Case 1601

Mediastinal schwannoma
Published on 31.07.2002

DOI: 10.1594/EURORAD/CASE.1601
ISSN: 1563-4086
Section: Paediatric radiology
Imaging Technique: CT
Case Type: Clinical Cases
Authors: J.K. Mordani, M.A. Siddiqui, D.B. Bakalinova
Patient: 13 years, female

Clinical History:
The patient presented with a history of cough and shortness of breath. A chest X-ray revealed a mediastinal mass.

Imaging Findings:
The patient presented with a history of cough and shortness of breath. A chest radiograph (PA and lateral) was performed and showed a smooth rounded mass in the middle/posterior mediastinum on the right side, causing deviation of the trachea to the left. No obvious calcification or bony change was noted. On contrast-enhanced CT a well-marginated, rounded mass was seen in the posterior mediastinum measuring 5cm x 5cm and extending from root of neck to 3rd thoracic vertebra lying anterolateral to the vertebral bodies on the right side causing shift of the trachea to the left. The mass was of soft tissue attenuation with some areas of low attenuation in the centre consistent with necrosis or cystic change. There was some peripheral enhancement. No extension into the spinal canal, or bony changes were noted.
A differential diagnosis of a neurogenic tumour was made with the possibility of a ganglioneuroma. The tumour was completely excised surgically and a histological diagnosis of schwannoma was made.

Discussion:
Neurogenic tumours comprise 9% of mediastinal masses in adults and 29% in children. A high proportion of posterior mediastinal masses in children are neurogenic. They can arise from peripheral nerve and nerve sheath (schwanomma, neurofibroma, malignant tumour of nerve sheath origin (MTNSO)), sympathetic ganglia (ganglioneuroma, ganglioneuroblastoma and neuroblastoma) or from paraganglia (chemodectoma, phaeochromocytoma). 85% of tumours in children are of ganglionic origin, while 75% of adult tumours are of peripheral nerve and nerve sheath origin. Paraganglia tumours are rare (4%).
Pathologically, schwannoma are either composed of densely packed spindle cells (Antoni A) or loosely organised in myxoid stroma (Antoni B). They arise from Schwann cells and extrinsically compress the nerve. They are encapsulated and can be heterogeneous, especially when large, because of areas of cystic degeneration, low cellularity, haemorrhage, and lipid laden myelin or punctuate calcification. Neurofibromas on the other hand are usually homogenous, well-marginated but non-encapsulated tumours that arise from the entire peripheral nerve component (Schwann cells, myelinated and unmyelinated nerve fibres and fibroblasts). Despite these differences, they both manifest grossly as lobulated spherical masses. Plexiform neurofibroma is a well-defined, non-encapsulated variant which infiltrates along the whole nerve trunk or plexus. This is considered pathognomonic of von Recklinghausen's disease.
On plain radiographs both schwannoma and neurofibroma are usually sharply margined, spherical and lobulated paraspinal masses that span one or two posterior rib interspaces, but can attain larger sizes. In 50% of cases they produce benign pressure erosion or deformity of ribs, vertebral bodies and neural foramina. Calcification is rarely detectable.

On CT they are well-circumscribed, homogenous or heterogeneous masses. Punctate calcification is occasionally detected and low attenuation areas corresponding to hypocellularity, cystic degeneration, haemorrhage and myelin are detected. They may demonstrate mild homogenous, heterogeneous or peripheral contrast enhancement. 10% grow through intervertebral foramina into the spinal canal and have a dumbbell or hourglass appearance.

On MRI they have slightly greater signal intensity than muscle on T1-weighted images and markedly increased signal intensity on T2-weighted images, often in an inhomogeneous fashion. Schwannomas may have a high intensity centre and relatively low intensity outer wall correlating with cystic change on T2-weighted images. Neurofibromas may have a central region of lower intensity than the periphery because of tumour in the centre and peripheral myxoid degeneration.

Sympathetic ganglia tumours originate from the nerve cell rather than the nerve sheath. Ganglioneuroma and ganglioneuroblastoma are most common in the posterior mediastinum. 50% of neuroblastoma arise from the adrenal gland and 30% in the mediastinum, the most common extra-abdominal location. Pathologically ganglioneuroma are composed of single or clustered mature ganglion cells in a dense stroma and manifest grossly as encapsulated, oblongated, homogenous tumours, occasionally having a dumbbell component. They typically occur in children older than 3 years, adolescents and young adults. Radiologically they are well-marginated, oblong masses with a broad base, spanning 3-5 vertebrae. CT shows a homogenous or heterogeneous elongated mass, while MRI shows homogenous intermediate intensity on T1-weighted images with occasional whorled appearance and heterogeneous high signal intensity on T2-weighted images.

Ganglioneuroblastomas have composite features of ganglioneuroma and neuroblastoma. Radiologically they might be sharply demarcated or irregular, locally invasive or widely metastatic.

Neuroblastomas affect children of less than 5 years of age and they are composed of small round cells arranged in sheets or pseudorosettes. They are non-encapsulated and contain extensive areas of necrosis, haemorrhage and cystic degeneration. Two thirds of children affected have distant metastasis at presentation. They can be metabolically active and secrete catecholamine and vasoactive intestinal peptides. Plain film shows calcification in 30% of cases. On CT, the tumour may be smooth or irregular, heterogeneous, and 80% show calcification. MR shows homogenous or heterogeneous signal intensity on all sequences and post-contrast enhancement.

**Differential Diagnosis List:** Mediastinal schwannoma

**Final Diagnosis:** Mediastinal schwannoma

**References:**

Khanlou H, Khanlou N, Eiger G.
Schwannoma of posterior mediastinum: a case report and concise review.

Strollo DC, Rosado-de-Christenson ML, Jett JR.
Primary mediastinal tumors: part II. Tumors of the middle and posterior mediastinum.
Chest 1997 Nov 5;112(5):1344-57. (PMID: 9367479)

Marchevsky AM.
Mediastinal tumors of peripheral nervous system origin.
Semin Diagn Pathol 1999 Feb;16(1):65-78. (PMID: 10355655)
Armstrong P, Wilson AG, Dee P, Hansell DM.
Imaging of diseases of the chest, 3rd edition.
Mosby, St Louis (2000).
Naidich DP, Muller NL, Zerhouni EA.
Computed tomography and magnetic resonance of the thorax, 3rd edition.
Lippincott Williams & Wilkins, Philadelphia (1998).
Description: PA radiograph showing a well-defined spherical mass located in the posterior mediastinum causing deviation of the trachea to the left. Origin:
Description: Lateral radiograph showing a well-defined spherical mass located in the posterior mediastinum. Origin:
Figure 2

Description: Images 2a-2g: Contrast-enhanced CT showing a well-margined, rounded mass in the posterior mediastinum measuring 5cm x 5cm and extending from root of neck to 3rd thoracic vertebra, lying anterolateral to the vertebral bodies on the right side and causing a shift of the trachea to the left. It is of soft tissue attenuation with some areas of low attenuation in the centre consistent with necrosis or cystic change. There is some peripheral enhancement. No extension into the spinal canal or bony changes are seen. Origin: