An Unusual Cause for an Acute Massive Pleural Effusion

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Abstract
Although pleural effusion is a well-known complication of acute pancreatitis, its occurrence is much rare in chronic pancreatitis. Formation of pancreatic pleural fistula (PPF) is a rare complication of chronic pancreatitis which may give rise to a massive symptomatic pleural effusion. Patients often present predominantly with pulmonary symptoms, resulting in a delay in diagnosis of the pancreatic pathology. We report a 16-year-old boy with a past history of intermittent abdominal pain who presented with progressive dypnoea and left sided pleuritic chest pain. The chest radiography revealed a massive left sided pleural effusion. Biochemical analysis of pleural fluid showed an exudative effusion with very high amylase level, consistent with a pancreatic pleural effusion. Computed tomography and magnetic resonance cholangiopancreatography revealed chronic pancreatitis complicated with a pancreatic pseudocyst in relation to the tail of the pancreas, which had ruptured into the left pleural cavity causing a massive pleural effusion. The patient made a good recovery with conservative medical management. This case highlights the importance of suspecting pancreatic disease as an unusual, but important cause of massive pleural effusion in the appropriate clinical setting

Key words: *massive pleural effusion, pancreatitis, pseudocyst*

Introduction

Pleural effusion is a common respiratory condition with an array of differential diagnosis. However, majority of pleural effusions are associated with pulmonary infections or malignancy. Rarely extra thoracic diseases such as hepatic or subdiaphragmatic abscesses, acute or chronic pancreatitis can also be complicated with pleural effusion. Among pancreatic diseases, acute pancreatitis is well known to be complicated with pleural effusion in approximately 4-17% of patients (1). On the contrary, pleural effusion in chronic pancreatitis is rare, and usually occur secondary to pancreaticopleural fistula (PPF)which is an extremely rare diagnosis seen in 0.4% of patients with pancreatitis (2,3). A PPF may be formed as a result of either leakage in an incompletely formed pseudocyst, a rupture of a mature one, or rarely due to direct pancreatic duct leak. (4,5) The fistulous tracts form through the esophageal or aortic diaphragmatic orifices or less commonly trans diaphragmatically (6) and give rise to massive recurrent pleural effusions or mediastinal fluid collections. However, the clinical manifestations are often misleading as patients typically present with pleuropulmonary symptoms with absent or minimal abdominal symptoms, which often leads to delayed diagnosis (5). Here we report a case of a young male who presented with a massive left sided effusion as the sole manifestation, due to ruptured pancreatic pseudocyst. Although abdominal symptoms were absent at presentation, the background history of intermittent abdominal pain led to the ultimate diagnosis of pancreatic pleural effusion.

Case report

A 16-year old boy presented to the respiratory unit of National Hospital Kandy with a 3-week history of left sided pleuritic type chest pain, shortness of breath on exertion, dry cough and constitutional symptoms. His symptoms worsened over the preceding 2 days, accompanied by high grade intermittent fever. He complained of significant loss of appetite and a weight loss of 2kg during the period he was symptomatic but there was no evening pyrexia, night sweats or a contact history of tuberculosis. His systemic review was significant for a chronic intermittent abdominal pain for 5 years which was mainly epigastric with radiation to the back and partially relieved by bending forward. The patient had sought medical advice several times from local health

care institutes but each time was treated symptomatically. He was a student and did not consume alcohol. His family history was not significant.

The chest x-ray done at the local hospital revealed a massive left side pleural effusion with mediastinal shift to the right side. (figure 1) An intercostal drainage tube was promptly inserted and the patient was referred to National Hospital Kandy for further management.

On examination, he was ill looking, febrile (101F), not pale or icteric. The vital signs were as follows; heart rate 104 bpm, respiratory rate 24 cycles/min, blood pressure 110/70mmHg and room air saturation 94%. A left sided intercostal drainage tube was in-situ which drained brownish fluid. His trachea was in the midline with a stony dull percussion note and reduced breath sounds in the left mid and lower zones. Abdominal examination revealed severe tenderness and guarding in epigastric and left hypochondriac regions. There were no palpable abdominal masses, hepatosplenomegaly or intra-abdominal free fluid. Cardiovascular and neurological system examination revealed no abnormalities.

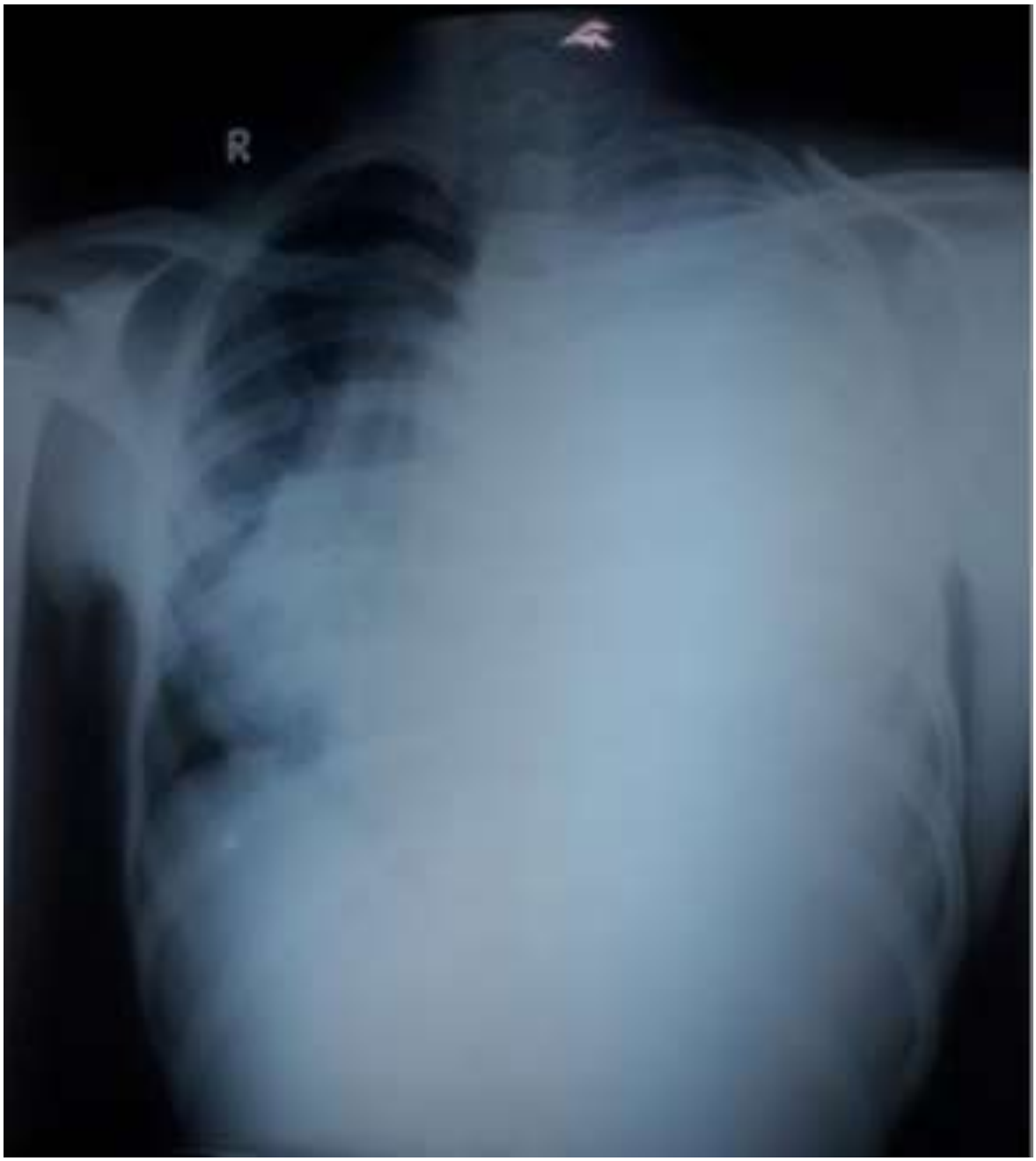

Figure 1 – chest radiography showing a left sided massive pleural effusion with mediastinal shift to the right side.

The laboratory investigations revealed a neutrophil leukocytosis (total white cell count of 14,000 cells/mm^3 with 81% neutrophils), a raised C-reactive protein level of 276mg/l and a raised erythrocyte sedimentation rate of 75mm in the first hour. The chest radiography revealed a large pleural effusion on the left side with an intercostal drainage tube in situ. Renal and liver function tests were normal. Sputum for gram stain, pyogenic culture, acid fast bacilli and XpertMTB were negative. Blood cultures did not reveal any growth. With the given history of chronic abdominal pain and left hypochondriac tenderness, underlying pancreatic pathology was strongly suspected. Ensuing investigations revealed a serum amylase level of 363U/L (30-110) and normal fasting blood glucose level. Pleural fluid analysis revealed brown colour fluid which was neutrophilic exudative (Pleural fluid protein 3.5g/dl, total white cell count of 10,400 cells/ mm^3 with 96% neutrophils) with pleural fluid amylase level of >12,000U/L. Pleural fluid for pyogenic culture, acid-fast smear and culture, Xpert/MTB were negative. Fiberoptic thoracoscopy revealed inflamed visceral, parietal and diaphragmatic pleurae with multiple adhesions and loculations in the pleural cavity with thick, gelatinous pleural fluid. Contrast enhanced computed tomography (CECT) scan of the chest and abdomen revealed features of chronic pancreatitis with an atrophied pancreatic gland complicated with a pancreatic pseudocyst which had ruptured into the left pleural cavity causing a massive pleural effusion. Multiple gas loculi were present in the pleural effusion which suggested superadded infection and there was near total collapse of the, left lung. (Figure 1)

A diagnosis of pancreatic pleural effusion secondary to PPF was made based on the presence of ruptured pseudocyst and high amylase in pleural fluid. He was referred for gastrointestinal surgical unit for subsequent management and commenced on intravenous antibiotics, anagesics, pancreatic rest, subcutaneous octreotide therapy and pancreatic enzyme replacement therapy. The patient improved clinically, with gradual reduction in the chest drainage after 48 hours. He subsequently underwent Magnetic resonance cholangiopancreaticography (MRCP) which revealed a 7.5cm x 6.5cm size cystic lesion in relation to the tail of the pancreas with duct dilatation involving the head, body and tail, with a short segmental luminal narrowing at the ampulla of Vater representing a benign stricture. No evidence of gall stones or congenital pancreatic anomalies were noted. (Figure 2A and 2B). The serum triglyceride level, serum calcium and immunoglobulin G levels, Anti-nuclear antibody (ANA) and rheumatoid factor (RF) were negative. His family history was not suggestive of hereditary pancreatitis or cystic fibrosis. In the absence of any other discernible aetiology, the benign stricture noted at the ampulla of Vater was considered as the most likely cause for chronic pancreatitis. However genetic studies to assess mutations of the SPINK 1 gene and CFTR gene were not performed due to unavailability and financial constraints.

The patient made a complete recovery with conservative management alone. Therefore, it was decided to follow up the patient closely with a plan to proceed with surgical interventions if the patient develops a relapse of the disease.

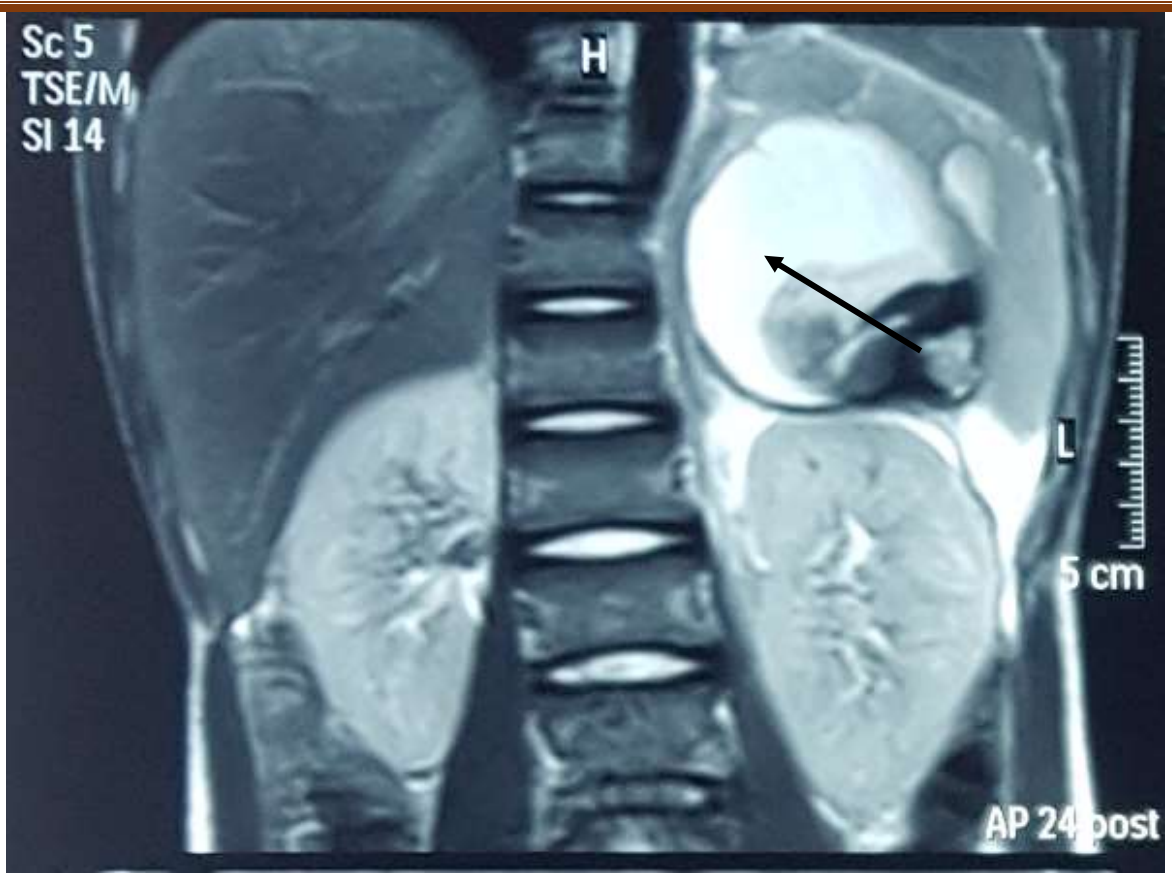

Figure 2A – MRCP showing the pancreatic pseudocyst.(arrow)

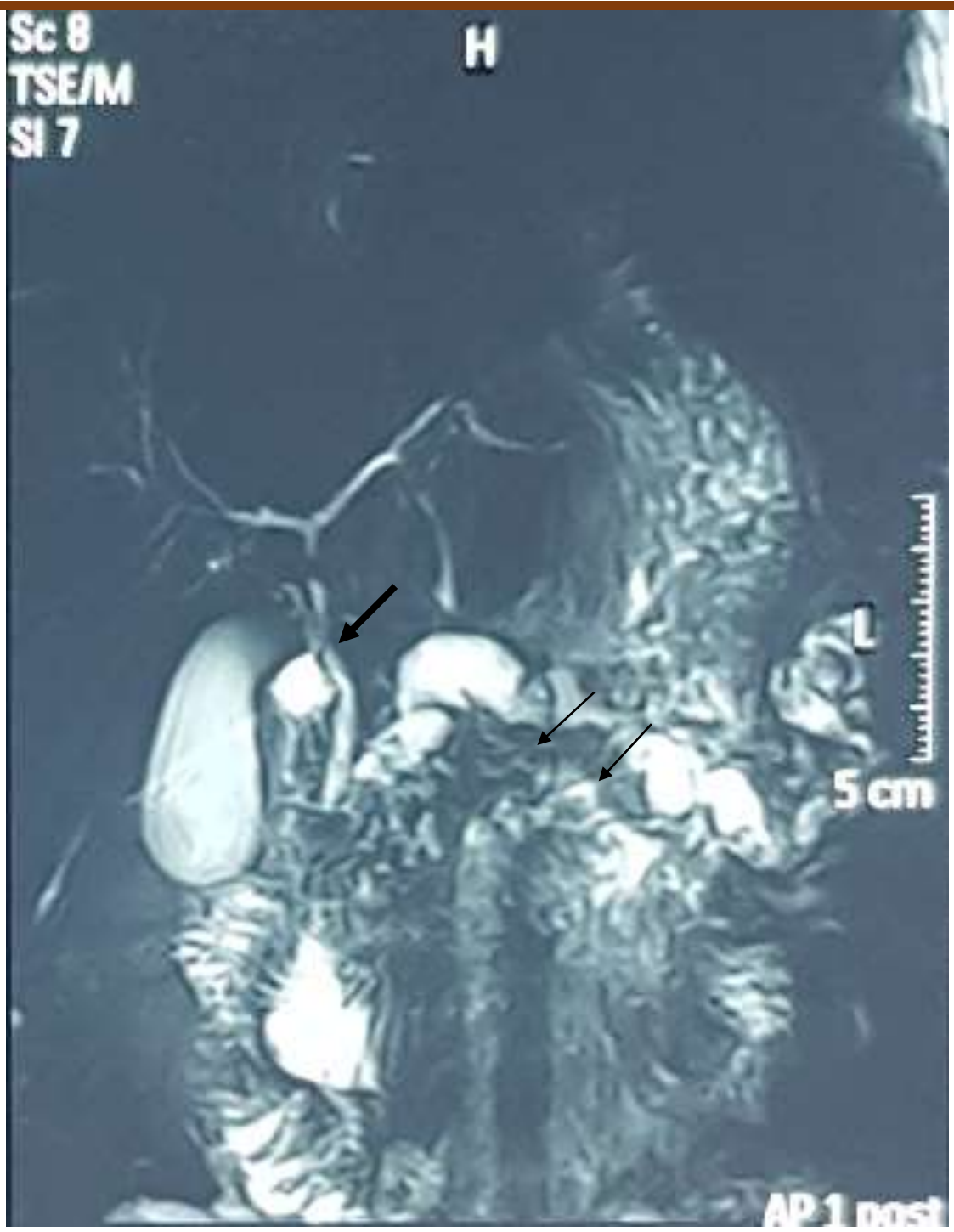

Figure 2 B - MRCP showing pancreatic duct dilatation involving the head (thick arrow), body and tail (thin arrow).

Discussion

A pancreatic pseudocyst is a well circumscribed collection of fluid in the pancreatic tissue or the adjacent pancreatic space. The fluid contained is rich in amylase and other pancreatic enzymes. They occur most commonly as a complication of acute pancreatitis, but can complicate chronic pancreatitis as well. (7) The rupture of a pseudocyst can occur into one of several possible locations which include the gastrointestinal tract, general peritoneal cavity, vascular system and pleural cavity. (8,9,10) The common differential diagnoses in a

young boy presenting with a unilateral acute massive pleural effusion include haemothorax due to trauma to chest, parapneumonic effusion/empyema, chronic infection like tuberculosis and metastases to the pleural cavity of which lymphoma is of particular

importance. However, the presence of recurrent episodes of abdominal pain in the history raised the suspicion of a possible pancreatic aetiology for the pleural effusion. This was further reinforced by the dramatically elevated pleural fluid amylase level. Although definite diagnostic level is lacking, according to published literature the mean pleural fluid amylase level exceeds 10000 U/L in patients with PPF. (11) Other differential diagnoses for amylase rich pleural fluid include esophageal rupture, thoracic malignancy and acute pancreatitis, (11,12) but these could reliably be excluded by careful analysis of the clinical presentation and further investigations. CECT chest and abdomen was particularly invaluable here in confirming the diagnosis as well as excluding the differential diagnoses.

Pleural effusions secondary to PPF are usually large, recurrent and more commonly seen on the left side (42 - 67%). Less commonly it is seen on the right side as well as bilaterally (12) These effusions should be differentiated from the self-limiting, reactive left sided effusions that occur in acute pancreatitis.

As in this case, majority of patients with PPF present with dyspnoea. (65-76%) Other presenting symptoms include cough, chest pain and fever mimicking a thoracic aetiology and consequently many patients go through extensive pulmonary evaluation prior to diagnosis of pancreatic pathology. Therefor a high index of suspicion and careful analysis of history with regard to abdominal symptoms are essential especially when alcoholic patients present with massive pleural effusions. Pleural fluid amylase is a widely available, important diagnostic tool which can be utilized routinely when analyzing pleural effusions in patients with risk factors for pancreatic disease.

Conclusion

Chronic pancreatitis is a rare cause of acute massive unilateral pleural effusion. Due to preponderance of pulmonary symptoms, the diagnosis is often delayed. Therefore, it is essential to exclude important non-pulmonic aetiologies when evaluating patients with unexplained pleural effusions. Pleural fluid amylase level is a widely available diagnostic tool which can provide an important clue for diagnosis.

Authors' contributions

LB drafted the manuscript. DM supervised and involve total management and manuscript, SR support writing ,IN,AL,PW involve the management.

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Conflict of interest: None

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