

Varied foci of primary benign schwannoma of the thorax: a case series

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Abstract

Schwannomas are the most common tumor of peripheral nerves. It comprises 1 to 2% of total thoracic tumor. Posterior mediastinum is the most common site in thorax. Hereby we are reporting two cases of schwannoma with different sites of origin. The classical presentation of schwannoma is an asymptomatic mass found on chest radiograph. This tumor is usually benign and slow growing. Imaging followed by histopathological examination is key to the diagnosis of this neoplasm. Resection of tumor cure the disease.

Introduction

Schwannomas are the most common tumor of peripheral nerves. It is the most common benign mediastinal neurogenic tumor originating from neural sheath Schwann cell. This tumor commonly occurs on the flexor surfaces of the extremities, neck, mediastinum, retroperitoneum, posterior spinal roots, and cerebellopontine angle [1]. They are generally benign, asymptomatic, slow-growing lesions. Very rarely pleuro-pulmonary schwannomas can be a manifestation of the metastasis from peripheral sites. This tumor comprises of 1 to 2% of total thoracic tumors [2]. Here we are reporting two cases of schwannoma with varied sites of origin.

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Case #1

A nineteen-year male presented to Pulmonary Medicine Outpatient Department (OPD) with chief complaints of right sided chest pain for 1 month. His past history is unremarkable. He was a non-smoker and a medical student. There was no family history of neurofibroma or ash leaf macule or neurofibroma were not found on general survey. At the time of presentation his vitals were stable. There were no palpable lymph nodes. Complete blood count and metabolic panel, urinalysis, and electrocardiography were normal. The tuberculin skin test was negative. Chest x-ray was showing mass lesion in right mid and lower zone. Contrast enhanced computed tomography (CECT) thorax was showing non enhancing heterogeneous soft tissue attenuating pleural based mass lesion of size 11×10.9×8.3 cm along right lower lobe. Medially lesion abutting pericardium, right atrium, and IVC. Laterally abutting lateral chest wall. Differential diagnosis of the pleural based mass considered were solitary fibrous tumor of pleura, mesothelioma and metastatic adenocarcinoma. Ultra sonographic guided biopsy was taken it was showing multiple fragmented core of tissue comprised of lesion with in cell arranged in hypo and hypercellular area (Antony A and Antony B area). With that diagnosis of pulmonary schwannoma was made after multidisciplinary discussion in tumor board clinic. Patient was sent to cardiothoracic vascular surgery (CTVS) department for operative management. Open thoracotomy and resection of tumor was done. The patient had an uneventful postoperative recovery with no adjuvant therapy. Follow up Chest X ray after 6 months showed no residual tumor (Figures 1 to 3).

Case # 2

A forty-two-year male presented to pulmonary medicine OPD with chief complaints of left sided chest pain for 2 months. There was no history of trauma, no associated comorbidities. He was a smoker with 20 pack years and farmer by occupation. His vitals were stable at the time of presentation. Chest x ray was taken showing pleural based mass lesion in right upper and midzone with erosion of left second rib. On CECT thorax, well-defined irregular heterogeneously enhancing soft tissue attenuating lesion with few non enhancing areas noted involving left chest wall measuring 7.7x6.2x6.5cm with destruction and expansion of left second rib and pressure erosion of anterior aspect of left first rib. Complete blood count and metabolic panel, urinalysis, and electrocardiography were normal. Differential diagnosis considered in this case were solitary fibrous tumor of pleura, Ewing's sarcoma, localized malignant mesothelioma and metastatic adenocarcinoma. CT guided biopsy was taken which showed predominantly

hyper cellular and a few hypocellular areas. The hypercellular areas show tumor cells which are spindled, palisading with moderate cytoplasm, hyperchromatic nucleus with infrequent mitosis. Immunohistochemistry revealed diffuse positivity for S-100 and negative for CD34 and p53 suggestive of cellular schwannoma. The case was discussed in multidisciplinary tumor board clinic. Patient was referred to CTVS for surgical management and wide local excision of tumor was done along with ribs. The patient had an uneventful postoperative recovery. Follow-up x-ray showed no tumor with segmental collapse of left upper lobe (Figures 4 to 7).



Figure 1. Chest X-ray showing well circumscribed mass lesion in right lower zone.

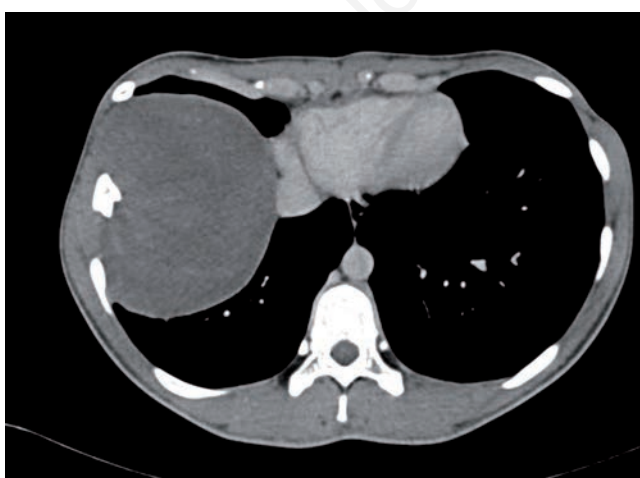


Figure 2. CECT thorax was showing non enhancing heterogeneous soft tissue attenuating pleural based mass lesion of size 11*10.9*8.3 cm along right lower lobe.

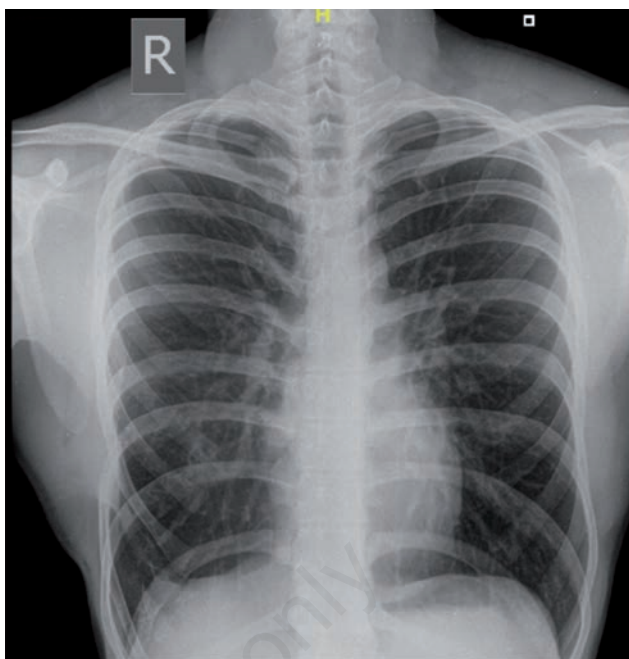


Figure 3. Chest X-ray after excision of the tumor from right lower lobe after 6 months.

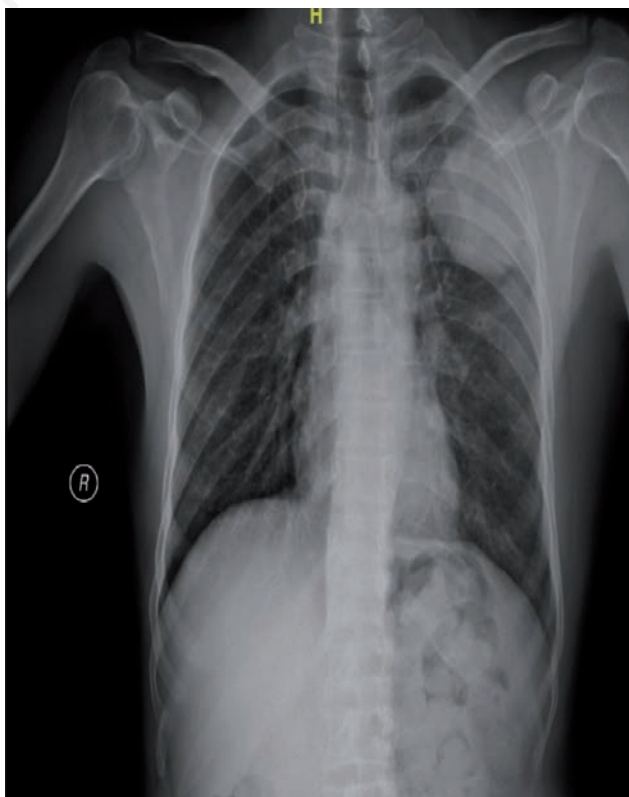


Figure 4. Chest X-ray showing pleural based mass lesion in left upper and mid zone with erosion of left second rib.

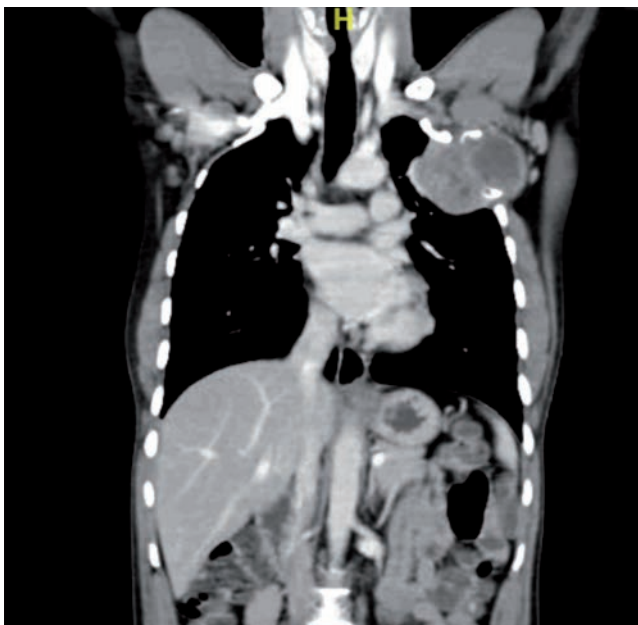


Figure 5. CECT thorax showing well-defined irregular heterogeneously enhancing soft tissue attenuating lesion with few non enhancing areas noted involving left chest wall measuring 7.7x6.2x6.5 cm with destruction and expansion of left second rib.

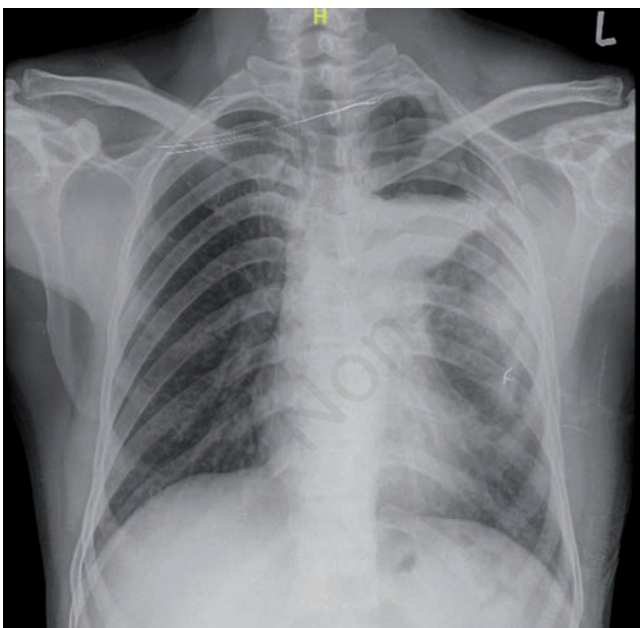


Figure 6. Chest X-ray after excision of tumor.

Discussion

Neurilemmoma, also termed schwannoma, presents as a well circumscribed and encapsulated mass in the human body and is almost always solitary [3]. Schwannoma is a tumor which arises from the specialized nerve cells which produce myelin called

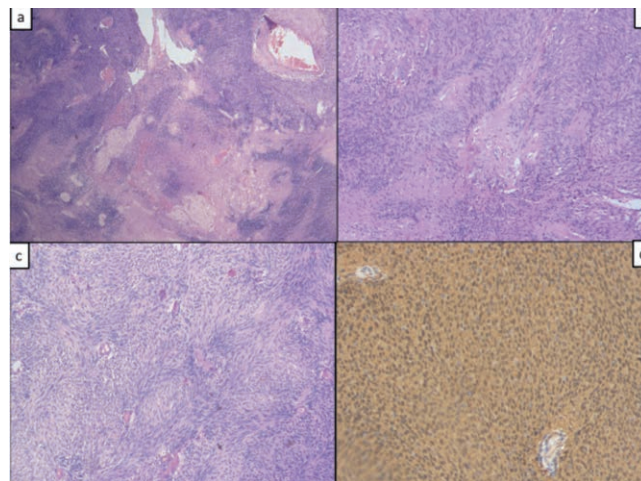


Figure 7. Photomicrographs of histopathological sections. a) Tumor composed of hypercellular and hypocellular areas along with hyalinized blood vessels (H&E, 40x). b) Sheets of tumor cells with peripheral palisading (H&E, 100x). c) Cells are in spindle shaped with mild nuclear atypia (H&E, 200x). d) Immunohistochemistry with S100 shows diffuse strong nuclear positivity (DAB, 200x).

Schwann cells. This kind of tumor is a relatively common mediastinal neurogenic tumor, accounting for about 25.3% of intrathoracic neurogenic tumors [4]. Posterior mediastinum is the most common site of schwannoma in thorax [5]. 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.

According to Otshuka *et al.*, schwannoma affects most commonly males [5]. It is predominant in the third or fourth decades of life [6]. This is a report of 2 male cases with schwannoma and they presented at different ages, one at 2nd decade and other at 4th decade of life. They also stated that 52% of intra-bronchial and pulmonary schwannoma are symptomatic. [5] On the other hand, most peripheral intrapulmonary schwannoma is asymptomatic [5]. The classical presentation is an asymptomatic mass found on chest radiograph. The major complaints of the patient as described in case series included slight hemoptysis, cough, fever, chest pain and shortness of breath [7]. Both of our patients presented with chest pain.

On radiographic images, peripheral pleural schwannoma appears as a round mass with well-defined margins with minimal contrast enhancement [8]. Malignant pleural schwannomas have been reported in literature but rare [9]. Malignant lesions have been described in patients with neurofibromatosis type 1 and patients with positive history of previous radiation therapy. Malignant lesion can present as pulmonary pleural nodule, pleural effusion, and mass lesion [10]. Benign schwannomas may be large and demonstrate intense heterogeneous FDG activity, mimicking malignancy on 18 FDG PET/CT. Benign and malignant peripheral nerve sheath tumor has similar FDG activity [11]. MRI is also one of the imaging modalities which is used to diagnose schwannoma in which T1 weighted images were isointense with skeletal muscle and T2 images were hyperintense. However, MRI is also not able to differentiate between benign and malignant lesions and this pattern of signal intensity was not specific for neural tumors [12]. So PET CT and MRI were not frequently advised for pulmonary schwannoma.

Histopathology is essential for diagnosis of benign schwannoma. In histopathology Antoni A and Antoni B areas are revealed in majority of cases. Antoni A represents areas of hypercellularity with Verocay bodies. Antoni B areas of myxoid hypocellularity exhibit degenerating changes (*i.e.*, cyst formation, hemorrhage, calcification, xanthomatous infiltration, and hyalinization). Resection of the tumor is the definitive treatment, and it may be ranging from wedge or sleeve resection, lobectomy or pneumonectomy. This benign lesion rarely recurs after resection [13]. Metastasis to lung from the peripheral tumor has been reported but it's extremely rare [14]. Signs of malignant invasion are absence of capsule, perineural invasion, increased mitotic figures, ill-defined cell borders, areas of hemorrhagic and cystic degeneration [15].

Conclusions

Schwannoma is an extremely rare neoplasm of thorax. Most of the patient will be asymptomatic. Imaging followed by histopathological examination is key to the diagnosis of this neoplasm. PET-CT rarely help to differentiate between malignant and benign tumors. This benign tumor has good prognosis after resection. Hence, schwannoma should be kept as a differential diagnosis when we are dealing with thoracic tumors.

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