An Unusual Schwannoma

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Abstract
Mediastinal masses are commonly encountered and have multiple differentials. Although histopathological examination is gold standard, the location of the mass narrows the diagnosis. While thyroid, thymus, germ cell tumour or lymph node related masses are common in superior mediastinum, vascular or pleuro-pericardial masses are seen in middle mediastinum. Posterior mediastinal masses are commonly neurogenic tumours, schwannoma being the commonest. We discuss a case of cystic schwannoma presenting as superior mediastinal mass.

Introduction
Median masses are commonly encountered with a spectrum of clinical presentation and varied radiology. Though the site of lesion can narrow the differential diagnosis, a definitive diagnosis is possible only on histopathological examination of the biopsy sample. Superior mediastinum is a common site for thyroid, thymus, germ cell tumour or lymph node related neoplasms. We report a case of neurogenic tumour presenting as the superior mediastinal mass.

Case Report
A 37 year old male non-smoker was referred to our outpatient department in view of intermittent chest pain and exertional breathlessness since 3 years. He had no medical or surgical illness in the past. General and systemic examination was unremarkable.

Investigations
Biochemical and haematological investigation were within normal limit. Chest X-ray (CXR) was suggestive of right suprahilar paratracheal opacity causing mediastinal widening (Fig. 1). On lateral CXR, the lesion was localised to the antero-superior mediastinum. A contrast enhanced computerised tomography (CT) of the thorax was suggestive of 4.8 cm x 4 cm non enhancing soft tissue density lesion in the superior mediastinum right paratracheal in location with uniform water attenuation. The lesion was abutting the trachea, prevertebral fascia and superior vena cava. The lesion extended from T2 to T4 vertebra (Figures 2a, 2b). Computerised tomography assisted fine needle aspiration of the mass lesion yielded clear fluid wherein cytological examination was inconclusive. Hence, surgical excision of the lesion was done through a right thoracotomy approach and the excised specimen was subjected to histopathology. The surgical excision specimen was an encapsulated sharply demarcated well defined lesion on gross examination. It had a yellow cut surface with areas of dark red/black discolouration due to haemorrhage and cystic degeneration (Figure 4a). The microscopic examination revealed the presence of spindle cells with bland looking elongated nuclei in the cyst wall with multiple verocay bodies (Figure 4b). The final histopathological diagnosis was cystic schwannoma. Post surgery, the patient recovered uneventfully.

Clinical Diagnosis and Discussion
Right superior mediastinal mass lesion.

The mediastinum is subdivided into various sections as per the anatomist (Figure 3).1,3 The differential diagnosis of superior mediastinal masses is vast and can be remembered by the mnemonic, 5 Ts, i.e. 1) Thymus related (thymoma, invasive thymoma,
thymic carcinoma, thymolipoma/thymoliposarcoma, thymic cyst, benign thymic hyperplasia, thymic carcinoid); 2) Thyroid and parathyroid related (neoplasms and goitre); 3) Terrible Lymphoma (Hodgkin lymphoma /non-Hodgkin lymphoma); 4) Teratomas (Germ cell tumours, mediastinal teratoma, teratocarcinoma, mediastinal seminoma, mediastinal embryonal cell carcinoma, mediastinal yolk sac tumour, mediastinal choriocarcinoma, mediastinal mixed cell type germ cell tumour); 5) Thoracic aortic aneurysm.4 The diagnosis in an individual case can be aided by thorough clinical examination and judicious use of imaging and ancillary investigations.5

Diagnosis in our case was revised to cystic schwannoma as per histopathological correlation. Schwannoma is a benign slow growing peripheral nerve sheath tumour;6,7 earlier known as neurilemmomas and neurinomas of Verocay. They are encountered in the region of head, neck, flexor aspect of upper and lower extremities, retroperitoneum, posterior spinal roots and cerebellopontine angle. Forty-five percent of schwannomas occur in the head and neck, with 9% occurring in the mediastinum. Schwannomas are the most common mediastinal neurogenic tumours, which generally involve the posterior mediastinum. They are well encapsulated and well-marginated masses found in the costovertebral sulci.8 Rarely they occur in the middle mediastinal compartment arising from the vagus or phrenic nerves. In about 10% of cases mediastinal schwannomas may extend to the spinal canal (dumbbell tumours); occasionally they may also extend to the cervical region or, even more rarely, may be associated with other synchronous mediastinal lesions with a different histology. Men and women are
equally affected in their third and fourth decades. When they occur in patients with neurofibromatosis, schwannomas usually present by the 3rd decade. Usually, they are asymptomatic and benign, and very rarely malignant or multiple. Radiologically they are sharply demarcated with rare calcifications. Contrast enhanced CT scan of the thorax shows in accordance, a sharply demarcated mass with low densities and mild enrichment, rarely with calcifications and no fat. On MRI the schwannomas have low - to intermediate signal intensity on T1-weighted images and may have intermediate - to high - signal intensity on T2-weighted sequences.

Mediastinal schwannomas are indistinguishable from congenital or acquired cysts, if cystic degeneration is extensive even with CT and MRI imaging and diagnosis is established post surgery in such cases. Histopathologically, schwannomas are derived from the myelinating cell of the peripheral nervous system and are composed almost entirely of Schwann cells. Schwannomas typically grow within a capsule and remain peripherally attached to the parent nerve. Antoni A and B tissue types represent distinct histologic architectural patterns that aid in the diagnosis of schwannomas. Type A tissue is highly cellular and demonstrates nuclear palisading and associated Verocay bodies, which reflects their prominent extracellular matrix and secretion of laminin. Type B tissue is loosely organized with myxomatous and cystic changes and may represent degenerated Antoni A tissue. The Verocay bodies can also be seen in some neoplasms in the skin.

In cases of benign neoplasms, complete excision of the lesion itself is generally sufficient. Video-assisted thoracoscopic surgical resection is now commonplace for these benign tumours. Shorter hospital stay and more rapid return to work have been demonstrated with this method.

**Conclusion**

In our case the schwannoma presented as a superior mediastinal mass mostly having arisen from the phrenic or vagus nerve which is a rare location with cystic degeneration. Surgical excision and pathological examination led to the diagnosis.

**References**


