CLINICAL IMAGE

A RARE CAUSE OF DYSPNEA

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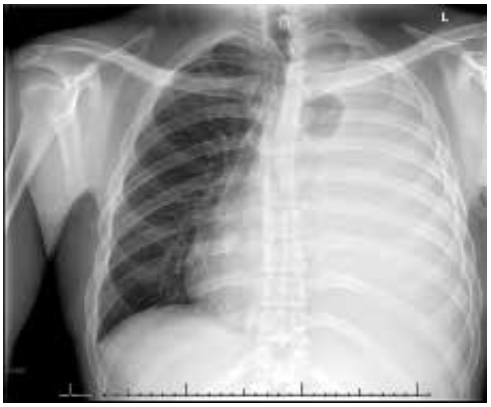
INTRODUCTION

A 38-year-old male with a past medical history of necrotizing pancreatitis and splenic vein thrombosis, diagnosed six months prior, presents to the emergency department with a chief complaint of dyspnea of three days duration. The patient experienced sudden onset difficulty breathing while working on his truck at home, with associated palpitations, both resolving after giving minutes of rest. After three days, the patient reported mild improvement in his dyspnea after using his significant others albuterol breathing treatment. However, due to persistent discomfort, the patient reported to the emergency department, where he underwent a chest x-ray and computed tomography (CT) of the chest with IV contrast. The patient denied any fevers or cough at home. The patient works part-time on the weekends at a turkey processing farm, with appropriate use of protective equipment, including barrier garments and a mask. He smokes six–seven cigarettes a day for the last ten years, admits to drinking alcohol occasionally and denies any other drug abuse, except for marijuana. The patient has a past medical history of liver cirrhosis secondary to alcohol abuse and surgical history of appendectomy. He denies any URI symptoms, chest pain, abdominal pain, nausea or vomiting, headaches or vision changes. He denies any history of travel and reports the only medication he takes is Motrin® for pain relief.

FIGURE 1:

The patient's chest x-ray.



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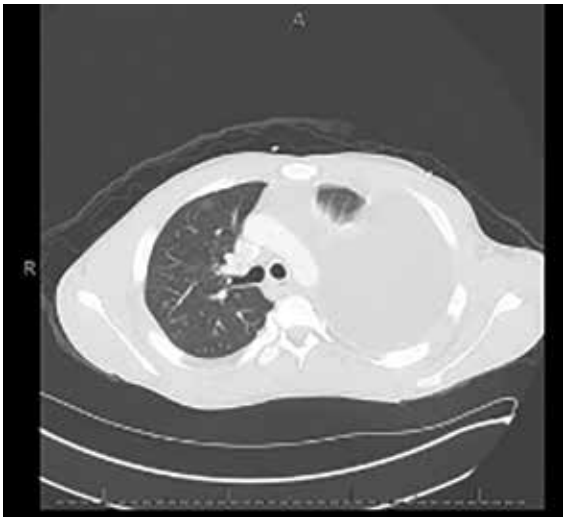
 Print ISSN: 1877-573X
DOI: 10.33181/13026

FIGURE 2:

The patient's computed tomography (CT) of the chest with IV contrast.



QUESTIONS:

1. Based on the patient's past medical history, clinical presentation and imaging, which of the following is the most likely underlying etiology for the pleural effusion?

A. Congestive heart failure

B. Pancreaticopleural fistula (PPF)

C. Pneumonia

D. Pulmonary embolism

2. What is the best imaging study of choice for diagnosing this disease?

A. Endoscopic retrograde cholangiopancreatography (ERCP)

B. Magnetic resonance cholangiopancreatography (MRCP)

C. Lung biopsy

D. Computed tomography (CT) of chest-abdomen-pelvis

3. What is the next best intervention to provide symptomatic relief for this patient's pleural effusion?

A. 40mg IV Lasix

B. Thoracentesis

C. CT guided lung biopsy

D. IV antibiotics

ANSWERS:

1. Based on the patient's past medical history, clinical presentation and imaging, which is the most likely underlying etiology for this patient's pleural effusion?

Correct answer:
B) Pancreaticopleural fistula (PPF)

PPF is a rare diagnosis that occurs in approximately 0.4% of patients with pancreatitis.[1] It is caused by chronic pancreatic inflammation in which pancreatic secretions drain directly into the pleural cavity through an abnormal connection between the pancreatic duct and the pleural space, causing an amylase-rich pleural effusion.[1] It usually requires a high index of suspicion as patients typically present with pulmonary symptoms such as dyspnea caused by the pleural effusion rather than abdominal symptoms caused by pancreatitis.[1] Other than resulting from a chronic inflammatory process, PPF can also occur from acute inflammation or traumatic or iatrogenic rupture of the pancreatic duct.[1] Congestive heart failure is a progressive disease in which the heart's function as a pump is inadequate to meet the body's needs. It is a less common finding in young individuals, especially with no prior cardiac history. CXR findings would be consistent with cardiomegaly, cephalization and Kerley lines. Pneumonia and pulmonary embolism can be common causes of acute dyspnea. Pneumonia could be a likely diagnosis in this patient with a history of exposure to birds; however, the rest of the clinical findings and imaging do not support this choice. Although a CXR would not be indicative for a PE, a CTPE was performed on this patient, revealing no blood clots in the pulmonary vasculature, making PPF the correct answer choice.

2. What is the best imaging study of choice for diagnosing this disease?

Correct Answer:
B) Magnetic resonance cholangiopancreatography (MRCP)

PPF is a rare complication of chronic pancreatitis consequent to disruption of the pancreatic duct. Although CT is the standard modality for evaluating acute pancreatitis and its complications, such as pseudocysts, it is not sensitive in assessing the anatomy and morphology of the fistulous tracts.[2] ERCP has emerged as both the diagnostic and therapeutic option in select patients with PPF; however, MRCP still remains the radiological investigation of choice as it not only helps in diagnosing the presence and site of the fistulous tract but also helps determine management options.[3]

3. What is the next best intervention to provide symptomatic relief for this patient's pleural effusion?

Correct answer:
B) Thoracentesis

Massive pleural effusion caused by PPF is a very rare complication of pancreatitis, most commonly associated with alcoholic chronic pancreatitis.[2] In cases of a PPF resulting in a hydrothorax, approximately 75% occur on the left side, although they may also be right-sided or bilateral.[2] Thoracentesis should be performed in patients with a pleural effusion. It not only helps with diagnosis by obtaining fluid for biochemistry, but it also alleviates pulmonary symptoms such as dyspnea.[2] Pleural effusions associated with PPF are usually exudative in nature and although there is no cutoff level to establish a diagnosis, pleural amylase is usually >1000 U/L.[4] CT guided lung biopsy would aid in diagnoses of malignancy related pleural effusion and also would not provide any type of symptomatic relief to this patient. While IV antibiotics and IV Lasix are potential adjunctive treatments, they would not provide immediate relief of the pulmonary symptoms this patient is experiencing.

DISCUSSION

Pancreaticopleural fistulas are a rare complication of acute or chronic pancreatitis and are most commonly associated with alcoholic pancreatitis.[2] They can also occur due to gallstone pancreatitis, idiopathic pancreatitis, trauma or pseudocysts. They occur in less than 1% of patients with acute pancreatitis, 0.4% of patients with chronic pancreatitis and 4.5% of patients with pseudocyst.[1] Chronic pancreatitis is a progressive and irreversible inflammatory process characterized by the transformation of pancreatic parenchyma into fibrotic tissue.[5] Additionally, chronic alcohol consumption can cause acute focal inflammation of the pancreatic ducts and form protein plugs, which can subsequently cause temporary blockage of the small duct branches and lead to main pancreatic duct dilation.[6] Due to this inflammation, an abnormal connection is known as a fistula, which sometimes forms between the pancreatic duct and the pleural space, causing pancreatic secretions to traverse into the pleura leading to a pleural effusion that is rich in amylase.[5]

The pathophysiology of PPF consists of ductal disruption on the posterior surface, which then results in the thoracic fluid collection as the fluid spreads retroperitoneally through pathways of least resistance at the aortic or esophageal hiatus.[2] For this reason, patients with PPF are more likely to present with pulmonary symptoms related to the pleural effusion rather than abdominal symptoms related to pancreatitis leading to a delay in diagnosis.[7] Many patients may even undergo extensive lung investigation before the pancreas is identified as the dysfunction's primary organ. The diagnosis is usually performed by thoracentesis after a chest radiography, with laboratory findings of elevated amylase levels, generally above 1,000 U/L, although no diagnostic level has been recognized.[7] Once the diagnosis has been established, imaging is used as an adjunct to characterize the anatomy and location of the fistulous tracts. Even though CT scan is the preferred choice for the evaluation of pancreatitis, for assessment of the fistulous tracts, MRCP is deemed superior.[3] Additionally, ERCP was previously preferred due to it being diagnostic and therapeutic in nature; however, MRCP is non-invasive when compared to ERCP and it helps with visualizing the pancreatic duct beyond simple strictures, making it the current imaging modality of choice in patients with PPF.[7]

Regarding the treatment of PPF, no treatment modality has been deemed superior to others. PPF can be treated medically, endoscopically or surgically, based on individual patient presentation. Immediate thoracentesis with chest tube placement to drain the recurrent pleural effusion is often the first step and provides immediate relief of symptoms. Endoscopic treatment with ERCP stenting to the pancreatic duct has also been shown

to be successful, with the main goals of blocking the abnormal connection of the pancreatic duct with pleura as well as to keep the duct open for the pancreatic secretions to flow down the path of least resistance into the duodenum.[1] Obvious complication includes anatomical disruption of the duct and recurrent fluid accumulation. If and when medical and ERCP treatment fails, surgical treatment is the best resort. The main principle of surgical treatment is to form a pancreatic enteric connection to achieve adequate drainage of the pancreatic sections with or without pancreatic resection.[1] The most common surgery reported in the treatment of PPF is distal pancreatectomy with pancreaticojejunostomy.[1] King *et al*. reported a 94% success rate for surgical treatment compared to a 31% success rate for medical treatment.[8] The same study also reported that medical treatment takes 50% more time than the time required for surgical recovery and that 70% of the reported surgical complications occurred in patients treated initially with medical management. Generally, the rule seems to be to treat normal or mildly dilated pancreatic duct with the trial of medical therapy, those with ductal disruptions or strictures to undergo endoscopy and reserve the surgical option for those with a complete ductal obstruction or if stenting is impossible based on image findings.[9]

CONCLUSION

Pancreatic pleural fistulas remain to be rare, however, they still present as a significant problem in patients with pancreatic disease. Dyspnea is a symptom that clinicians often encounter in their practice and many usually place cardiopulmonary etiology at the top of the differential.[10] Nevertheless, it is always important to recognize the patient's history, rather than relying on their symptoms alone. The single most important diagnostic procedure is a pleural tap to determine the level of pleural fluid amylase.[9] ERCP used to be the preferred investigative technique for confirming the diagnosis; however, MRCP is now deemed superior due to its noninvasive nature. For treatment, as always, deciding which route to proceed is different based on individual patient presentation and a thorough risk and benefit assessment needs to take place between the physician and the patient. Strict follow-up with primary care physicians to reduce complications such as PPF should be encouraged for pancreatic disease patients.

AUTHOR DISCLOSURES: No relevant financial affiliations or conflicts of interest. If the authors used any personal details or images of patients or research subjects, written permission or consent from the patient has been obtained. This work was not supported by any outside funding.

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