May 2018 Pulmonary Case of the Month

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History of Present Illness

A 70-year-old man presented with new anemia and heme-positive stools. Esophagogastroduodenoscopy was performed which revealed gastritis. Ascites developed and a chest x-ray noted a left pleural effusion. He was initially managed with weekly high-volume thoracentesis and paracentesis. He was referred to pulmonary medicine.

Past Medical History, Social History and Family History

He has a history of coronary artery disease having undergone coronary bypass grafting in 2016. He also has type 2 diabetes mellitus managed by diet and recently diagnosed orthostasis. He smokes about ½ pack of cigarettes per day but does not drink alcohol. He denies any inhalational exposures. He is Native American and works as a judge. There is no family history of any similar disorders.

Physical Examination

- No acute distress
- Slight bruise to left eye
- No lymphadenopathy
- Decreased breath sounds on left
- Protuberant distended abdomen
- Significant left leg edema
- Discoloration of a few nails

A point of contact ultrasound is performed (Figure 1).

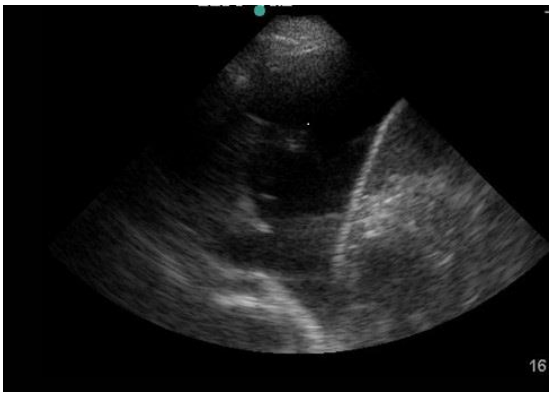

Figure 1. Image from the point of contact ultrasound.

What should be ***done next***?

1. Needle biopsy of pleural mass
2. Thoracentesis
3. Thoracic surgery consultation for video-assisted thorascopic surgery (VATS)
4. 1 and 3
5. All of the above

Correct!
2. Thoracentesis

The ultrasound shows a pleural effusion (Figure 2A). The diaphragm can be identified as the nearly vertical bright line (arrow). The dark space to the left of the diaphragm is the pleural effusion (star). The patient has no apparent contraindications and a thoracentesis is performed. The result fluid is shown in Figure 2B.

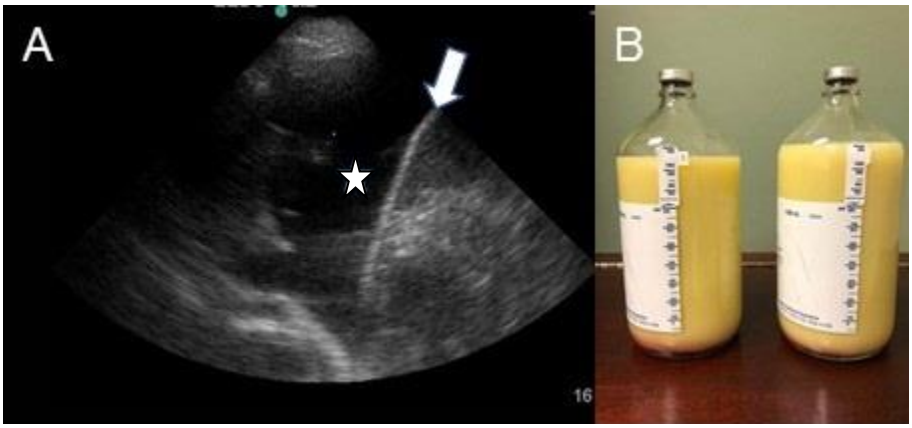

Figure 2. A: Point of contact ultrasound of the left pleural effusion showing the diaphragm (arrow) and pleural effusion (star). B: Fluid from thoracentesis.

What ***tests should be ordered*** on the pleural effusion?

1. Adenosine deaminase
2. pH
3. Triglycerides
4. 1 and
5. All of the above

Correct!
3. Triglycerides

The fluid has the characteristic appearance of a chylous effusion or chylothorax which results from an accumulation of chyle or lymph fluid in the pleural space. The appropriate test to diagnose a chylous effusion is a triglyceride level. Our patient's triglyceride level was markedly elevated at 967 mg/dL (normal usually < 110). The adenosine deaminase (ADA) is a test for tuberculosis which is not routinely ordered in the US because of the low incidence of tuberculosis. The pleural fluid pH can be useful in the diagnosis including empyema where it is usually below 7.2. The patient's course does not really fit with empyema but empyema on occasion can have the same milky appearance when it is packed with polymorphonuclear leukocytes.

Which of the following are ***true regarding chylous pleural effusions***?

1. About half are secondary to trauma (surgical, penetrating trauma)
2. Chylous effusions are common after coronary bypass grafting but usually resolve
3. May be secondary to an obstruction of the thoracic duct or elevated venous pressures
4. 1 and 3
5. All of the above

Correct!
4. 1 and 3

Chylothorax results when the flow of lymph from the abdomen is interrupted during its normal passage along the thoracic duct before draining into venous system usually at the subclavian veins (1). About half the cases are secondary to trauma where the thoracic duct leaks chyle and about half are due to medical conditions or are congenital. Obstruction of the thoracic duct from direct obstruction (mediastinal masses, mediastinitis, and radiation therapy) or elevated right-sided heart pressures essentially resulting in thoracic duct obstruction may cause a chylothorax. The thoracic duct runs posteriorly along the esophagus and is rarely injured during coronary artery bypass grafting. Most surgical injuries are secondary to esophageal, head and neck or aortic operations.

Based on the above pathophysiology which of the following may be used to diagnose the ***cause of the chylous effusion***?

1. Cardiac ultrasound (echocardiogram)
2. Lymphangiogram or lymphoscintigraphy
3. Thoracic CT scan
4. 1 and 3
5. All of the above

Correct!
5. All of the above

A thoracic CT scan to look for mediastinal masses and a cardiac ultrasound to look for a cause of elevated right-sided pressures are fairly easily performed and may be useful. If these fail to identify a source or a thoracic duct leak is suspected, two imaging techniques have been used in visualizing the lymphatic system in patients with chylothorax: conventional lymphangiography and lymphoscintigraphy (1). Lymphangiography typically employs bilateral pedal injection of contrast medium (lipiodol) while lymphoscintigraphy utilizes labeled colloids to generate morphological and functional imaging data about the lymphatic drainage. The role of these imaging modalities in the evaluation and management of patients with chylothorax remains poorly defined. In addition, these radiologic techniques may not be available at many medical centers.

A lymphangiogram was performed and was interpreted as normal (Figure 3).

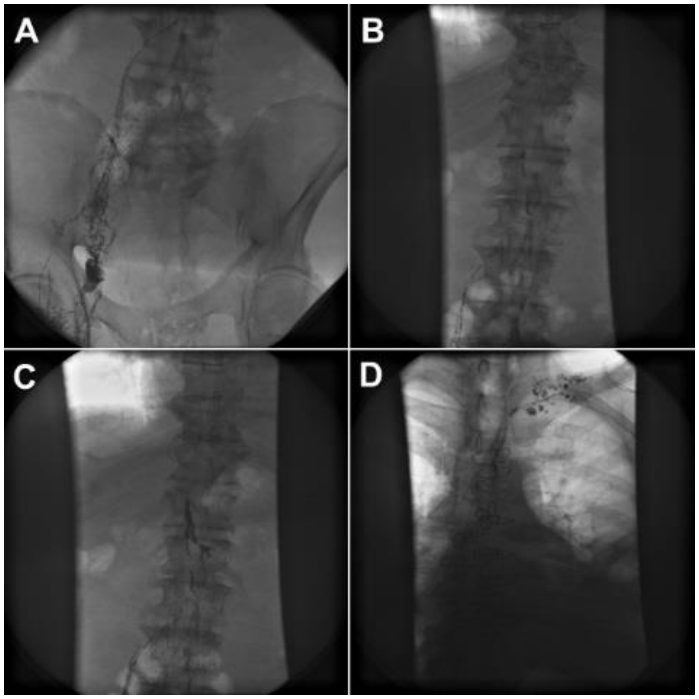

Figure 3. Representative images from the lymphangiogram. The images were interpreted as normal.

Which of the following ***should be done***?

1. Dermatology consult for yellow nails syndrome
2. PET scan
3. Review the cardiac ultrasound
4. 1 and 3
5. All of the above

Correct!
4. 1 and 3

The yellow nails syndrome is a rare disorder characterized by the triad of yellow and thickened nails, lymphedema and chylous pleural effusion (1). The pathogenesis of this rare syndrome remains poorly understood, but the evidence to date suggests that the lymphatic dysfunction observed is an acquired disorder. Yellow nail syndrome has been described in association with a variety of conditions including lymphoproliferative disorders, immunodeficiency states, connective tissue disorders, endocrinopathies, obstructive sleep apnea, Guillain–Barré syndrome, xanthogranulomatous pyelonephritis and tuberculosis. It seems likely that most of these associations are probably spurious.

A dermatology consult was ordered. The dermatologist thought the patient's nails were more consistent with onychomycosis rather than yellow nail syndrome (2).

The cardiac ultrasound was reviewed and showed slow ventricular filling and abnormal heart muscle relaxation. The ventricular and septal walls were thickened. Consistent with the diagnosis of cardiomyopathy the NT-pro brain natriuretic peptide was markedly elevated at 23,000 pg/ml (normal < 300). An abdominal fat biopsy showed increased Congo red staining and apple green birefringence under polarizing microscopy (Figure 4).

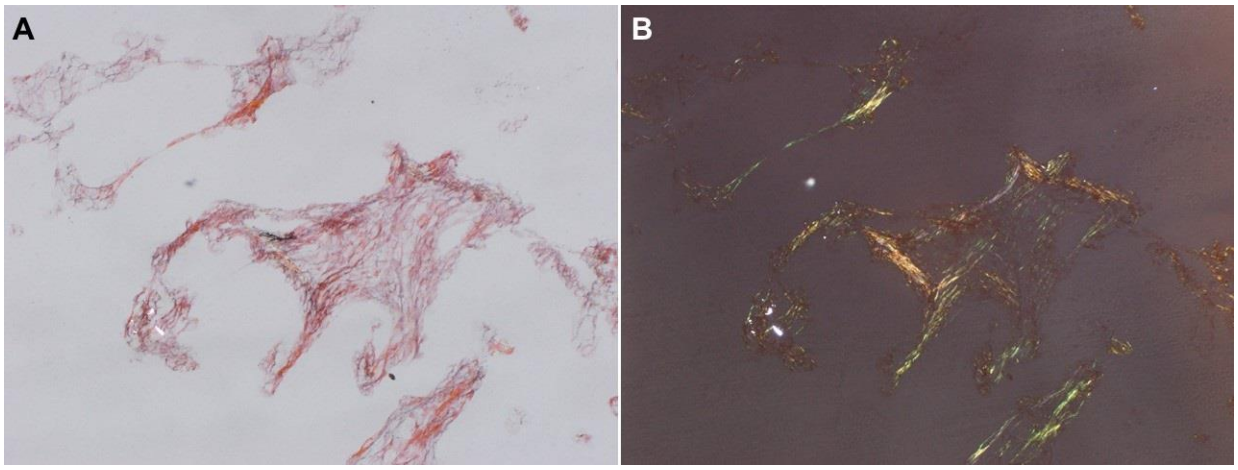

Figure 4. Abdominal fat showing (A) increased Congo red staining and (B) birefringerence under polarizing microscopy.

Which of the following disorders are associated with amyloidosis?

1. Bronchiectasis
2. Multiple myeloma
3. Tuberculosis
4. 1 and 3
5. All of the above

Correct!
5. All of the above

A variety of chronic inflammatory diseases have been associated with secondary amyloidosis. Chest diseases include bronchiectasis, tuberculosis and cystic fibrosis. However, a common cause is multiple myeloma (3). Our patient had an elevated kappa free light chain and bone marrow biopsy confirmed the presence of increased plasma cells consistent with multiple myeloma. The patient is currently undergoing treatment for his multiple myeloma. His chylothorax is being managed conservatively with the substitution of dietary fat by medium-chain triglycerides which are absorbed directly into the portal circulation and by octreotide.

References

1. Ryu JH, Tomassetti S, Maldonado F. Update on uncommon pleural effusions. Respirology. 2011 Feb;16(2):238-43. [CrossRef] [PubMed]
2. Wesselius LJ. Medical image of the week: yellow nail syndrome. Southwest J Pulm Crit Care. 2017;15(4):230-1. [CrossRef]
3. Lawler PR, Bergmark BA, Laubach JP, Lakdawala NK. Having a heavy heart: approaches to infiltrative cardiomyopathy. Circulation. 2014 Apr 22;129(16):1703-11. [CrossRef] [PubMed]