are usually located adjacent to or within the esophageal wall in the lower posterior mediastinum and neuroenteric cysts are rare posterior mediastinal lesions that may be connected to the meninges through a midline defect in one or more vertebral bodies [17]. The imaging features against a bronchogenic cyst are the

unusual location, the presence of septal calcifications and septal enhancement. The calcifications in bronchogenic cyst are usually peripheral or milk of calcium but not septal [18]. Thick or irregular wall is not a typical feature of bronchogenic cyst and suggests superadded infection, hemorrhage or a necrotic neoplasm.

Imaging studies play an important role in diagnosis and localization of mediastinal lesions. In chest X-rays these lesions manifest usually as mediastinal lesions or mediastinal contour abnormality. Computed tomography (CT) can greatly assists in determining the exact location of the mediastinal tumor, its components and its relationship to adjacent structures. Magnetic resonance imaging (MRI) provides superior soft tissue characterization and can help better delineate the relationship of an identified mediastinal mass to nearby intrathoracic vascular structures [15].

Understanding the anatomy of intrathoracic nerves is important for correct interpretation of mediastinal lesions. The phrenic nerves lie along the lateral aspect of mediastinum and run from the thoracic inlet to the diaphragm. They course through the upper chest, medial to the mediastinal pleura, medial to the apex of the right or left lung. The right phrenic nerve lies lateral to the right brachiocephalic vein and the superior vena cava. The left phrenic nerve courses along the lateral aspect of the aortic arch. Then the phrenic nerves pass anterior to their respective pulmonary hila and then inferiorly in a vertical plane along the margin of the heart between the fibrous pericardium and the mediastinal pleura. The left phrenic nerve passes over the pericardium adjacent to the left ventricle and pierces the diaphragm separately. The right phrenic nerve passes adjacent to the right atrium and then leaves the thorax by passing through the vena cava hiatus. The phrenic nerves can occasionally be visualized at cross-sectional imaging. In our case the tumor arises from a branch of the left phrenic nerve adjacent to pericardium of left ventricle. Although no specific imaging features of phrenic nerve schwannoma is described in cross sectional imaging, but mediastinal neurogenic tumors should be considered in the differential diagnosis of rare mediastinal lesions following the course of mediastinal nerves which could be helpful in pre-surgical planning for preservation and repair of the nerve [19].

Conclusion

Cross sectional imaging plays an important role in the diagnosis of cystic mediastinal lesions. Cystic schwannoma of the intrathoracic phrenic nerve, although very rare, might be considered in the differential diagnosis of cystic mediastinal lesions if the lesion is following the course of phrenic nerve.

Conflict of Interest

The authors have no conflicts of interest to declare.

Bibliography

1. Strollo DC., *et al*. "Primary Mediastinal Tumors: Part ll. Tumors of the Middle and Posterior Mediastinum". *Chest* 112.5 (1997): 1344-1357.

2. Wang Jingwei., *et al*. "Giant neurogenic tumors of mediastinum: report of two cases and literature review". *Chinese Journal of Cancer Research = Chung-kuo Yen Cheng Yen Chiu* 25.2 (2013): 259-262.

3. Moinuddeen K., *et al*. "Diaphragmatic eventration: an uncommon presentation of a phrenic nerve schwannoma". *Chest* 119.5 (2001): 1615-1616.

4. Cavanaugh D., *et al*. "Benign schwannoma of the phrenic nerve: case report". *Military Medicine* 144.6 (1979): 406.

5. D'souza B., *et al*. "A rare case of Schwannoma of the intrathoracic phrenic nerve". *ANZ Journal of Surgery* 80.11 (2010): 841-842.

6. Gilani S., *et al*. "A rare presentation of phrenic nerve schwannoma". *European Annals of Otorhinolaryngology, Head and Neck Diseases* 129.6 (2012): 331-333.

7. Orki A., *et al*. "A schwannoma in the middle mediastinum originated from the phrenic nerve". *Indian Journal of Surgery* 74.2 (2012): 199-200.

8. McClenathan JH., *et al*. "Peripheral tumors of the intercostal nerves". *The Annals of Thoracic Surgery* 78.2 (2004): 713-714.

9. Ortigara L., *et al*. "Resection of a mediastinal schwannoma using video-assisted thoracoscopy". *Jornal Brasileiro de Pneumologia* 32.2 (2006): 172-175.

10. Davis Jr RD., *et al*. "Primary cysts and neoplasms of the mediastinum: recent changes in clinical presentation, methods of diagnosis, management, and results". *The Annals of Thoracic Surgery* 44.3 (1987): 229-237.

11. Nakazono T., *et al*. "MRI findings of mediastinal neurogenic tumors". *American Journal of Roentgenology* 197.4 (2011): W643-W52.

12. Feldhaus RJ., *et al*. "A rare endobronchial neurilemmoma (Schwannoma)". *Chest* 95.2 (1989): 461-462.

13. Elstner K., *et al*. "Schwannoma of the pulmonary artery". *Heart, Lung and Circulation* 22.3 (2013): 231-233.

14. Kapoor A., *et al*. "Mediastinal schwannoma: A clinical, pathologic, and imaging review". *South Asian Journal of Cancer* 4.2 (2015): 104-105.

15. Laurent F., *et al*. "Mediastinal masses: diagnostic approach". *European Radiology* 8.7 (1998): 1148-1159.

16. Jeung M-Y., *et al*. "Imaging of cystic masses of the mediastinum". *Radiographics* 22.1 (2002): S79-S93.

17. Ödev K., *et al*. "Imaging of cystic and cyst-like lesions of the mediastinum with pathologic correlation". *Journal of Clinical Imaging Science* 2 (2012): 33.

18. McAdams HP., *et al*. "Bronchogenic cyst: imaging features with clinical and histopathologic correlation". *Radiology* 217.2 (2000): 441-446.

19. Takeda S-i., *et al*. "Intrathoracic neurogenic tumors-50 years' experience in a Japanese institution". *European Journal of Cardio-Thoracic Surgery* 26.4 (2004): 807-812.

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