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Pancreaticopleural Fistula: A Rare Complication of Pancreatitis in Children - A Case Report

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Abstract

Pancreaticopleural fistula (PPF) is a rare complication of pancreatitis, which can present as massive pleural effusion. We herein report a six-year-old male, a follow-up case of acute pancreatitis, who later presented with bilateral pleural effusion due to PPF. Initially, tuberculosis was suspected as the cause of pleural effusion due to the patient's maternal history of contact with tuberculosis and chronic history and chronic history. However, further investigations revealed a diagnosis of PPF. Management and outcome: He was managed conservatively for a prolonged period, following which there was a complete resolution of pleural effusion. This pediatric case highlights a rare complication of acute pancreatitis and the role of early initiation of management in these patients, thereby resulting in good outcomes even in a resource-limited setting.

Keywords: Pancreaticopleural fistula, high pleural fluid amylase, octreotide

Introduction

Pancreaticopleural fistula (PPF) is a rare complication of pancreatitis in children. The actual pediatric incidence of PPF is not known. In children, available literature is limited to case reports and case series. In adults, as is reported in 0.4% of cases of pancreatitis and 4.5% of patients with pancreatic pseudocyst[1]. A PPF develops when a pancreatic collection, usually a pseudocyst, ruptures into the pleural space, most commonly through the aortic or esophageal hiatus in the diaphragm, causing pancreatic fluid to leak directly into the pleural cavity due to a disruption in the pancreatic duct, leading to pleural inflammation and fluid accumulation; this is typically a complication of pancreatitis, with the most common underlying cause being chronic pancreatitis.

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Many times, diagnosis is delayed due to its presentation exclusively with pulmonary symptoms[2]. We report a case of a six-year-old boy with PPF who showed complete resolution of the condition on prolonged medical management in our resource-limited settings.

Clinical Description

A six-year-old boy who was previously diagnosed as a case of acute pancreatitis with pseudocyst eight months ago and was asymptomatic during follow-up with a diminishing pseudocyst size for the last 2 months from last discharge. He presented with sudden-onset breathlessness, chest pain (right-sided), and cough for 5 days. Dyspnea was present at rest as well as when lying down. Chest pain was bilateral, dull, aching, and non-radiating. The cough was non-productive and intermittent. There was a history of continuous, diffuse, non-radiating, dull aching abdominal pain for the last three days. There was no associated history of fever, hemoptysis, vomiting, loose stools, abdominal distension, decreased urine output, rash, abnormal body movement, and cyanosis. His past medical and treatment history revealed that he was diagnosed with seizure disorder at age five and was on treatment with sodium valproate. He had a history of severe abdominal pain and vomiting eight months ago, for which he was diagnosed with acute pancreatitis secondary to sodium valproate based on high serum amylase, serum lipase, and ultrasound findings. After the onset of pancreatitis, he had three short admissions over the course of next three months for which he was managed conservatively with analgesics, dietary management, proton pump inhibitors and antibiotics. During that time on ultrasound examination, pseudocyst formation was noted after 1 month of onset of acute pancreatitis. The serial sizes of pseudocyst on follow-up ultrasounds done at 4 weeks, 8 weeks and 16 weeks post-illness were as follows: 5×4×5 cm; two ppc of size 4.9×4.6×6 cm & 3.1×3.1×2.7 cm and 3.1×5.3×3 cm respectively. He was on conservative pain and dietary management along with vitamins and calcium supplements. There was a history of tuberculosis contact in the mother 2 years ago. The social, birth and developmental history were non-contributory. He consumed a mixed diet, which was deficient in both calories and proteins. On examination, there was tachypnea, tachycardia, and respiratory distress characterized by subcostal retractions and nasal flaring. The child was hemodynamically stable. His height and weight were less than the 3rd centile and body mass index was 11.2 kg/m^2. He appeared pale, but there was no icterus, clubbing, edema, lymphadenopathy, or rash. The trachea was deviated to the left with reduced chest movements on the right side. Stony dull percussion was present in bilateral subscapular, right infra-axillary, and axillary regions. Air entry was reduced in the lower part of the chest bilaterally with no added sound. Abdomen examination showed mild distension without any guarding, rigidity, organomegaly, fluid thrill and shifting dullness. Bowel sounds were normal. The rest of the systemic examination was unremarkable. A clinical diagnosis of bilateral pleural effusion was made. Since the child was afebrile and was a follow-up case of acute pancreatitis with pseudocyst, and also had a history of tuberculosis in the mother, clinically an etiological diagnosis of pleural effusion secondary to acute pancreatitis, tuberculosis or malignancy was made.

Management and Outcome

His initial investigations were as follows. Complete blood counts: Hemoglobin - 9.2 gm%. Total leukocyte count - 13,400/mm^3, differential leukocyte count - P62%/L32%, and platelet count - 2.7×10^5/mm^3. The Kidney Function test, Lipid profile, vitamin D, calcium, and phosphate levels were normal. The liver function tests showed the following: total bilirubin - 0.9 mg/dL; serum glutamic-oxaloacetic transaminase - 57 IU; serum glutamic pyruvic transaminase - 48 IU; alkaline phosphate - 208 IU. Serum amylase was 1032 IU/L and lipase was 730 IU/L. Chest X-ray showed massive bilateral pleural effusion (**Figure 1A**). Ultrasonography showed a bulky pancreas with an obscured tail, a peripancreatic fluid collection measuring 5×4×5 cm, bilateral B/L pleural effusion (with separations of 5 cm on the right side and 3 cm on the left side) and mild ascites. Diagnostic or therapeutic paracentesis was performed. Pleural fluid analysis showed cytology 10-12 RBCs/HPF, with no atypical cells, and 250 WBCs/mm^3 with 85% lymphocytes. Pleural fluid biochemistry showed a sugar level of 50 mg/dL, proteins at 5.2 g/dL, and albumin at 2 g/dL. The pleural fluid amylase level was high (32,962 U/L). A provisional diagnosis of PPF with bilateral pleural effusion and an underlying acute pancreatitis with pseudocyst was made. He was kept nil per os and was started on proton pump inhibitors, antibiotics, and octreotide infusion. Due to the non-availability of endoscopic retrograde cholangiopancreatography (ERCP) or surgical intervention for a small child, he was managed with conservative therapy over the course of three months only. During this period, a chest tube was inserted on the right side. Calories and proteins were optimised for his age, to avoid delayed improvement. There was subsequent improvement, marked by a reduction in drain output, prompting the removal of the chest tube after 10 days (**Figure 1B**). Octreotide infusion was continued as part of the treatment plan. Although efforts were made to introduce NG feeds, the patient struggled to tolerate them, necessitating the initiation of total parenteral nutrition (TPN). After three weeks of sustained improvement in pleural effusion and symptoms, the octreotide infusion was gradually tapered off and eventually stopped. Magnetic resonance cholangiopancreatography (MRCP) done during this time revealed walled-off necrosis in a peripancreatic region of size 5.9×6.7×6.3 cm near the tail of the pancreas, with extensions into the lesser sac anteromedially and cranially, and posteriorly to the right retrocrural region, and a fistulous tract (18 mm) originating from the pseudocyst and extending to the right hemithorax (**Figure 2**). A diagnosis of PPF was confirmed.

However, symptoms of respiratory distress along with pleural effusion reappeared within one week of the discontinuation of octreotide, prompting the reintroduction of octreotide infusion and chest tube drainage. This regimen was continued for another three weeks before being tapered off and discontinued. Following this, he was watched for the presence of any adverse reactions.

The patient had not experienced abdominal pain, vomiting or respiratory distress for about next five days. A chest X-ray done before discharge showed complete resolution of effusion (**Figure 1C**). On subsequent follow-up at 1 week, 1 month, 2 months, visits, the child was gaining weight and had no further symptoms. Pancreatic pseudocyst size on follow-up on abdominal ultrasound had also decreased to 3x3x2.5 cm. The patient's prolonged illness, presence of an indwelling ICD, and pancreatic pseudocyst led to early satiety, restricting oral intake. Consequently, the patient remained on a liquid diet for an extended period, resulting in nutritional deficiencies. However, as the pancreatic pseudocyst gradually decreased in size and nutritional deficiencies were corrected, the child began to gain weight.

Discussion

PPF is a unique complication of pancreatitis occasionally reported in adults and rarely in pediatric patients. On review of literature, only 33 cases of PPF have been reported amongst pediatric patients, whereas more than 300 cases of PPF have been reported amongst adults[1]. PPF is significantly rarer in children compared to adults, with the primary difference being that in adults, PPF is most commonly associated with chronic alcoholic pancreatitis, while in children, the cause is often unclear and may be related to genetic mutations causing chronic pancreatitis or trauma, making the diagnosis and management in children more complex; additionally, children may present with less typical symptoms due to their age and developmental stage. While adults may present with classic symptoms of chronic pancreatitis like abdominal pain, along with respiratory symptoms such as chest pain and shortness of breath due to pleural effusion, children may have less prominent abdominal pain. They may present primarily with respiratory symptoms like cough or difficulty breathing, which can lead to delayed diagnosis. When disruption of the pancreatic duct occurs posteriorly, amylase-rich pancreatic secretion can enter the retroperitoneal space, entering through various diaphragmatic orifices into the mediastinum, and further rupture into the pleural space to form PPF[1-3]. PPF usually presents as rapidly accumulating massive pleural effusion and is resistant to therapeutic thoracentesis. It needs to be distinguished from the self-limiting pleural effusion which is seen in 3% to 17% of cases of acute pancreatitis, that is unilateral, of mild to moderate volume and often resolves on its own during conservative management[4]. The major clinical symptoms of PPF are dyspnoea (52%), cough (24%), and chest pain (20%). Pulmonary symptoms create a diagnostic challenge and delay timely diagnosis. Other symptoms include abdominal pain (20%), vomiting (8%), and anorexia (8%). The most characteristic feature of PPF is a high pleural amylase level which can also be observed in tuberculosis, oesophageal perforation, lymphoma, liver cirrhosis or malignancy. The amylase level of PPF is grossly elevated, as seen in previously reported case series, ranging from 1200 U/L to 156,200 U/L[5].

For diagnosing PPF, imaging is essential. Due to bowel gas artifacts and inadequate respiratory cooperation, transabdominal ultrasound has limited effectiveness for diagnosing PPF[6]. Abdominal computed tomography is a commonly used imaging method in evaluating pancreatic parenchymal atrophy, pseudocyst, calcification, and ductal dilatation. But its sensitivity in diagnosing PPF is low ranging from 33% to 47%. MRCP has been recommended as the first-choice imaging modality for diagnosing PPF as it can visualize pancreatic duct anatomy (stricture or obstruction) in 80% of cases. ERCP can help in identifying precise pancreatic ductal anatomy and any duct disruption, but the major limitation is its low diagnostic sensitivity (38.5%)[7,8].

There is no well-established treatment for PPF in the pediatric population and management is limited to previously reported cases in pediatrics and adults. Initial management includes conservative therapy with medications received for 10 to 60 days (average 2 to 3 weeks), followed by endoscopic procedures or surgery for those failing conservative management. Medical therapy including, octreotide which is the mainstay acts by reducing large-volume pancreatic secretion, therefore, helping in the closure of disrupted pancreatic duct is seen in 31% to 65% of PPF patients combined with TPN also recognised in the index case after 3 weeks. The children usually

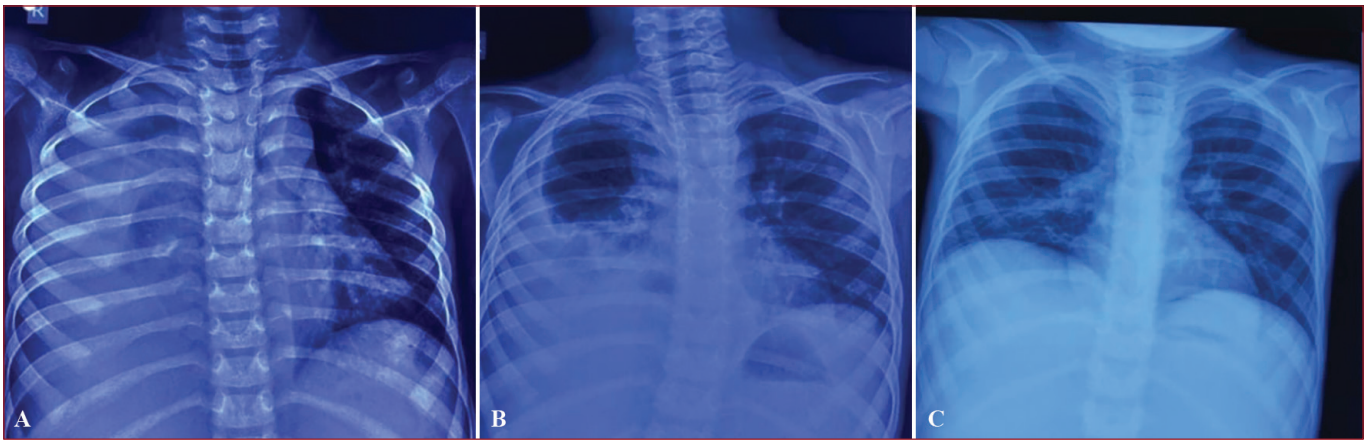

Figure 1. (A) Chest X-ray at the time of admission showing a large volume right sided pleural effusion presenting clinically as dyspnoea, **(B)** reducing pleural effusion during treatment in hospital, **(C)** significant improvement noticed before discharge

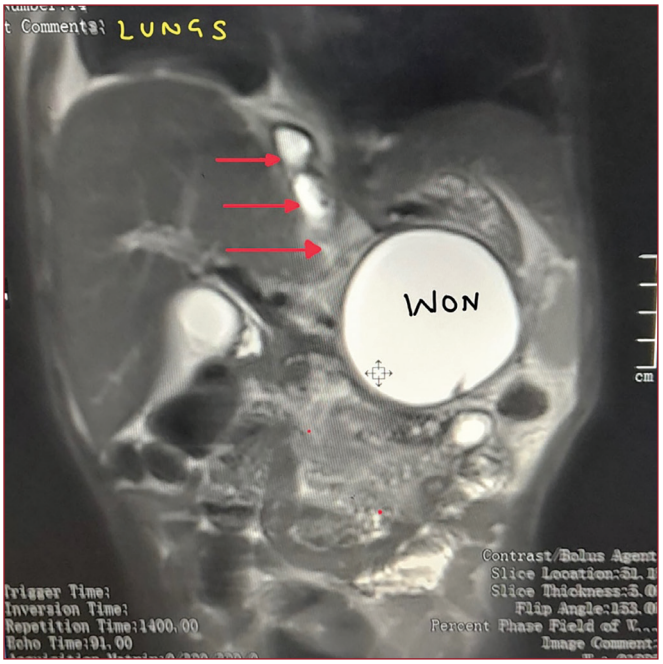

Figure 2. MRCP showing a well-defined thick-walled collection (WON) noted in region of tail of pancreas. The collection is extending into lesser sac anteromedially, cranially and posteriorly to right retrocrural region. A communication tract (PPF) seen crossing the crura and opening in right pleural cavity as shown with red arrow head

MRCP: Magnetic resonance cholangiopancreatographic

respond to standard octreotide therapy in 3 weeks in the presence of a closed thoracic drainage tube[9,10]. During conservative therapy, the child may develop malnutrition, septicemia, and other complications that need to be dealt with utmost priority[11]. Children who fail conservative management may further require endoscopic or surgical management with varying success rates. Endoscopy therapy includes placement of an ERCP stent, an evolving non-surgical method of management of PPF with an excellent success rate of 100% and 96.4% as reported by Khan et al.[12] and Pai et al.[13] respectively. ERCP is a less invasive procedure with shorter postoperative recovery time and with fewer complications (infections, bleeding, damage to the pancreatic duct, repeated accumulation of fluid and pancreatitis) is gradually developing as a procedure of choice which was comparable to surgical success rate of 94% reported by King et al.[8] with the only limitation of the requirement of substantial technical experience. In our case, PPF was managed in our resource-limited setting. This case highlights the role of clinical examination leading to early initiation of conservative management, resulting in good outcomes. This case presented with bilateral pleural effusion, with a significantly high level of amylase in the pleural fluid. Diagnosis of PPF was confirmed by MRCP. Due to the non-availability of surgical and endoscopic intervention, he was managed conservatively with octreotide infusion, and had a prolonged course lasting nearly six weeks. During the past six months of follow-up after discharge, the child has been in good health and is symptom-free with normalized serum amylase levels.

Conclusion

This case highlights the rare complication of acute pancreatitis and the role of examination, imaging and early initiation of conservative management in a case of PPF resulting in positive outcomes in a resource-limited setting. Illustrates the fact that when a patient has massive pleural effusion. Early diagnosis of these rare lesions is important to avoid a preventable fatal outcome.

Ethics

Informed Consent: Written informed consent was obtained from the parents before writing this case report.

Footnotes

Author Contributions: Mishra NM: Surgical and Medical Practices, Concept, Design, Analysis or Interpretation, Data Collection or Processing, Literature Search, Writing; Kumar S: Surgical and Medical Practices, Concept, Design, Analysis or Interpretation, Data Collection or Processing, Literature Search, Writing; Dewan V: Surgical and Medical Practices, Concept, Design, Analysis or Interpretation, Literature Search, Writing; Mishra VK: Surgical and Medical Practices, Concept, Design, Analysis or Interpretation, Literature Search, Writing; Mohan N: Surgical and Medical Practices, Analysis or Interpretation, Data Collection or Processing, Literature Search, Writing; Lamba A: Surgical and Medical Practices, Data Collection or Processing, Literature Search.

Conflict of Interest: The authors have no conflicts of interest to declare.

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