A Nervous Breakdown:

Multimodality Imaging of Thoracic Neurogenic Tumors

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GOALS AND OBJECTIVES

- Illustrate the complex anatomy of the nervous system within the chest wall and mediastinum
- Detail the important epidemiologic and pathophysiologic features to use in an approach to neurogenic tumors
- Identify key imaging features of neurogenic tumors occurring in the chest, focusing on distinguishing characteristics
- Explain the relative advantages of imaging modalities when examining neurogenic tumors
Sensation and motor innervation of the thorax
- Ventral nerves T1 – T11 lateral and anterior cutaneous branches
- Anterior branch has an accessory branch
- T1 contributes to the brachial plexus
- Special nerves such as phrenic nerve course through the thorax
- T12 part of the lumbar plexus called the subcostal nerve
Sympathetic Nervous System

- Sympathetic chain preganglionic sympathetic fibers arrive via white rami communications of ventral primary rami of spinal nerves T1 – L2
- Form greater thoracic splanchnic nerve, lesser thoracic splanchnic nerve, and least thoracic splanchnic nerve
**NEUROANATOMY OF THE THORAX**

**Parasympathetic Vagus nerve**

- Forms several different plexi providing parasympathetic innervation including, esophageal plexus, cardiac plexus, pulmonary plexus

[Diagram showing the recurrent laryngeal nerves, right vagus nerve, left vagus nerve, and esophageal plexus]
A recent study (Japanese Association for Research on Thymus) to define the mediastinum, divided it into the following components:

- Superior portion of the mediastinum
- Anterior mediastinum (prevascular)
- Middle mediastinum (peri-tracheoesophageal zone)
- Posterior mediastinum (paravertebral)
• Superior portion of the mediastinum borders:
  – Superior border thoracic inlet
  – Inferior border horizontal plane at the intersection of the caudate margin of the brachiocephalic vein with the trachea
  – Anterior sternum
  – Lateral parietal pleural reflections
  – Posteriolaterally by a vertical line against the posterior rim of the chest wall at the lateral rim of the thoracic vertebral transverse process
Anterior mediastinum (prevascular) borders:

- Superior boundary inferior boundary of the superior portion of the mediastinum
- Inferior diaphragm
- Anterior sternum
- Lateral parietal pleural reflections
- Posterior the percardium, anterior rims of the left brachiocephalic vein, superior vena cava, superior and inferior pulmonary veins, ascending aorta and the lateral rim of the aortic arch

Differential diagnosis based on paper results:
Thymoma (192), thymic carcinoma (52), thymic lymphoma (24), mature teratoma (24), malignant germ cell tumor (30), pericardial cyst (10) out of 343 tumors.
**Middle mediastinum (peritracheoesophageal zone) borders:**

- Superior boundary inferior boundary of the superior portion of the mediastinum
- Inferior diaphragm
- Anterior posterior rim of the left brachiocephalic vein, superior vena cava, ascending aorta, bilateral main pulmonary arteries and the heart
- Posterior, the anterior rim of the descending aorta and a vertical line connecting a point on each thoracic vertebral body at 1 cm behind it anterior margin

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Differential diagnosis based on paper results:
Bronchogenic cyst (25), neurogenic tumors (6), pericardial cyst (5), enteric cyst (2) out of 38 tumors
KEY FEATURES: LOCATION

- Posterior mediastinum (paravertebral) borders:
  - Superior boundary
  - Inferior boundary of the superior mediastinum
  - Inferior border diaphragm
  - Anterior boundary of the middle mediastinum
  - Posterior-lateral vertical line against the posterior rim of the chest wall at the lateral rim of the lateral process of the thoracic spine

Differential diagnosis based on paper results:
Neurogenic tumors (29) and bronchogenic cyst (7) out of 37 tumors

Axial contrast CT at the level of the heart
Another study evaluating location of neurogenic tumors found:

- Large airways – extremely rare: 26 reported cases from 1950 to 2003, 23 schwannomas and 3 neurofibromas
- Lung – extremely rare: 62 as pulmonary schwannomas, 17 as primary pulmonary paraganglioma
- Mediastinum – one of the most commonly encountered mediastinal tumors (comprising 15-25%), predominantly located in the posterior mediastinum
- Chest wall – one of the more usual chest wall tumors characterized by pressure deformity and displacement of the adjacent ribs and vertebrae
**KEY FEATURE: DEMOGRAPHICS**

**Differentiation based on age**

- Around approximately 15 years of age neurogenic tumors shift from ganglion cell tumors in patient’s younger than 15, to nerve sheath tumors in patient’s older.
- Similarly at around the same age the prevalence of malignant neurogenic tumors in patient’s less than 15 years shift to a greater prevalence of benign neurogenic tumor in patient’s older.
Neurofibromas and schwannomas typically demonstrate a lobulated spherical contour

- May have a dumbbell or hourglass configuration when projecting through the spinal canal

Ganglia tumors typically demonstrate oblong mass with a broad base anterolateral aspect of the spine
Morphological differences between schwannomas and ganglioneuromas in the mediastinum

- The craniocaudal length to major axis ratio (CC/M) may be used to differentiate between schwannoma and ganglioneuroma

Since the plane of growth of ganglion tumors is parallel to the spine the craniocaudal axes is longer than with schwannomas as their plane of growth is perpendicular to the spine
Morphological differences between schwannomas and ganglioneuromas in the mediastinum

- Plane of growth of ganglion tumors is parallel to the spine; the craniocaudal axes is longer than with schwannomas as their plane of growth is perpendicular to the spine.
IMAGING FEATURES OF NEUROGENIC TUMORS

MRI characteristics and usefulness

- MRI – schwannomas and neurofibromas typically have low to intermediate signal intensity on T1 weighted images and have areas of intermediate to high signal intensity on T2 weighted sequences.
- MRI – ganglia cell tumors homogenous intermediate signal intensity on all sequences, occasionally with a “whorled” appearance on T1 weighted images and heterogenous high signal intensity on T2 weighted sequences.
- MRI is performed to exclude intraspinal tumor extension.

Axial T2 non fat saturation image of the thorax demonstrates heterogenous internal signal intensity more commonly found in nerve sheath tumors.

Axial T2 non fat saturation image of the thorax demonstrates a more homogenous signal intensity mass in the posterior mediastinm more consistent with a ganglion cell tumor.
GANGLION TUMORS

Ganglioneuroma

- Fully differentiated neuronal tumor
- Patients generally older than 10, male predominant
- Composed of ganglion cells, schwann cells, and fibrous tissue

Key Imaging Features

- Generally occur in the posterior mediastinum (approximately 40%), then retroperitoneum, and cervical regions

Posteroanterior and lateral chest radiographs of a 7 year old with ganglioneuroma demonstrate a large mass in the posterior mediastinum. The mass is elongated and oriented in a vertical axis in the direction of the sympathetic chain.
Ganglioneuroma in a 7-year-old. Enhanced CT scan at the level of the heart shows a left paravertebral enhancing soft tissue mass displacing the heart to the right. The low internal attenuation is due to the abundant myxoid matrix. The mass is oriented along the sympathetic chain.

Multiplanar multisequence MRI demonstrates an intermediate signal intensity mass on all sequences with a somewhat whorled appearance caused by curvilinear or nodular bands of low signal intensity on both T1- and T2-weighted images.
GANGLIONEUROMA — CT FINDINGS

7 year old with a ganglioneuroma. Bone logarithm demonstrates punctate calcifications. Calcifications may be present in approximately 20% of tumors and is more common in sympathetic ganglia tumors than nerve sheath tumors.

Ganglioneuroma in a 7 year old. Coronal se illustrate the broad base along the anterolateral aspect of the spine spanning 3 to 5 vertebrae a common finding.

Coronal and sagittal images illustrate the broad base along the anterolateral aspect of the spine spanning 3 to 5 vertebrae a common finding.
Multiplanar multisequence MRI demonstrates an intermediate signal intensity mass on all sequences with a somewhat whorled appearance caused by curvilinear or nodular bands of low signal intensity on both T1- and T2-weighted images.
GANGLION TUMORS

Ganglioneuroblastoma

- Intermediate tumor between ganglioneuroma and neuroblastoma
- Variable imaging characteristics on a spectrum ranging from a similar appearance to ganglioneuroma → neuroblastoma
- Affect male and female subjects equally
- Presents in patients younger than 10 years old

Key Imaging Features

- May manifest as a sharply marginated oblong paraspinal mass or may be irregular, locally invasive, and widely metastatic
GANGLION TUMORS

Neuroblastoma

- Most common extracranial solid childhood malignancy
- Typically occurs in infants and very young children (mean age of presentation 22 months)
- Most commonly present in the abdomen, then mediastinum, neck and lower extremity

Key Imaging Features

- Non encapsulated, frequently containing extensive areas of hemorrhage, necrosis, and cystic degeneration
- May be further evaluated with mIBG, PET, or Tc-99m bone scan.

Posterioanterior radiograph of a 10 month old with neuroblastoma who presents with cough and fever demonstrates a paravertebral mass displacing the azygoesophageal reflection.
Axial contrast enhanced CT in a 10 month old demonstrates a paravertebral posterior mediastinal mass homogenous solid mass.

Sagittal T2 image demonstrates a homogenous paraspinal T2 hyperintense signal posterior mediastinal mass.

Coronal T1 fat saturation contrast enhanced image demonstrates a homogenous paraspinal enhancing posterior mediastinal mass.

Coronal I-123 mIBG demonstrates radiotracer localization within the mass.
PARAGANGLION TUMORS

Paraganglioma

- Tumor of paraganglion cells
- Found in sympathetic or parasympathetic chains
- Benign and malignant are histologically indistinguishable
- Component of hereditary paraganglioma-pheochromocytoma syndrome

Key Imaging Features

- Typically arise adjacent to the pericardium or heart, within the interatrial septum or left atrial wall, or from autonomic tissue along the paravertebral sulci
- Tumors often hypervascular on imaging and demonstrate central necrosis and hemorrhage

Coronal contrast-enhanced CT of a patient with paraganglioma in a characteristic location adjacent to the pericardium demonstrates a well-circumscribed, elongated mass with enhancement and prominent internal vascularity.
PERIPHERAL NERVE SHEATH TUMORS

• Derived from schwann cells
• Arise eccentrically from the parent nerve
• Symptoms related to compression of the parent nerve
• Peak presentation 40 and 50 years of age
• Histologically composed of two cell types: Antoni A (cellular) and Antoni B (myxoid)

SCHWANNOMAS

Key Imaging Features

• Well circumscribed encapsulated masses
• Adjacent bone changes may include splaying of the ribs, or enlargement of an intervertebral foramen.

Posteroanterior and lateral chest radiographs of a patient with schwannoma demonstrates a smoothly marginated, oval paraspinal mass.
Schwannoma – CT Findings

Axial and coronal non-contrast CT images demonstrate expansion of the T2 neuroforamen and erosion of the T2 vertebral body by a smoothly marginated mass in this patient with schwannoma.
SCHWANNOMA – MRI FINDINGS

Axial T2-weighted MR image of this same patient with schwannoma demonstrate characteristic neuroforaminal expansion, vertebral body erosion, and avid enhancement. Regions of high T2 signal intensity as well as central non-enhancing component are also typical features for schwannoma.

T1-weighted MR image of this same patient with schwannoma demonstrate characteristic neuroforaminal expansion, vertebral body erosion, and avid enhancement. Regions of high T2 signal intensity as well as central non-enhancing component are also typical features for schwannoma.

T1 post-gadolinium MR image of this same patient with schwannoma demonstrate characteristic neuroforaminal expansion, vertebral body erosion, and avid enhancement. Regions of high T2 signal intensity as well as central non-enhancing component are also typical features for schwannoma.
PERIPHERAL NERVE SHEATH TUMORS

NEUROFIBROMAS

Localized neurofibroma
- Most common form of neurofibroma (90%)
- Primarily affect superficial cutaneous nerves
- Slow growing
- Usually < 5 cm in size
- Peak presentation between 20 and 30 years of age
- Benign neoplasms composed of schwann cells and fibroblasts, containing a rich network of collagen fibers

Key Imaging Features
- Unencapsulated, infiltrating between nerve fascicles
- May undergo malignant transformation to malignant peripheral nerve sheath tumor

Anteroposterior and lateral chest radiographs of a patient with mediastinal neurofibromas demonstrate a smoothly lobulated mass in the aorticopulmonary window and widening of the left superior mediastinum.
Axial contrast enhanced CT of this same patient with mediastinal neurofibromas demonstrates multiple conglomerate mediastinal masses.
PERIPHERAL NERVE SHEATH TUMORS

MALIGNANT PERIPHERAL NERVE SHEATH TUMORS (MPNST)

- Account for 5-10% of all soft tissue sarcomas
- Metastasize hematogenously, frequently to the lungs
- Most are high grade tumors (85%)
- Half associated with NF1
- May arise de novo or de-differentiate from an existing peripheral nerve sheath tumor

Key Imaging Features

- Malignant features:
  - Large lesion (> 5 cm)
  - Irregular borders
  - Rapid growth on interval imaging

Axial and coronal FDG PET/CT images of a patient with MPNST demonstrate a large right chest wall mass with intense activity. Central area of decreased activity correspond to cystic and necrotic components.
Axial and coronal contrast-enhanced CT images of a patient with MPNST demonstrate a large, enhancing mass in the right chest wall with vascular and cystic components.
MALIGNANT PERIPHERAL NERVE SHEATH TUMOR – MRI FINDINGS

Coronal STIR demonstrates a heterogenous T2 signal mass along the right chest wall, which was greater than 5 cm with irregular borders.

Coronal T1 demonstrates a heterogenous T1 signal mass with areas of increased T1 signal likely due to hemorrhage, along the right chest wall, which was greater than 5 cm, with irregular borders.

Sagittal proton density demonstrates a heterogenous mass along the right chest wall.

Axial STIR demonstrates a heterogenous T2 signal mass with fluid-fluid levels along the right chest wall, which was greater than 5 cm with irregular borders.

Axial precontrast T1 non fat sat demonstrates a heterogenous T1 signal mass along the right chest wall.

Axial postcontrast T1 fat sat show avid heterogenous enhancement with areas of necrosis.
NERVE SHEATH TUMORS

NEUROFIBROMATOSIS TYPE 1

• Autosomal dominant disorder with a frequency of 1 in 3000.
• Plexiform neurofibroma is pathognomonic

Key Imaging Features

• Rib deformity
• Scoliosis
• Posterior scalloping of vertebral bodies from dural ectasia
• Thin-walled bullae, fibrosis

Coronal T2-weighted MR image with fat saturation of a patient with neurofibromatosis type 1 shows multiple T2 hyperintense masses, many of which demonstrate low signal intensity centrally.
Mediastinal Mass (Superior or Posterior)

Suspect neurogenic tumor

Shape

- Plane of growth parallel to the spine – Nerve sheath tumor
- Plane of growth perpendicular to the spine – ganglion cell tumor

Imaging characteristics

- T2 heterogenous internal signal intensity more commonly found in nerve sheath tumors
- T2 homogenous signal intensity mass in the posterior mediastinum more consistent with a ganglion cell tumor

Age

- Posteroanterior radiograph of a patient <15 years of age with ganglioneuroma demonstrate a large mass in the posterior mediastinum.
- Posteroanterior radiograph of a patient >15 years of age with schwannoma demonstrates a smoothly marginedated, oval paraspinal mass.

Aggressive or non aggressive features

Benign versus malignant
Mediastinal Mass (Superior or Posterior)  

Suspect neurogenic tumor  

Shape  
- Plane of growth parallel to the spine or perpendicular to the spine – Nerve sheath tumor  
- Plane of growth perpendicular to the spine – ganglion cell tumor  

Imaging characteristics  
- Heterogenous tumoral T2 signal, homogenous tumoral T2 signal  

Aggressive or non aggressive features  
- Benign versus malignant  

Age  
- Less than 15 years of age or greater than 15 years of age  

Axial T2 non fat saturation image of the thorax demonstrates heterogenous internal signal intensity more commonly found in nerve sheath tumors  

Axial T2 non fat saturation image of the thorax demonstrates a more homogenous signal intensity mass in the posterior mediastinum more consistent with a ganglion cell tumor  

Posteroanterior radiograph of a patient <15 years of age with ganglioneuroma demonstrate a large mass in the posterior mediastinum.  

Posteroanterior radiograph of a patient >15 years of age with schwannoma demonstrates a smoothly marginated, oval paraspinal mass.
Neurogenic tumors of the chest may be a diagnostic challenge both clinically and radiologically.

A detailed anatomic understanding of the chest nervous system will serve as a foundation for evaluating these tumors.

Attention to key imaging features on imaging will suggest neurogenic tumors in a differential diagnosis.

The complementary features of imaging modalities, when used deliberately and protocolled appropriately, will further refine a differential diagnosis and ultimately improve patient care.