The Breadth of the Diaphragm: Congenital and Acquired Derangements Including 3D Imaging

Disclosures

• The authors have no financial disclosures related to this presentation

Goals and objectives

• Review the normal imaging appearance of the diaphragm
• Discuss updates regarding development of the diaphragm
• Review congenital abnormalities of the diaphragm
• Review both benign and malignant abnormalities of the diaphragm
Introduction

• The diaphragm is a unique skeletal muscle that enables respiration

• Diaphragm abnormalities are often initially discovered on chest radiographs
  • Alteration in position
  • Alteration in shape

• Evaluation of structural defects and intrinsic/adjacent pathology
  • Performed with CT and occasionally MRI

• Evaluation of diaphragm motion
  • Performed with fluoroscopy and occasionally MRI
Normal diaphragm

- Normal position of the diaphragm can vary substantially.
- The right hemidiaphragm dome is typically between T10-T12.
- Left hemidiaphragm dome is typically ½ vertebral body lower.
  - 5-10% of patients are at the same level or higher.
- The diaphragm tends to be lower with increasing age and higher with increasing weight.

Right dome at T10  Left dome slightly below
Normal diaphragm

• Signs distinguishing right and left hemidiaphragms
  • Entire anterior-posterior extent of right diaphragm should be present

• Variable segment of anterior left diaphragm is obscured by heart diaphragm interface

• Gastric bubble under left

• Right ribs are magnified more than left

Right hemidiaphragm is associated with the magnified ribs
Normal diaphragm

- The diaphragm is best seen on CT when surrounded by fat
- More difficult to see when adjacent to liver or spleen, particularly without contrast

- The right crura is typically larger than the left
- Decussation of the right crura forms the esophageal hiatus
- The diaphragm muscle bundles can take a more nodular appearance at their insertion point near the ribs
Development of the diaphragm

- Development occurs during 4-12 weeks of embryonic life
- Complex interplay of structures:
  - Septum transversum
    - Initial structure separating the thorax and abdomen
  - Posthepatic mesenchymal plate (PHMP)
    - Eventually seals the chest from the abdomen
  - Pleuroperitoneal folds
    - Scaffolding positioned posterolaterally in the cavity
  - Dorsal esophageal mesentery
    - Forms the muscular portion of the diaphragm

Early diaphragm
Development of the diaphragm

- A study by Mayer et al. in 2011 showed the PHMP plays a central role in sealing the diaphragm

- Abnormal development of the PHMP is central in the formation of congenital diaphragmatic hernias

- Mayer et al. demonstrated in rats that although the pleuroperitoneal folds and septum transversum appear normal, the PHMP is smaller and malformed

- This results in persistence of the pleuroperitoneal canals with resultant formation of a diaphragmatic defect
Congenital abnormalities
Bochdalek hernia

• Results from incomplete closure of the pleuroperitoneal membrane in utero

• Typically posterolateral defects and usually on the left

• Variable size
  • Small – only retroperitoneal fat
  • Large – can contain viscera

• Incidental finding in adults
  • Increasing frequency with age suggests these can be acquired
Congenital abnormalities

Bochdalek hernia

Congenital Bochdalek hernia containing non-obstructed large bowel

Subsequent development of bowel obstruction in same patient
Congenital abnormalities

Bochdalek hernia

- Large Bochdalek hernias (congenital diaphragmatic hernia) in the neonatal period can represent a surgical emergency
  - Common with incidence of ~1:3,000
  - Chest radiograph
    - Opacification of hemithorax with abdominal viscera
    - Mediastinal shift
    - Ultrasound can be used to diagnose prenatally
- Can result in lung hypoplasia
Congenital abnormalities
Morgagni hernia

- Related to abnormal development of the septum transversum
- Uniquely, the Morgagni hernia is covered with a sac of peritoneum and pleura
  - Typically on the right
- Variable size
  - Small – contains omentum
  - Large – contains viscera
    - Can result in volvulus and strangulation
Acquired abnormalities
Hernias
Hiatal hernia (type I)

- Hiatal hernia is the most common hernia in adults
  - GE junction slips into thorax
  - More common than paraesophageal type
  - Prevalence increases with age
- Associated with obesity and pregnancy
  - Increased abdominal pressure
- Considered a risk factor for GERD
- Radiograph
  - Retrocardiac mass with air-fluid level
- CT and MRI
  - Extension of the proximal aspect of the stomach into chest

Air fluid level
Intrathoracic stomach
Acquired abnormalities
Hernias
Paraesophageal (type II-IV)

- Less common than hiatal hernia (type I)
- Includes a peritoneal sac as opposed to the simple sliding hiatal hernia, which does not
- Can be asymptomatic or symptomatic
  - When symptomatic, at higher risk of developing incarceration
- Classified into three types:
  - Type II – stomach herniates but gastroesophageal junction remains normally positioned
  - Type III – combination of type I and II
  - Type IV – stomach along with other abdominal organs herniate into chest
Acquired abnormalities
Traumatic diaphragm hernia

• Secondary to blunt or penetrating injury

• Diaphragm injuries occur more frequently on the left
  • Blunt: related to protection by liver on the right
  • Penetrating: related to most people being right handed

• Injury typically affects the posterior aspect of the diaphragm or posterolateral diaphragm attachments

• Depending on side of injury, stomach or liver may herniate
  • If large, may include bowel or spleen

• Early diagnosis is important because defects can enlarge
Acquired abnormalities
Traumatic

- Chest radiograph
  - Sensitivity ranges from 20-71%

- Radiographic findings
  - Herniation of abdominal viscera
  - NG tube within intrathoracic stomach
  - Elevated hemidiaphragm
    - Particularly on the right
    - Suspect injury if right is elevated 4-5 cm above left

Elevated diaphragm
Acquired abnormalities
Traumatic

• Chest CT with multi-planar reformats
  • Sensitivity: 61-100%
  • Specificity: 77-100%

• Findings:
  • Dependent viscera – abnormal contact of the abdominal viscera with the posterior ribs
  • Collar sign – constriction of herniated contents
  • Hump sign – focal bulge of liver
  • Absent diaphragm sign – diaphragm is not present in a location where it should contact an organ
  • Abrupt discontinuity of the diaphragm
  • Asymmetric thickening of the diaphragm

Dependent viscera sign with lacerated spleen
Collar sign
Acquired abnormalities
Traumatic

- Indications of penetrating trauma
  - Wound tract
  - Herniation of fat through diaphragmatic defect
  - Focal thickening of diaphragm
    - Blood and/or edema

Wound tract
Hematoma

Active extravasation of contrast on delayed imaging
Acquired abnormalities
Positional – elevation

- Unilateral elevation causes
  - Volume loss
  - Eventration
  - Phrenic nerve paralysis
  - Abdominal disease
  - Mimics include
    - Subpulmonic effusion, pleural mass, diaphragmatic hernia
Acquired abnormalities
Positional – elevation

- Bilateral elevation causes
  - Volume loss
  - Abdominal mass effect
  - Bilateral eventration
  - Bilateral subpulmonic effusion
  - Neuromuscular disease
  - Connective tissue disease
  - Phrenic nerve paralysis

Shallow inspiration with elevation of both hemidiaphragms
Acquired abnormalities
Positional – depression

- Unilateral depression causes
  - Pneumothorax
  - Bullous emphysema
  - Large pleural effusion
  - Congenital lobar emphysema

Large tension pneumothorax with depression of the right hemidiaphragm
Acquired abnormalities
Positional – depression

• Bilateral depression causes
  • COPD
  • Deep inspiration
  • Bilateral large pleural effusion
  • Mechanical ventilation at high pressures
  • Cystic fibrosis
  • Pulmonary langerhans cell histiocytosis
  • Lymphangioleiomyomatosis

COPD with marked hyperinflation and depression of both hemidiaphragms
Acquired abnormalities
Paralysis

- Can result from any abnormality along the neuromuscular axis of the diaphragm
  - Invasion by neoplasm
  - Trauma related to surgery
  - CNS conditions such as multiple sclerosis, syringomyelia, neurofibromatosis
  - Mass effect
    - Aortic aneurysm, goiter, lymphadenopathy
  - Idiopathic

Pre-op

Development of left hemidiaphragm paralysis following CABG
Acquired abnormalities
Paralysis

- Fluroscopic sniff test
  - Gold standard
  - Evaluates for paradoxical upward diaphragm motion with rapid inspiration through the nose
  - Normal diaphragm excursion is > 2.5 cm
  - Excursion < 2.5 cm may be normal or represent diaphragm weakness
  - Paradoxical upward motion of the diaphragm > 2 cm suggests paralysis

- Confounding factors
  - Sniff test is best performed when reversible conditions affecting the diaphragm have resolved
    - Pneumonia, pleuritis, peritonitis, etc…
  - Severe weakness or complete eventration can be difficult to distinguish from paralysis

MRI sniff test, which has the added ability to assess for extra-diaphragmatic pathologies not seen with fluoroscopy
Acquired abnormalities
Tumors – mimics

- Lateral arcuate ligament pseudotumor
  - Ligament that courses along the posterior ribs
  - Can take on a nodular appearance simulating a retroperitoneal tumor
  - Can be bilateral, which is helpful in excluding a tumor
  - When unilateral, typically has a curvilinear appearance and is contiguous with the diaphragm

Diaphragm lateral muscle bundles

A single muscle bundle can simulate a tumor. FDG PET/CT demonstrated non-avidity
Acquired abnormalities
Tumors – benign

- Rare
- Benign tumors occur with similar frequency as malignant tumors
- Most commonly reported tumor is a benign cyst
  - Bronchogenic or mesothelial in origin
- Lipomas are the most commonly reported solid benign tumor
  - Does not disrupt the integrity of the diaphragm as opposed to a Bochdelak hernia
Acquired abnormalities
Tumors – benign

Diaphragmatic lipoma

Diaphragm cyst

Diaphragm cyst

Axial
Axial
T2 weighting

Coronal
Coronal
Post contrast
Acquired abnormalities
Tumors – malignant

- Rare
- Typically sarcomas of fibrous or muscular origin
- The most common malignant tumor is rhabdomyosarcoma
- Other types
  - Schwannoma
  - Chondroma
  - Pheochromocytoma
  - Endometriosis
  - Hemangiopericytoma
- The diaphragm is a thin structure, which can make assessing the origin of a tumor difficult
  - Tumors of the lung, pleura, or abdominal viscera are far more common
Acquired abnormalities
Tumors – malignant

Rhabdomyosarcoma

FDG avid on PET/CT
Acquired abnormalities
Tumors – malignant

Fibromyxoid sarcoma

Heterogeneously hyperintense on T2 weighting

Hypointense on T1 fat sat

Heterogeneously enhancing on T1 post-gad
Acquired abnormalities
Tumors – malignant

Myxoid chondrosarcoma

Hyperintense on T2 weighting
Hypointense on T1 fat sat
Enhancing on T1 post-gad
3D modeling

• Often described as an imaging revolution

• 3D modeling offers an incredibly powerful pre-surgical planning and education tool

• High resolution CT or MRI is obtained based on relevant anatomy
  • Slice thickness is important in determining the level of model detail
  • Choice of reconstruction algorithm (i.e. bone vs soft tissue) influences how the model looks

• Acquired data set from CT or MRI is transferred to an independent workstation

• A 3D model is created using a combination of hand tracing and automatic segmentation

• The finalized 3D model is exported in STL file format, which is recognized by 3D printers
3D printing

• At our institution, a multi-material printer is used, which generates objects using additive printing

• A photopolymer is laid down by the printer and hardened with UV light

• Materials can be hard or soft and colored differently

• Material costs can range from hundreds to thousands of dollars and print time can take hours to days

• Models can be used for pre-operative planning or a reference during surgery

• Models can function as tools for both patient and medical education

Printing of diaphragm hernia for repair
References
