


Bridging the Divisum: A Rare Case of Pancreaticopleural Fistula in the Setting of Complete Pancreatic Divisum Treated Endoscopically

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Abstract

A pancreaticopleural fistula (PPF) is a rare complication of chronic pancreatitis that occurs either due to a pancreatic duct disruption or a pseudocyst extension. A pancreatic divisum, on the other hand, is a common anatomic variant of the pancreas that is rarely symptomatic. We describe a case of recurrent pleural effusion in a patient with a history of chronic pancreatitis. Investigations revealed the presence of a PPF and a concomitant complete pancreatic divisum. There was resolution of the pleural effusion on endoscopic therapy. This is the fourth reported case of a PPF in the setting of complete pancreatic divisum and the first reported case in a middle-aged female.

Keywords

pancreas, biliary tract, pancreatic divisum, endoscopy

Introduction

Pancreaticopleural fistulas (PPFs) are rare complications of chronic pancreatitis.¹ Pleural effusions due to PPFs occur in <1% of patients and are seen in men aged between 40 and 50 years with a history of alcohol consumption.² Common clinical symptoms include dyspnea, cough, and chest pain.¹ Abdominal pain is rare, resulting in a diagnostic dilemma.³

Case Report

Our patient is a 51-year-old female with a history of chronic pancreatitis due to alcohol consumption complicated by a pseudocyst who presented to the emergency department with shortness of breath for over a week.

She had been experiencing nonpositional, nonexertional progressive shortness of breath for over a week. She denied fevers, chills, chest pain, cough, and urinary or bowel complaints. Her vitals revealed a pulse of 116 beats per minute and a respiratory rate of 25 breaths per minute. Physical examination revealed dullness to percussion over the right lung field accompanied by decreased breath sounds. Aside from mild epigastric tenderness, her abdominal examination was benign. The remainder of her examination was unremarkable.

Laboratory tests on presentation were significant for a white blood cell count of 14900/ μ L, alkaline phosphatase level of 131 IU/L, serum amylase of 360 IU/L, and serum lipase of 113 IU/L. A chest X-ray revealed a moderate right-sided pleural effusion (Figure 1). Computed tomography of the abdomen showed diffuse atrophy of the pancreatic parenchyma but no evidence of active pancreatitis. She was admitted for management of her pleural effusion.

A thoracentesis on admission revealed a non-hemorrhagic aspirate. Analysis revealed a lactate dehydrogenase level of 617 IU/L (upper limit of normal serum lactate dehydrogenase of 222) and a pleural protein level of 2.4 g/dL (serum protein of 6.4). Per Lights criteria, the aspirate was exudative. Further analysis revealed a fluid lipase level of 23 088

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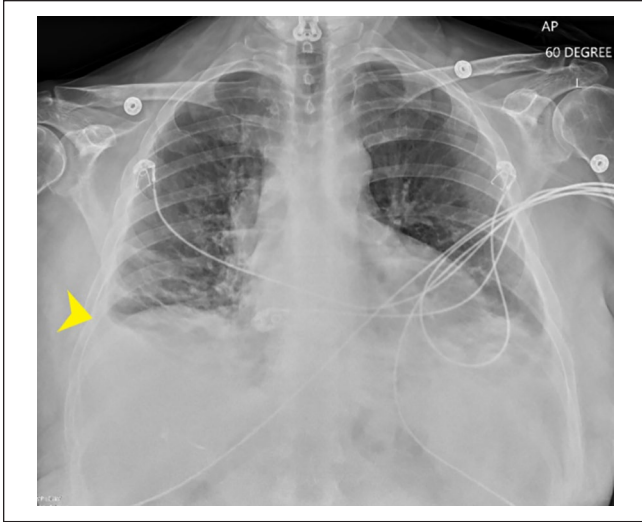


Figure 1. Chest X-ray demonstrating right-sided pleural effusion (yellow arrow head).

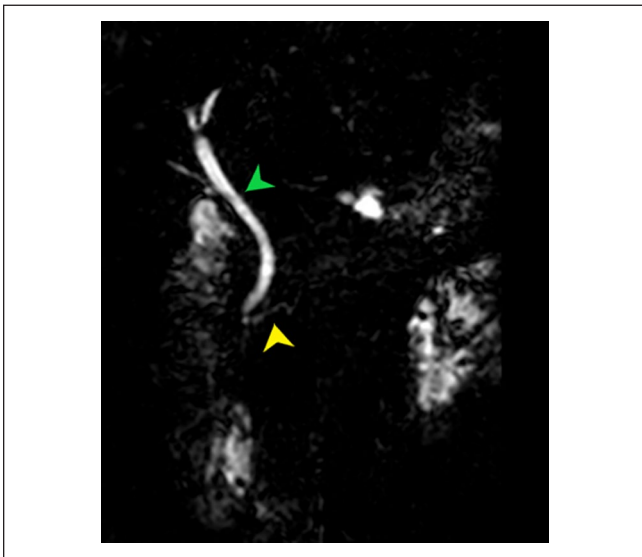


Figure 2. Magnetic resonance pancreatography demonstrating the common bile duct (green arrow head) joining with the ventral pancreatic (yellow arrow head) duct to empty into the duodenum.

U/dL and an amylase level of 10973 U/dL. A diagnosis of a PPF was made and gastroenterology was consulted.

A magnetic resonance pancreatography (MRCP) was obtained, which was unable to identify a PPF. However, it did reveal a complete pancreatic divisum (Figures 2 and 3), which had not been reported prior. Given high suspicion for a PPF, the patient underwent an endoscopic retrograde cholangiopancreatography (ERCP) with contrast that revealed contrast leakage tracking upwards from the dorsal pancreatic duct confirming a PPF. The absence of communication between the dorsal and ventral ducts confirmed complete

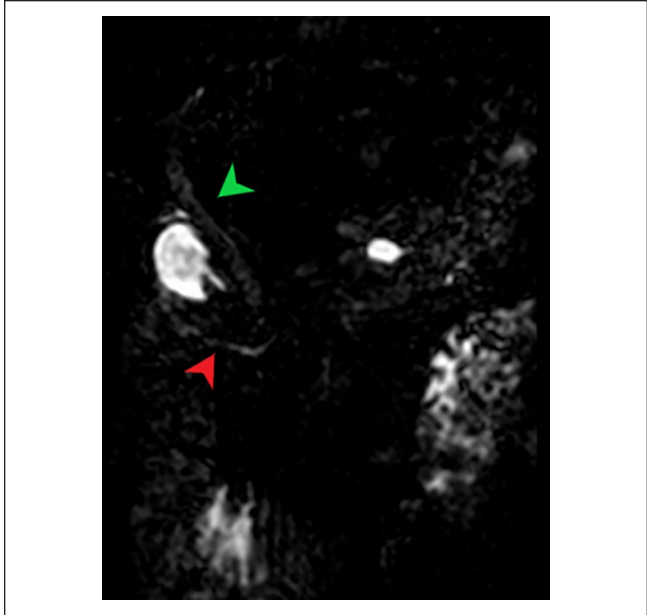


Figure 3. Magnetic resonance pancreatography demonstrating dorsal pancreatic duct (red arrow head) traversing anterior to the common bile duct (green arrow head) and draining into minor papilla depicting complete pancreatic divisum.

pancreatic divisum. A 2-mm dorsal pancreatic sphincterotomy was made with a monofilament traction sphincterotome using electrocautery. A 5 Fr by 9 cm pancreatic duct plastic stent was placed in the minor papilla (Figure 4).

Post procedure computed tomography confirmed appropriate placement of the pancreatic stent (Figure 5). She reported improvement in her dyspnea. Repeated chest X-ray at a 2-month follow-up revealed complete resolution of her pleural effusion (Figure 6).

Search Strategy

A PubMed search was conducted with the terms “Pancreaticopleural fistula” AND “Pancreas Divisum” with the filters being set to “full text” and language being set to “English.” Initial search result revealed 1 article.⁴ A separate PubMed search was then conducted with mesh term “Pancreaticopleural fistula” with filters being set to full text and English language, which yielded 120 results. After careful filtering of the articles based on abstract summary and article headings 118 articles were excluded. Thus, 2 more articles were added.^{5,6} A Google Scholar search with the term “Pancreaticopleural fistula” yielded 1 more article.⁷ Clinical features of the 4 other cases have been summarized in Table 1.

Discussion

It is theorized that PPFs occur from pancreatic ductal rupture. An anterior rupture results in pancreatic ascites whereas a

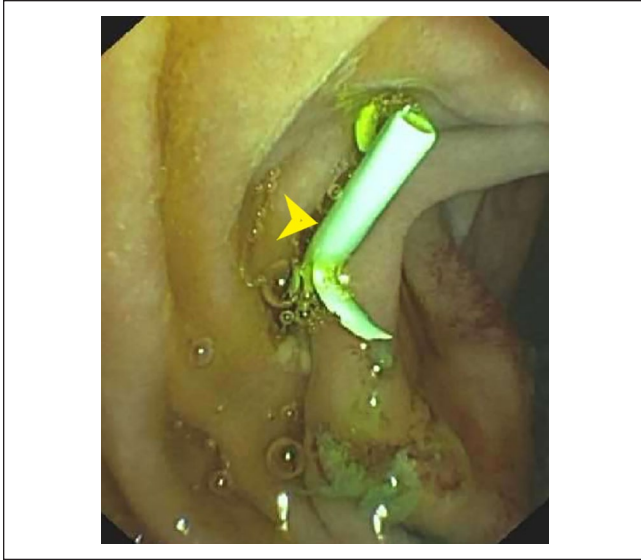


Figure 4. Endoscopic imaging shows the pancreatic duct stent (yellow arrow head) in the minor papilla.

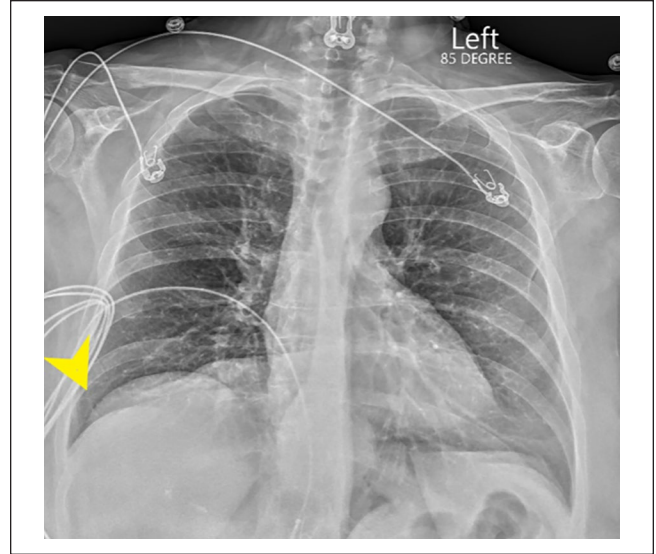


Figure 6. Chest X-ray demonstrating resolution of pleural effusion (yellow arrow head).

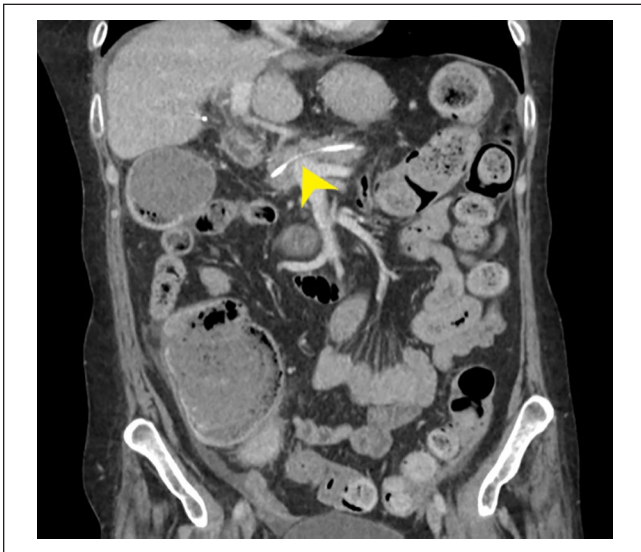


Figure 5. Computed tomography scan of the abdomen revealing placement of pancreatic duct stent (yellow arrow head) in the dorsal pancreatic duct.

posterior rupture results in pseudocyst or fistula formation.^{2,8} PPFs occur in about 3% to 7% of patients with pancreatitis.²

The classic presentation is of a middle-aged male with a history of alcohol consumption who complains of respiratory symptoms.^{2,3,8} Ali and colleagues¹ described that shortness of breath accounted for almost two thirds of the presenting complaints whereas abdominal pain accounted for less than 29%. Although our case fit the typical age, symptom description, and radiological presentation, it differed in the fact that

our patient was female, whereas more than 80% of the literature cases are of men.^{1,3,9-12}

Regarding laboratory testing, our review revealed that the hallmark of a PPF lies in the pleural fluid amylase level.² Pleural fluid amylase levels are severely elevated, often more than 10 000 IU/L with comparatively modest elevations in serum amylase levels.^{8,13,14} Our patient's pleural fluid amylase of 10 973 U/dL and serum amylase of 260 IU/L was in line with the reported literature.

While MRCP is the diagnostic imaging of choice, it is not entirely sensitive. It has been reported that MRCP alone missed 20% of PPFs.¹ If suspicion of a PPF persists despite a negative MRCP, the next best step is an ERCP.^{8,14} Given the strong clinical suspicion of a fistula despite a negative MRCP we opted to proceed with an ERCP.

Management of a PPF has evolved over the years. Previously, management was conservative followed by a surgery after 2 to 4 weeks if symptoms persisted.¹³ Nowadays, management is guided by findings from MRCP or ERCP.^{8,14} If the pancreatic duct is mildly dilated and without strictures, the initial treatment is medical therapy via thoracentesis and somatostatin.³ The presence of ductal disruption in the pancreatic body or the presence of strictures warrants using procedures such as ERCP-guided stent placement, endoscopic ultrasound-guided rendezvous ERCP, or a nasopancreatic drainage with stenting.^{3,12,15}

Contraindications and complications to endoscopic therapy require surgical management.¹ The ductal disruption seen via ERCP in our patient led to a minor papilla sphincterotomy with a dorsal pancreatic duct stent placement. This resulted in resolution of the pleural effusion.

Table 1. Clinical summaries of Case reports involving Pancreaticopleural fistula and Pancreas divisum.

| S no. | Study | Year | Age (years) | Gender | Initial presenting symptom | Side involved |
|-------|----------------------------------|------|-------------|--------|----------------------------|---------------|
| 1 | Nacoti et al ⁶ | 2006 | 18 months | Male | Shortness of breath | Right lung |
| 2 | Park and El-dika ⁵ | 2013 | 31 years | Female | Epigastric pain | Left lung |
| 3 | Vanderbruggen et al ⁴ | 2019 | 47 years | Male | Shortness of breath | Left Lung |
| 4 | Sarkar and Jagroop ⁷ | 2020 | 52 years | Male | Abdominal pain | Left lung |

Another key finding in our patient was the presence of a complete pancreatic divisum, which is an embryological defect in the fusion of the dorsal or ventral pancreatic ducts present in 5% to 10% of the population.^{16,17} This results in pancreatic fluid being diverted to the minor pancreatic duct leading to an obstruction in outflow, predisposing to pancreatitis. Although typically asymptomatic, it has been reported to increase the risk of pancreatitis.¹⁸ It is likely that our patient's anatomic predisposition increased her risk for a PPF as has been noted in literature.^{4,6,7,19}

To conclude, pleural effusions due to PPF can be especially difficult to diagnose.⁴ Suspicion should rise when there is persistent pleural effusion despite adequate treatment for common causes, such as heart failure or pneumonia. The presence of a preexisting pancreatic anomaly such as pancreatic divisum should strengthen clinical suspicion. Given the rarity of the condition, more literature would be necessary to establish a causal association between pancreatic divisum and the development of PPF. To our knowledge, this is the fifth reported case of a PPF in the setting of pancreatic divisum and the first case reported in a middle-aged female.⁴⁻⁷

Authors' Note

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Author Contributions

Rachel Peterson: Collection of data, literature review, drafting of manuscript, editing of images, and final revision.

Samiran Mukherjee: Conceptualization, collection of data, literature review and drafting of manuscript, and supervision of draft.

Prateek Harne: Interpretation of literature, drafting of manuscript, and critical review.

Sahir Quraeshi: Acquisition of data, drafting of manuscript, and critical review.

Anuj Sharma: Conceptualization, supervision, and critical review of manuscript.

Umair Masood: Supervision and critical review of manuscript.

Divey Manocha: Mentoring, supervision, and critical review of manuscript.

Article guarantor: Samiran Mukherjee.

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Ethics Approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed Consent

Verbal informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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