July 2018 Imaging Case of the Month

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Clinical History: An 81-year old non-smoking woman presented with complaints of shortness of breath for one month, more so when laying down. The patient had a history of Sjögren syndrome established 13 years earlier. She notes a history of dryness of the eyes and upper airways. Her medications included 5 mg prednisone daily as well as various vitamins and supplements. While she complained of several medication “allergies,” none were serious and most appeared to represent side effects or untoward reactions to medications as opposed to true allergic reactions. Her past medical history included arthritis, possible obstructive sleep apnea, kidney stones, and orthostatic hypotension, the latter thought to be related to her Sjögren syndrome. Her surgical history included a sternotomy for thymoma resection years earlier.

Her physical examination was unremarkable except for diminished breath sounds at the left base; her vital signs were within normal limits.

Frontal and lateral chest radiography (Figure 1) was performed.

Figure 1. Frontal (A) & lateral (B) chest radiography.

Which of the following represents **the most accurate assessment** of the chest radiographic findings?

1. Chest radiography shows an elevated left hemidiaphragm
2. Chest radiography shows bibasilar fibrotic-appearing opacities
3. Chest radiography shows cavitary pulmonary lesions
4. Chest radiography shows multifocal bronchiectasis
5. Chest radiography shows small pulmonary nodules

Correct!

1. Chest radiography shows an elevated left hemidiaphragm

Chest radiography (Figure 2) shows elevation of the left hemidiaphragm, suggesting paresis or paralysis.

Figure 2. Frontal (A) & lateral (B) chest radiography shows an elevated left hemidiaphragm, presumably reflecting phrenic nerve palsy related to the previous sternotomy. A dilated esophagus, manifesting as a prominent lucency (arrows), is visible. On the lateral projection, the combined posterior tracheal membrane and anterior wall of the esophagus, referred to as the posterior tracheo-esophageal stripe (arrowheads), are visible.

No nodules are evident, and there is no evidence of basilar fibrosis. No definite features to suggest bronchiectasis. No cavitation is evident.

At this point, which of the following represents the **most appropriate step** in this patient's management?

1. ^{18}F FDG-PET scan
2. Bronchoscopy with transbronchial biopsy
3. Comparison to prior studies
4. Functional fluoroscopic study of the chest
5. Thoracic MRI

Correct!

3. Comparison to prior studies

Thoracic MR (considered separately from cardiac MR) can be useful for examination of the mediastinum, occasionally the pleura, but is generally not very useful for examination of the lungs. As no clear significant lung or mediastinal pathology is evident at chest radiography, bronchoscopy and ^{18}F FDG-PET scanning are not indicated at this point. A functional fluoroscopic, or “sniff” test, is appropriate to evaluate for the possibility of either weak or paralyzed left diaphragm, given the left diaphragm elevation, but, as is almost always the case, comparison to remote studies is the first priority when chest radiographic abnormalities are encountered.).

Prior chest radiography from 2008 (Figure 3) was located.

Figure 3. Chest radiography performed 10 years earlier (B) and the current (A) study.

Which of the following represents **the most accurate assessment** of the current and prior chest radiographic findings?

1. Chest radiography shows linear and reticular abnormalities suggesting fibrosis on the prior (Figure 2) that had resolved as of the *current* study (Figure 1)
2. Chest radiography shows multifocal consolidation on the *prior* (Figure 2) that had resolved as of the *current* study (Figure 1)
3. Chest radiography shows multifocal pleural disease on the *prior* (Figure 2) that had resolved as of the *current* study (Figure 1)
4. Chest radiography shows nodules on the *prior* (Figure 2) that had resolved as of the *current* study (Figure 1)
5. Chest radiography shows persistent elevation of the left diaphragm on the *prior* (Figure 2) and the *current* study (Figure 1), essentially unchanged

Correct!

5. Chest radiography shows persistent elevation of the left diaphragm on the prior (Figure 2) and the current study (Figure 1), essentially unchanged

Both the previous radiographs show no evidence of pulmonary nodules, areas of consolidation, features of fibrotic lung disease, or significant pleural disease. The left diaphragm is persistently elevated- this finding was present on both chest radiographs- and was thought to be the result of the previous sternotomy for thymoma..

Which of the following statements represents **the most accurate assessment** of the current and prior chest radiographic findings?

1. Chest radiography shows a dilated esophagus has developed on the *current* study
2. Chest radiography shows an interval gastric pull-through has been performed since the *prior* study
3. Chest radiography shows more apparent medial paraseptal emphysema on the *current* versus the *prior* study
4. Chest radiography shows new cystic lung abnormalities
5. Chest radiography shows new pneumomediastinum on the *current* versus the *prior* study

Correct!

1. Chest radiography shows a dilated esophagus has developed on the current study

Chest radiography shows lucency projected over the mediastinum on the frontal projection, posterior to the heart on the lateral projection, consistent with the location of the esophagus. This finding does not represent pneumomediastinum, and the position is inconsistent with paraseptal emphysema. The lucent focus resides within the mediastinum and therefore does not represent cystic lung disease. A gastric pull-through following esophagectomy is a consideration, but typically such a surgery is not performed through a sternotomy but rather through a right thoracotomy, and most commonly for esophageal malignancy. This patient does not have a history of previous right thoracotomy or esophageal carcinoma, nor does she have a history of a gastric pull-through between the two examinations.

Which of the following is the **most appropriate** consideration for the finding on the **current** (Figure 1) chest radiograph?

1. Esophageal diverticula
2. Esophageal obstruction
3. Esophageal pneumatosis
4. Intrathoracic stomach with gastric torsion
5. Tracheo-esophageal fistula

Correct!
2. Esophageal obstruction

The appearance of the mediastinal gas is consistent with an esophageal location. The rather long cephalocaudad appearance of this lucency suggests distal esophageal obstruction. An intrathoracic stomach with torsion can present with esophageal dilation owing to obstruction, but a globular gas lucency, presenting gas within the obstructed stomach, would also be present. A trachea-esophageal fistula would not necessarily be associated with esophageal dilation, and may be accompanied by infiltrative lung abnormalities owing to the passage of material from the esophagus into the trachea and then into the pulmonary parenchyma. Esophageal diverticula may be seen as focal gas-containing lucencies, but generally not a tubular-shaped, long-segment gas-containing mediastinal structure. Esophageal pneumatosis would manifest as thin, linear streaks of gas tracking through the esophageal wall.

A prior thoracic CT (Figure 4) from 2 years earlier, performed in an emergency room for shortness of breath and suspected pulmonary embolism, was located.

Figure 4. Left A-I: Axial enhanced thoracic CT displayed in soft tissue windows obtained as part of acute pulmonary embolism evaluation for shortness of. Right A-I: lung windows.

Which of the following is the **most appropriate** consideration for the findings on this thoracic CT scan?

1. Barrett's esophagus
2. Esophageal carcinoma
3. Esophageal gastrointestinal stromal tumor
4. Esophageal intramural pseudodiverticulosis
5. Primary achalasia

Correct!
5. Primary achalasia

The thoracic CT shows diffuse esophageal distension extending to the level of the gastroesophageal junction, but there is no evidence of mass at the point of esophageal tapering, as would be expected if an esophageal malignancy were causing esophageal obstruction, particularly if carcinoma were the cause of such obstruction. Note, however, esophageal malignancy presenting primarily as stricture may cause obstruction with little evidence of actual esophageal mass at thoracic CT. Barrett's esophagus typically shows no discernable findings at thoracic CT. A gastrointestinal stromal tumor affecting the esophagus may appear as a solid mass of variable size, which may be eccentric and unassociated with obstruction, or may cause esophageal luminal narrowing resulting in obstruction. Intramural esophageal pseudodiverticulosis appears as numerous, multifocal, "flask-shaped" small intramural esophageal wall oral contrast collections at barium esophagram; these findings would be difficult to detect at thoracic CT. The appearance of extensive upper and mid-esophageal distension extending to a normal-appearing gastro-esophageal junction, without a visible mass, is typical of achalasia, and, while not this consideration cannot be specifically established as a definitive diagnosis at thoracic CT, achalasia is the leading consideration for the CT appearance in this patient.

The patient subsequently underwent barium esophagram (Figure 5) for evaluation of the presumed esophageal dilation seen at chest radiography and thoracic CT.

Figure 5. Upper gastrointestinal barium examination in the AP (A) and oblique (B) projection.

Which of the following represents **the most accurate assessment** of the barium esophagram findings?

1. The barium esophagram shows diffuse esophageal dilation tapering near the gastro-esophageal junction
2. The barium esophagram shows extraluminal extension of administered oral contrast consistent with an esophageal tear
3. The barium esophagram shows gastric torsion
4. The barium esophagram shows normal findings
5. The barium esophagram shows sacculation suggesting scleroderma-related esophageal dysmotility

Correct!

1. The barium esophagram shows diffuse esophageal dilation tapering near the gastro-esophageal junction

The barium esophagram shows a diffusely dilated esophagus tapering to a normal caliber at the gastro-esophageal junction. While the esophagus is dilated, the appearance does not resemble the “sacculation” typical of scleroderma involvement of the esophagus; note that the distal esophagus is not dilated as is typical of scleroderma, but instead tapers to a non-distended appearance. No evidence of pneumomediastinum or extraluminal contrast extension to suggest esophageal injury is present. No features to suggest acute gastric torsion are present; the stomach is largely decompressed.

The patient continued to complain of shortness of breath.

At this point, which of the following represents the **most appropriate step** in this patient’s management?

1. ¹⁸FDG – PET
2. Bronchoscopy with transbronchial biopsy and possibly lymph node biopsy
3. Pulmonary function testing
4. Thoracic MR
5. Video-assisted thoracoscopic biopsy

Correct!

3. Pulmonary function testing

Thoracic MR and ¹⁸FDG-PET scanning would be of little use for evaluating this patient's persistent shortness of breath in light of the lack of actionable findings in the mediastinum, lung parenchyma, and pleural space at thoracic CT. Bronchoscopy with biopsy and video-assisted thoracoscopic biopsy are also not appropriate as no clear target for tissue sampling is evident.

The patient underwent pulmonary function testing, which showed mixed results, including significant restriction, with a forced vital capacity of 1.67 L (58% predicted) and a forced expiratory volume in one second of 1.06 L (50% predicted). The forced expiratory flow rate was only 49% predicted. The flow-volume loop suggested some inspiratory flow restriction.

At this point, which of the following represents the **most appropriate step** in this patient's management?

1. ⁶⁸Ga-citrate scintigraphy
2. Cervical mediastinoscopy
3. Repeat contrast-enhanced thoracic CT
4. Repeat thoracic CT performed with high-resolution technique
5. Upper endoscopy

Correct!

4. Repeat thoracic CT performed with high-resolution technique

Upper endoscopy is reasonable and will likely be performed at some point to evaluate the patient's suspected achalasia, but upper endoscopy is unlikely to assist in the evaluation of the patient's shortness of breath and pulmonary function testing abnormalities. A repeat thoracic CT using intravenous contrast material is unlikely to provide incremental information to what is already known. However, repeating the thoracic CT using high-resolution technique, which includes post-expiratory and prone imaging, may well be of benefit. As there is no target for tissue sampling, cervical mediastinoscopy is not indicated. Finally, ^{68}Ga -citrate scintigraphy, once occasionally used for the assessment of diffuse lung disease to distinguish pulmonary infection from other non-infectious causes of diffuse pulmonary disease, is now rarely employed for imaging the chest.

The patient underwent repeat enhanced thoracic CT (Figure 6). The study was not ordered as a high-resolution chest CT, however.

Figure 6. Axial thoracic CT displayed in lung windows shows an extremely dilated esophagus (e).

Which of the following findings on this thoracic CT scan **may explain** this patient's shortness of breath?

1. Acute pulmonary embolism with right ventricular strain
2. Extensive airway thickening
3. Multifocal ground-glass opacity
4. New centrilobular nodules
5. Tracheal narrowing

Correct!
5. Tracheal narrowing

The contrast-enhanced thoracic CT shows no evidence of right heart failure or acute pulmonary embolism. No evidence of ground-glass opacity is seen. The central airways do not show abnormal thickening. No evidence of centrilobular nodularity is present. However, the upper thoracic trachea appears significantly narrowed, apparently compressed between the dilated esophagus and great vessels and anterior chest wall (Figure 6). This narrowing continues to the level of the carina. This finding is consistent with tracheomalacia, not readily seen on the previous thoracic CT (Figure 4).

It was noted that, although the patient did not offer complaints of dysphagia voluntarily, when questioned she admitted to such (particularly, and not surprisingly, solids) and suffered from significant protein-calorie nutrition. The patient subsequently underwent upper endoscopy for evaluation of her esophageal abnormalities, with botulinum toxin injection, which showed narrowing of the distal esophagus but without intrinsic abnormality or extrinsic obstruction. No upper- to mid-esophageal motility was noted, with a hypertonic lower esophageal sphincter, consistent with achalasia. Shortly after the procedure, the patient suffered hypoxic respiratory failure requiring intubation. The patient was subsequently stabilized, and extubated 30 minutes later. However, about 20-30 minutes following extubation, she again developed shortness of breath with hypoxia, requiring re-intubation. She was again stabilized and subsequently extubated. She did well over most of the ensuing night, but again developed shortness of breath and hypoxic respiratory failure, this time managed successfully with bag-mask ventilation. Arterial blood gas analysis suggested acute-on-chronic hypercarbic respiratory failure during these events. Pulmonary medicine was consulted and felt her complaints and respiratory failure events were a combination of restriction resulting from left diaphragmatic paralysis following her sternotomy, her malnutrition, and tracheomalacia resulting from compression by a megaesophagus. Pulmonary medicine did not feel tracheal stenting was appropriate for the patient, and efforts should be directed towards relieving the airway obstruction by the enlarged esophagus, avoidance of sedative medications, and use of positive pressure ventilation as needed. A percutaneous gastrostomy tube was placed to improve the patient's nutrition prior to a planned Heller myotomy to address the patient's achalasia, as this was felt to be the inciting factor for both the patient's malnutrition and her tracheomalacia-induced respiratory failure. However, general surgery felt the patient's clinical status presented significant surgical risks and was reluctant to perform a surgical intervention to address the patient's achalasia. The patient was treated with repeat upper endoscopy with botulinum toxin injection.

Diagnosis: Tracheobronchomalacia due to achalasia

References

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