An Unusual Presentation of Diaphragmatic Hernia

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Patient LG: Clinical Presentation, Nov. 2004

• 52 year old woman presents with new nonproductive cough & dyspnea
• PMH: Obesity, recurrent bronchitis
• ROS: No GI or GU complaints, no history of prior trauma or major surgery
• PE: Decreased breath sounds at left lung base
Patient LG: Chest Radiograph, 11/26/04

- Elevated hemidiaphragm
- Bowel gas
- Shifted mediastinum
- Crowded vessels?
- Atelectasis?
- Elevated hemidiaphragm
DDx: Elevated Hemidiaphragm

• Lung conditions “pulling” up diaphragm
  – Atelectasis
  – Prior lobectomy
  – Pulmonary disease, e.g., pulmonary fibrosis

• Abdominal (and other) conditions “pushing” up diaphragm
  – Organ enlargement, e.g., splenomegaly, distended stomach
  – Inflammatory or infectious process, e.g., subphrenic abscess
  – (Subpulmonic effusion—“pushes” up lung from above diaphragm; not a truly elevated diaphragm)

• Diaphragmatic defects
  – Eventration—muscular defect causing weakness
  – Paralysis—elevation and paradoxical movement
  – (Hernia—not a truly elevated diaphragm)
In fact, after a recent colonoscopy was limited by a redundant colon, patient LG underwent virtual CT colonoscopy which revealed her diagnosis…
Patient LG: Virtual Colonoscopy
Coronal CT, 8/3/04

Herniated bowel
Patient LG: Virtual Colonoscopy
Axial CT, 8/3/04
Amazingly, when this large diaphragmatic hernia was first diagnosed in August 2004, at age 52, LG was asymptomatic.

Her respiratory symptoms would only begin several months later.
From November to December 2004, LG continued to have progressive dyspnea and cough, increasingly productive of greenish sputum, and intermittent fevers.

Outpatient CT on 12/28/04 demonstrated pneumonia with LUL abscess and pleural effusion.

Patient was admitted to BIDMC later that day for further work-up and management.

CT-guided abscess drainage was performed on 1/1/05.
Patient LG: CT-Guided Abscess Drainage, 1/1/05

NON-CONTRAST AXIAL CT

Pigtail Catheter
Patient LG: Axial Contrast CT, 1/3/05

CT, 5 months ago

Abscess w/ catheter

Empyema

Kidney

Courtesy of Dr. Khasgiwala
Patient LG: Coronal and Sagittal Contrast CT, 1/3/05

CORONAL

Abscess w/ catheter
Colon

SAGITTAL

Empyema
Colon
Kidney
Diaphragm

Courtesy of Dr. Khasgiwala
Patient LG: Hospital Admission
12/28/04-1/7/05

• Diagnosis
  – Bochdalek diaphragmatic hernia
  – Streptococcus milleri pneumonia complicated by LUL abscess and multiloculated empyema

• Treatment
  – Abscess drainage
  – Antibiotics (CTX)

• Elective diaphragmatic repair was deferred pending resolution of infection
Let’s put our patient’s unusual presentation into a broader context
Diaphragmatic Hernias (DH): Classification

• Congenital
  – Bochdalek
  – Morgagni
  – Hiatus
• Idiopathic
• Acquired
  – Traumatic
  – Iatrogenic
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Development of Diaphragm

GESTATIONAL AGE

Week 7

- Pleuroperitoneal fold
- Septum Transversum

Week 9

- Body wall muscle ingrowth
- IVC

Week 14

- Dorsal mesentery of esophagus
- Es

Defect leads to
Bochdalek hernia

Defect leads to
Morgagni hernia

Defect leads to
hiatus hernia

Adapted from Sadler, Langman’s Medical Embryology, 9th Ed, 218.
Mature Diaphragm Diagram

INFERIOR SURFACE

Morgagni foramina

Esophageal Hiatus

Bochdalek foramina

R

L

Sternal fibers
Costal fibers
Central tendon

IVC
Aorta

Moore and Agur, *Essential Clinical Anatomy*, 2nd Ed, 188.
Congenital DH: Bochdalek

- Posterolateral defect through Bochdalek foramen
- Most common congenital diaphragmatic hernia (1:2200 live births)
- L>R>>bilateral
  - L: bowel, stomach, fat, spleen, kidney
  - R: liver, fat, kidney
- Neonates with large hernias often present with respiratory distress due to poor fetal lung development
Congenital DH: Morgagni

- Anteromedial defect through Morgagni foramen
- Rare (1:1,000,000 live births)
- R>L
- Often small, containing only fat, and asymptomatic
Congenital DH: Hiatus

- Herniation of stomach through esophageal hiatus
- Overall rare in children
- Of the three different subtypes of hiatus hernia (see below), paraesophageal is most common congenital form
  - Congenital paraesophageal hernias not often associated with complications (e.g., obstruction), as in adults

Type I
“Sliding”

Type II
Paraesophageal

Type III
Mixed

Moore and Agur, *Essential Clinical Anatomy*, 2nd Ed, 188.
Imaging Pediatric DH

- Prenatal ultrasound (US)
- Neonatal radiographs
  - Contrast studies may help
Companion Patient #1 Prenatal US: Congenital DH

Findings suggestive of DH:
- Abdominal organs in thorax
- Contralateral mediastinal shift
- Small abdominal circumference

Hedrick & Adzick, UpToDate, 2006.
Prenatal US: Congenital DH

• **Advantages**
  – Routinely performed
  – Safe for woman and fetus
  – Early diagnosis
    • Search for other associated anomalies (prenatal karyotype, echo)
    • Opportunity for prenatal intervention (e.g., fetal tracheal occlusion)
    • Plan for delivery and critical postnatal care at tertiary hospital
    • Prepare parents psychologically

• **Disadvantages**
  – User dependent
  – Limited resolution
  – May not detect smaller abnormalities
Companion Patient #2 Neonatal Radiograph: Congenital Bochdalek DH

**DDx:**
- Bochdalek diaphragmatic hernia
- Congenital cystic adenomatoid malformation
- Cystic pulmonary interstitial emphysema
Companion Patient #3 Neonatal Radiograph: Congenital Morgagni DH

AP

Lateral

Bowel

Mediastinum

Compressed

Lung

Bowel

Compressed

Lung

Courtesy of Dr. Khasgiwala
Companion Patient #4 Neonatal Radiograph: Congenital Hiatus DH

Cystic mass in posterior mediastinum

Companion Patient #5 Neonatal Barium Study: Congenital Hiatus DH

Barium study performed to distinguish from other posterior mediastinal cystic masses, such as:
- Epiphrenic diverticulum
- Pulmonary cyst
- Cystic tumor

Neonatal Radiograph: Congenital DH

- **Advantages**
  - Widely available and cheap
  - Demonstrates anatomy
  - Contrast studies (e.g., barium) may be used for confirmation
  - Can track progress and complications of interventions (e.g., lines, catheters, pulmonary disease, pre/post-op)

- **Disadvantages**
  - Exposes child to radiation
  - Limits to identifying involved structures
  - If small hernias are missed by US and asymptomatic, they will not be detected
And now let’s turn to other presentations of diaphragmatic hernias in adults, which sometimes recapitulate (if not represent) congenital phenotypes.
Diaphragmatic Hernias (DH): Classification

• Congenital
  – Bochdalek
  – Morgagni
  – Hiatus

• Idiopathic

• Acquired
  – Traumatic
  – Iatrogenic
Imaging Adult DH

- Radiographs ± contrast
- Cross-sectional studies: CT, MR
  - Characterize anatomy of hernia
  - Identify potential complications (e.g., respiratory, GI)
  - Directly identify diaphragmatic defect
Idiopathic DH: Hiatus Hernias

- Most common diaphragmatic hernia overall, usually of unclear etiology
  - Type I (sliding) predominates
    - About half present with GERD
    - Usually medically managed
  - Other types (e.g., paraesophageal) less common
    - May present with obstruction due to volvulus
    - Surgical repair is indicated, even if incidental and asymptomatic

Type I
“Sliding”

Type II
Paraesophageal

Type III
Mixed

Companion Patient #6 Barium Study: Type I Hiatus Hernia

- Esophagus
- A ring
- Gastric rugal folds
- Esophageal hiatus

Kahrilas, Pandolfino. *GI Motility online* 2006 | doi:10.1038/gimo48
Idiopathic DH: Other Types

• There are multiple case reports of Bochdalek and Morgagni hernias of unclear etiology in adults, which are diagnosed incidentally or because of symptoms.

• A retrospective review of 13,138 abdominal CT reports for adults patients at a large urban hospital identified incidental Bochdalek hernias in 0.17%, of which 27% involved solid or enteric organs.

Companion Patient #7 Radiographs and CT: Morgagni DH in Asymptomatic 64yo Female

**RADIOGRAPH**

- **PA**
  - Arrow = R anterior cardiophrenic mass

- **Lateral**
  - Arrow = R anterior cardiophrenic mass

**PARASAGITTAL CONTRAST CT**

- Asterisk = herniated mesenteric fat
- Arrowheads = anteromedial diaphragmatic defect

Acquired DH: Trauma

• Penetrating trauma
  – Direct injury to diaphragm causes rupture
  – Often undergo quick surgical repair

• Blunt trauma
  – Impact may lead to direct or indirect injury of diaphragm
    • Increased abdominal pressure may push abdominal structures through a weakened diaphragm
  – Many hernias are missed early, and patients can present late with respiratory illness or GI complication (e.g., obstruction)
  – L>R>bilateral (R-sided protection of liver)
Companion Patient #8 Radiograph & Axial CT: Traumatic DH in 47yo Male s/p MVA

PA CHEST RADIOGRAPH (CONED DOWN)

- Effusion
- Bowel
- Rib fractures

AXIAL CONTRAST CT

- Diaphragm defect
- Bowel
- Fat
- Hemothorax
- Rib fracture

Acquired DH: Iatrogenesis

• Thoraco-abdominal surgeries, esp. esophago-gastrectomy (e.g., for esophageal cancer), may lead to acquired DH
  – Similar presentation and complications to traumatic hernias
Diaphragmatic Hernia Repair

• Medical management suffices for most sliding hiatus hernias and small idiopathic hernias

• Surgical repair is indicated for most pediatric, acquired, and otherwise symptomatic adult DH
So what finally happened with our patient?
Patient LG: Clinical Course

- After resolution of infection with many months of antibiotic therapy (CTX followed by Levo/Clinda), LG underwent successful surgical repair of the diaphragm in July 2005.
- Surgery revealed:
  - extensive adhesions from previous empyema
  - 8 x 5 cm defect in the posterolateral diaphragm consistent with “a congenital Bochdalek type hernia” (per operative report)
Patient LG: Chest Radiograph Before and After Diaphragm Repair

**PRE-OP**
7/6/05
PA

**POST-OP**
9/8/05

Key findings:
- Repaired diaphragm
- Normal mediastinum
- Minor post-op changes

Linear atelectasis
Pleural effusion
Summary

• Congenital diaphragmatic hernias can be classified as Bochdalek, Morgagni, or hiatus types

• Prenatal imaging followed by neonatal radiographs represent the best tests to identify congenital diaphragmatic hernias and track the children’s clinical course

• For adults with suspected idiopathic or acquired diaphragmatic hernias, radiographs ± contrast and cross-sectional imaging can best characterize the defects and their associated complications
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References

• Hedrick HL and Adzick NS. Congenital diaphragmatic hernia: Prenatal diagnosis and management. *UpToDate* 2006.
• Kahrilas PJ. Hiatus Hernia. *UpToDate* 2005.
• Kahrilas PJ and Pandolfino JE. Hiatus hernia. *GI Motility online* 2006; doi:10.1038/gimo48.