A Case of Giant Mediastinal Neurogenic Tumor in a Child-Unusual Presentation

Khadka H*, Thapa B**
*Associate professor, Department of Radiology and Imaging, NAMS, **Associate professor, Department of Paediatric surgery, Kant Children Hospital, Maharajgunj.

ABSTRACT
Neurogenic tumors are commonly found in the posterior mediastinum, they may reach large size before becoming symptomatic. If the neurogenic tumor occupies more than half size of the thorax, it is called giant intrathoracic neurogenic tumors. Giant intrathoracic neurogenic tumors are relatively rare. We report a case of Right sided thoracic mass in a 10 year old child who presented with chest pain and fever for 1 week. On chest x-ray, there was an opacity in Right chest, mimicking pleural effusion. In CT scan of chest, there was a heterogeneously enhancing mass in Right hemithorax. In trucut biopsy, it turned out to be neuroblastoma.

INTRODUCTION
Neurogenic tumors are the most common mediastinal mass accounting for 12-21% of all mediastinal neoplasms and 75-90% of posterior mediastinal neoplasms. Nearly half of neurogenic tumors are asymptomatic, however as they become large, they can produce symptoms related to local compression, bone invasion and spinal cord involvement. Neurogenic tumors develop either from the peripheral nerves, ganglion cells or paraganglion cells. In thorax, they are located in posterior mediastinum. Neurofibromas and schwannoma develop from peripheral nerves and they are common in adults. Neuroblastoma. Ganglioneuroma and ganglioneuroblastoma arise from ganglion cells and are common in children. Pheochromocytoma and chemodectoma arise from paraganglion cells and they are rarest of the neurogenic tumors. Small neurogenic tumor is typically located in posterior mediastinum, therefore it is easy to diagnose. But when the lesion is big, it is very difficult to differentiate it from lung mass. In x-ray it is very likely to be misinterpreted as massive pleural effusion. The same thing happened in this case. This was the main region to publish this case as it was initially thought to be pleural effusion. Even later, cross sectional imaging by CT scan, it was very difficult to differentiate it from malignant lung mass. Only biopsy revealed the true histology of the tumor.

CASE REPORT
10 year old girl brought by her parents to pediatric speciality hospital, with complain of chest pain and fever for 1 week. On clinical examination, temperature was 100°F. On respiratory system examination, chest was dull on percussion on right side with decreased breath sounds. Few crepts were heard on same side. Provisional diagnosis of pneumonia was made and investigations were ordered. Chest x-ray showed a homogenous opacity in Right mid and lower zones with obliteration of right costophrenic angle and right hemidiaphragm (fig 1). Imaging diagnosis of right sided pleural effusion was made with these findings.
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Pleural tap of 10 ml fluid showed exudative nature. After 2 days of IV antibiotics, discharged on oral antibiotics.

But after 1 month she was again brought with complain of persistent right sided chest pain which was more severe than before. Antibiotics continued, with no improvement in her symptoms. Repeat chest x-ray showed increased right sided opacity (fig 2) with upper margin showing convexity upwards. CECT chest showed huge heterogeneously enhancing lobulated soft tissue mass in Right hemithorax with nonenhancing necrotic areas suggesting its malignant nature. The mass had abutted mediastinal structures as well as chest wall, anteriorly, posteriorly and laterally with midline cross (fig 3). Few enhancing peripherally located nodular lesions also seen in bilateral lungs suggestive of metastases.

Ultrasound guided trucut biopsy was done which revealed neuroblastoma.

DISCUSSION

There is no consensus on clinical diagnostic criteria of giant mediastinal neurogenic tumors. Generally, neurogenic tumors occupying half the size of thoracic cavity accompanied by mediastinal shift, tracheal compression or superior venacaval reflux disorder can be called giant neurogenic tumor. Because posterior compartments of mediastinum is rich in spinal nerves, vagus nerves and sympathetic chains, neurogenic tumors are most common in this compartment. Neurogenic tumors develop either from the peripheral nerves, ganglion cells(sympathetic chain ganglia) or paraganglion cells. In thorax, they are located in posterior mediastinum. Neurofibroma and schwannoma arise from peripheral nerves and are commonest in adults. Neurofibromas and schwannoma are both derived from Schwann cells but neurofibromas also have collagen and fibroblasts. Only about 1.6% of NF-1 patients have symptomatic spinal tumors. Patients with NF-2 usually have intraspinal intradural tumors and 30-40% of these patients have neurologic deficits and symptoms. Neuroblastoma, ganglioneuroma and ganglioneuroblastoma arise from ganglion cells, and are commonest in children. Phaeochromocytoma and chemodectoma arise from paraganglion cells, and are rares of the neurogenic tumors. Neurogenic tumors can be benign and malignant (70% vs 30%). Intrathoracic neural tumors are uncommon but account for about 75% of tumors.
of posterior mediastinum. The male to female ratio is 1:1.3. Age range can be from 7 mths to 63 yrs (average 33 yrs). They present Usually as an incidental posterior mediastinal mass in child or young adult, on a chest radiograph. Can have back pain. Dumbell tumor can cause spinal cord compression, usually neurofibromas. They are usually in neurofibromatosis. They are usually Well defined round or oval soft tissue mass in the paravertebral gutter usually projecting to only one side of mediastinum. Nerve sheath tumors are usually circular. Ganglion cell tumors are usually more elongated. Ganglion cell tumors show foci of calcification (spicules or nodules). Calcification is rare in nerve sheath tumors. In one study, by Davidson et al, over 70% of tumors were benign nerve sheath tumors, neurofibromas (45%), neurilemomas (13%) and a mixed pattern with both neurofibromas and neurilemoma elements in them (13%). The remainder were all nerve cell tumors. Benign ganglioneuromas were 24% and neuroblastomas were 5%. Involvement of bones can give splaying of thin posterior ribs, localized pressure erosion defect of vertebral body, enlargement of IV foramen, rib notching and scoliosis. Rapid increase in size in association of bone destruction and pleural effusion - s/o malignancy. In anteriorly situated neural tumors, there is a higher incidence of malignancy but such tumors are very rare.

**Diagnostic modality**

CT scan is useful for obvious mediastinal pathology, widened mediastinum, vascular mediastinal lesion, abnormal hilum, staging of malignancy, calcification, and bone destruction. CT is 100% sensitive in detection and delineation of tumor, 88% sensitive for intraspinal extension and 100% sensitive for chest wall involvement.

MRI is preferred investigation for neurogenic tumors because it allows simultaneous assessment of intraspinal extension, spinal cord abnormalities and longitudinal extent of tumor. MRI is 100% sensitive for lymphnode, intraspinal extension and chest wall involvement.

In summary, MRI is better than CT scan for intraspinal extension, much better for lymphnode detection. CT is superior to MRI for detection of calcification, CT and MRI equally sensitive for chest wall involvement, CT is sufficient to see the recurrence after treatment.

Nerve sheath tumors are usually benign. Schwannoma arise from the nerve sheath and they are encapsulated. They have areas of cystic degeneration and haemorrhage and small focal areas of calcification. They grow laterally along the parent nerve and compress the nerve. CT scan - lower attenuation than skeletal muscle due to cystic degeneration, high lipid content and interstitial fluid. Neurofibromas are homogenous and have higher attenuation because they have fewer of these histologic features. Heterogenous enhancement after IV contrast.

MRI-variable signal intensity but typically have signal intensity similar to spinal cord. T2-high signal intensity peripherally and low signal intensity centrally (target sign). They enhance after gadolinium. Neurofibromas are unencapsulated. They grow by diffusely expanding the parent nerve. Male:female ratio is one. CT/MRI-sharply marginated unilaterial, spherical or lobular posterior mediastinal masses. 50% can cause pressure erosion of adjacent ribs or vertebral body and enlargement of neural foramina. Punctate intrasosseal calcification occasionally present. 10% paravertebral neurofibroma and schwannoma extend into spinal canal and give dumbbell shaped masses with widening of neural foramen.

**Neuroblastoma** is the most common solid extracranial pediatric tumor. It arises from nerve cells of sympathetic ganglia. Adrenal gland is the commonest location. Most common extraabdominal location is posterior mediastinum (15-30%). It is highly malignant. Typically occurs <5 yrs. Posterior mediastinal mass in this age group is neuroblastoma unless proven otherwise. Neuroblastoma occurs at very young age, median age of diagnosing neuroblastoma is 17 months. Although primary intrathoracic neuroblastomas carry a less ominous prognosis than similar intra-abdominal tumors, it has been suggested that wide excision, even if incomplete may lead to increased survival. Neuroblastoma is an embryonal tumor of autonomic nervous system meaning that the cell of origin is thought to be a developing and incompletely committed precursor derived from neural crest tissue.

CT scan-predominantly a paraspinal heterogeneous soft tissue mass with areas of haemorrhage, necrosis and cystic degeneration. In one study, calcification was found to be 79%, characteristic rim calcification in 29%.
MRI-heterogenous signal intensity in all sequences. Heterogenous enhancement after contrast administration. Often widespread local invasion and tendency to cross the midline 49% of neuroblastoma in children were detected as incidental finding in chest roentgenogram done for non-tumor related symptoms-14% presented as acute respiratory symptoms.

Staging:

The “International Neuroblastoma Staging System” (INSS) established in 1986 and revised in 1988 stratifies neuroblastoma according to its anatomical presence at diagnosis.

Stage 1. Localised tumor confined to area of origin.

Stage 2A: Unilateral tumor with incomplete gross resection, identifiable ipsilateral or contralatera lymphnode negative to tumor.

Stage 2B: Unilateral tumor with complete or incomplete gross resection, with ipsilateral lymphnode positive for tumor, identifiable contralateral lymphnode, negative for tumor.

Stage 3: Tumor infiltrating across midline with or without regional lymphnode, or unilateral tumor with contralateral lymphnode, or midline tumor with bilateral lymphnode involvement.

Stage 4: Dissemination of tumor to distant lymphnodes, bone marrow, bone, liver or other organs except as defined by stage 4s.

Stage 4s: Age <1 year old with localized tumor as defined by stage 1 or 2, with dissemination limited to liver, skin or bone marrow (less than 10 percent of nucleated bone marrow cells are tumors).

Screening: Urine catecholamine level can be elevated in pre-clinical neuroblastoma.

Treatment: Aggressive multimodal therapy (intensive chemotherapy, surgery, radiation therapy, stem cell transplant, differentiation agent isotretinoin also called 13-cis-retinoic acid, and frequently immunotherapy with anti GD2 monoclonal antibody therapy).

CONCLUSION

Giant intrathoracic neurogenic tumors are rare, they are rarer in children. Neuroblastoma is commonest neurogenic tumor in paediatric age group. Giant intrathoracic neuroblastoma can mimic massive pleural effusion in chest x-ray. Even in CT scan or MRI, it is difficult to definitely tell their origin (mediastinum versus lung).

REFERENCES: