Mediastinal Pseudocyst and Cardiac Tamponade Due to Massive Pericardial Effusion in Pediatric Chronic Calcific Pancreatitis

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Abstract

Keywords

- ▶ chronic calcific pancreatitis
- ▶ mediastinal pancreatic pseudocyst
- ▶ cardiac tamponade
- ▶ Roux-en-Y cystojejunostomy

A 7-year-old male patient with a history of recurrent abdominal pain over 1 year presented with cardiac tamponade due to massive pericardial effusion, which was percutaneously drained. Contrast-enhanced computed tomography revealed a large posterior mediastinal cyst and calcified, heterogeneous pancreatic parenchyma. Elevated amylase and lipase levels of the cyst fluid confirmed the diagnosis of pancreatic pseudocyst, which was treated with an octreotide infusion and Roux-en-Y cystojejunostomy. The child was discharged on pancreatic enzyme supplement and was asymptomatic on follow-up.

Introduction

Pediatric chronic or relapsing pancreatitis is caused by genetic mutations, hyperlipidemia (types I, IV, and V), hyperparathyroidism, ascariasis, congenital anomalies of the pancreatic or biliary ductal system (such as pancreatic divisum and forme fruste choledochal cyst), and autoimmune and juvenile tropical pancreatitis. The following genetic mutations result in chronic pancreatitis¹:

1. *PRSS 1* or protease, serine-type 1 gene encoding for cationic trypsinogen, on the long arm of chromosome 7
2. *SPINK1* or serine protease inhibitor Kazal-type 1 gene encoding for pancreatic trypsin inhibitor, on chromosome 5
3. *CFTR* (cystic fibrosis) gene
4. *CTRC* (chymotrypsinogen C) gene
5. *PRSS2* or protease, serine-type 2 gene encoding for anionic trypsinogen
6. *CTSB* (cathepsin B) gene

Pseudocysts complicate 7 to 15% of episodes of acute pancreatitis and 20 to 25% cases of chronic pancreatitis,

and in rare instances, pseudocysts may also extend to the mediastinum.² The natural history, etiology, complications, and definitive treatment of mediastinal pseudocysts are still evolving due to their rare occurrence.² Pericardial effusion is a surprisingly common finding in acute pancreatitis, though cardiac tamponade is an extremely rare but serious finding.³ Herein, we report a rare case of pediatric chronic calcific pancreatitis with mediastinal pseudocyst formation and pericardial effusion leading to tamponade.

Case Report

A 7-year-old male patient presented to the emergency department with severe respiratory distress, progressively increasing over the past 2 days. The child was in shock, with thready peripheral pulses, cold extremities, tachycardia (150/minute), and tachypnea (40/minute). He had muffled heart sounds and bilateral vesicular breath sounds with diminished air entry over the left basal region. He had epigastric tenderness, no hepatosplenomegaly, and normal peristaltic sounds. Pallor, icterus, and significant lymphadenopathy were absent.

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Careful auscultatory blood pressure (BP) measurement revealed a fall in systolic BP > 10 mm Hg with inspiration (pulsus paradoxus). Emergency chest radiography (**Fig. 1**) revealed a globular-shaped enlarged cardiac shadow, left-sided pleural effusion, and mediastinal widening. Echocardiography revealed a large pericardial effusion, right atrial systolic collapse, and right ventricular diastolic collapse—signifying cardiac tamponade. We treated with fluid resuscitation, vasoactive medications (dopamine, $10 \mu\text{g/kg/min}$), and intravenous antibiotics. Under the echocardiographic guidance, a pigtail catheter was introduced into the pericardial space, and 300 mL of exudative fluid was removed over the next 48 hours, thereby relieving the signs and symptoms of obstructive shock. The pericardial fluid analysis revealed 3,000 leukocytes/ mm^3 (60% lymphocytes, 40% neutrophils), glucose 10 mg/dL, protein 4 g/dL, and negative cultures. The child's respiratory distress markedly improved following pericardiocentesis, but the child continued to complain of dull-aching pain in the upper abdomen. Detailed history revealed that the child had recurrent episodes of abdominal pain over the past year, which had been treated with oral antispasmodic agents and H_2 blockers. He had no history of chronic diarrhea or a similar disease in any family member. Ultrasound of the thorax (**Fig. 2**) and abdomen revealed a large retrocardiac cystic structure, bilateral pleural effusion, heterogeneous echotexture of the pancreas, and normal biliary and pancreatic ducts without any calculus.

Contrast-enhanced computed tomography of the abdomen revealed a large encysted mediastinal cystic lesion with trifoliate shape and lobulated outlines in the posterior mediastinum, more on the left, pushing the heart anteriorly, and to the right, extending from the level of upper pulmonary vein (**Fig. 3A**) to the level of the upper pole of the left kidney along the posterior mediastinal recess (**Fig. 3B**). The lesion had a thick enhancing wall with foci of calcifications

Fig. 1 Chest skiagram showing enlarged cardiac silhouette, mediastinal widening, and left pleural effusion (marked with an arrow).

Fig. 2 Ultrasound of the chest showing postdrainage residual pericardial effusion (marked PERI EFF), the heart (marked HT), and the retrocardiac huge cystic cavity (marked with an arrow).

(**Fig. 3B**). The bilateral lower lobes and the lingular lobe of the lungs had patchy consolidations. There was a mild pericardial effusion. Pancreatic echotexture was heterogeneous with foci of calcification throughout the parenchyma (**Fig. 3C**). The lesser sac contained a small encysted collection in the anterior peripancreatic region (**Fig. 3D**). Complete blood count revealed hemoglobin: 10 g percent, mild leukocytosis: 16,500/ mm^3 (neutrophil 70%, lymphocyte 28%, eosinophil 1%, and monocyte 1%), and platelet: 1.8 lakhs/ mm^3 . Liver and renal function tests were normal. Serum amylase: 458 U/L (reference value: 28–100 U/L), lipase: 2,500 U/L (reference value: 5–31 U/L), cholesterol: 150 mg/dL, triglyceride: 120 mg/dL, calcium: 10 mg/dL, phosphorous: 4 mg/dL, and plasma glucose: 80 mg/dL. Stool examination was negative for parasites. The sweat chloride test was normal. The cyst fluid was aspirated under computed tomography (CT) guidance. The serosanguinous fluid had amylase 27,900 U/L and lipase 206,500 U/L, thereby confirming it to be a pancreatic pseudocyst. Coxsackie, enterovirus, herpes simplex virus, human immunodeficiency virus, Epstein-Barr virus, hepatitis B and C, TORCH (Toxoplasma, Other infections, Rubella, Cytomegalovirus, Herpes simplex virus), mycoplasma, and chlamydia serologies were negative in serum. Antinuclear factor, antineutrophilic cytoplasmic antibody, and rheumatoid factor were negative. Blood culture was sterile. The child was treated with pantoprazole, pancreatic enzyme supplementation (500 U lipase/kg/meal), octreotide infusion ($2 \mu\text{g/kg/h}$), and tramadol for analgesia.

The child was operated 10 days after admission. Under general anesthesia, a thoracoabdominal incision was given through the left seventh intercostal space and the abdomen was explored. No prominent cyst was found within the abdomen. Exploration of the pancreatic region showed no

Fig. 3 (A) Axial section of computed tomography of the thorax showing the upper extension of the cystic cavity (marked with a block arrow) at the level of the upper pulmonary vein (marked with a line arrow). (B) The sagittal section of a computed tomogram of the chest and abdomen showing the extension of the retrocardiac cyst (marked with a block arrow) along the posterior mediastinal recess into the abdomen. The calcifications in the cyst wall (marked with a horizontal line arrow) and pancreatic parenchyma (marked with a vertical line arrow) is seen. (C) The axial section of a computed tomogram of the abdomen showing focally calcified and heterogeneous pancreatic parenchyma (marked with a block arrow). (D) The axial section of the computed tomogram abdomen showing the presence of a small collection in the peripancreatic region (marked with a line arrow).

sign of acute pancreatitis. Resection of the seventh rib revealed a huge cyst with thick walls and adventitious vessels in the posterior mediastinum. The adjacent lung tissues were collapsed and the heart was shifted anteriorly. The cyst was opened in between stay sutures (►Fig. 4) and 1.5 L of fluid drained. The cyst wall was shaggy. Through a rent in the diaphragm, the jejunal loop was brought inside the thoracic cavity and then inside the cyst through a rent in the inferior wall (►Fig. 4). Suturing the rim of the internal lumen of jejunum around the cyst wall created the Roux-en-Y cysto-jejunostomy loop. The diaphragm was repaired, seventh rib approximated, intercostal tube drain placed, and the abdomen closed in layers. He received one unit of packed erythrocyte transfusion intraoperatively. Postoperatively, the child received octreotide infusion for 4 days and stopped afterwards. Histopathology revealed that the cyst wall contained lymphoid aggregates, hemosiderin-laden macrophages, and lacked epithelium. There was no evidence of granuloma or

malignancy. The child was discharged on the 14th postoperative day with oral pantoprazole, pancreatic enzyme supplements, and multivitamins. We planned monitoring of the long-term complications of chronic pancreatitis. He was asymptomatic on follow-up with normal blood glucose and calcium levels. Serial ultrasounds showed regression of the mediastinal cyst. After 8 months, MRCP (magnetic resonance cholangiopancreatography) showed resolution of the cyst, heterogeneous and focally calcified pancreatic parenchyma, without pancreatic or biliary ductal dilatation or calculus.

Discussion

Galligan and Williams first reported pediatric mediastinal pancreatic pseudocyst in 1966.⁴ The most common cause of pseudocyst in children is trauma.^{5,6} Visrutaratna and Ukarapol reported mediastinal pancreatic pseudocyst in chronic pancreatitis in a 14-year-old girl in 2010.⁷ Pathophysiologically,

Fig. 4 The intraoperative picture showing the ileal loop (marked with a black arrow) being brought into the cyst cavity for anastomosis. The cyst has been opened between stay sutures; the margin of the opening is marked with line arrows.

mediastinal pseudocysts develop after rupture of the pancreatic duct posteriorly into the retroperitoneal space. Because the more common pathways of spread of the pancreatic fluid offer, greater resistance due to fibrosis from previous inflammation, the spread of fluid occurs along the path of least resistance. In such cases a mediastinal extension of the pancreatic fluid occurs along the following pathways: peripancreatic space → retromesenteric plane → left extraperitoneal space → esophageal hiatus → retrocrural space → posterior mediastinum. Peripancreatic space → retromesenteric plane along the left perirenal bridging septa or across the left fascial trifurcation → left retrorenal plane → further track into retrocrural space via esophageal hiatus. Fluid may also extend from the peripancreatic space into the retrocrural space and the right mediastinum via the aortic hiatus and the inferior venacaval hiatus, respectively.⁸ The fluid collection may enlarge slowly and hence delay the appearance of symptoms as in our case.²

Complications due to mediastinal pancreatic pseudocyst can occur due to compression of lymphatic flow from lungs, leading to lymphatic interstitial edema, obstruction of the trachea or major bronchus resulting in respiratory compromise, retrocardiac compression leading to elevated filling pressures causing congestive heart failure, and erosion into the pericardial sac causing life-threatening tamponade.^{2,9,10}

Medical management of pseudocyst includes somatostatin analogue octreotide, which by inhibiting pancreatic secretions facilitates closure of the pancreatic fistula.² Transcutaneous external drainage with CT-guided stent placement has a recurrence rate of 8% in children.¹¹ Endoscopic retrograde cholangiopancreatography with transpapillary duct drainage and endoscopic stent placement and transgastric or transesophageal endoscopic drainage has been described in adults.² For complicated pediatric cases of pancreatic pseudocysts, internal drainage options include cystogastrostomy, cystoduodenostomy, cystojejunostomy, the recurrence rates being 4.7,

50, and 7.6%, respectively.^{7,11} However, till 2015 cystoenterostomy operation for pediatric thoracoabdominal pancreatic pseudocyst has been described in only two cases.¹²

Numerous mechanisms have been suggested for the mechanism of pericardial effusion in pancreatitis, including chemical pericarditis due to pancreatic enzymes carried by lymphatic vessels or available in circulation, inflammation of vascular walls in fatty areas leading to subpericardial fat necrosis, and the fistulous connection between abdominal and pericardial cavities. Chylous pericardial effusion has been reported in acute necrotizing pancreatitis. The simultaneous occurrence of pericarditis and pancreatitis secondary to viral infection can occur. Usually, pericardial effusion disappears spontaneously after clinical recovery except for effusion associated with fistula formation.⁹ Pericardial effusion that presents as cardiac tamponade following pancreatitis is extremely rare, but has been reported by Bhatt et al³ and Parekh et al¹³ in adults. To the best knowledge of the authors, this is the first case report of cardiac tamponade due to massive pericardial effusion along with a posterior mediastinal pseudocyst in a child.

Endocrinal, metabolic, infective, and anatomic structural anomalies were excluded as the cause of chronic pancreatitis in this case. Juvenile idiopathic pancreatitis/tropical calcific pancreatitis is the most common cause of chronic pancreatitis in developing tropical countries.¹ Although initially reported from South India, the entity is seen in all parts of our country. The disease is characterized by recurrent episodes of abdominal pain, protracted malnutrition, ketosis-resistant diabetes in 33%, and pancreatic cancer in 10% patients.¹⁴ North Indian patients differ in having a higher prevalence of pain and lower frequency of diabetes, calcification, and intraductal calculi.¹⁴ In noncalcific variants of idiopathic chronic pancreatitis in North India, pseudocyst formation, and segmental portal hypertension is frequent, pseudocyst being the most common local complication.¹⁵ In calcific variants, diabetes mellitus and abnormal fecal fat excretion occur more frequently.¹⁵ However, the index case, although ethnically originating from the North Indian state of Uttar Pradesh and having calcific chronic pancreatitis, developed pancreatic pseudocyst. Initially malnutrition with protein deficiency, cassava toxicity, and impaired immune response to viral infections were postulated as the etiology for juvenile tropical idiopathic pancreatitis. However, strong genetic susceptibility and comparative phenotype of idiopathic chronic pancreatitis in India suggest that the term tropical calcific pancreatitis is a misnomer.¹⁶ *SPINK 1* and chymotrypsinogen gene mutations are strongly associated with this form of pancreatitis.^{1,16,17}

Thus, the present case stresses the need to diagnose the relatively uncommon entity of chronic pancreatitis in the pediatric population and to be vigilant about the rare but potentially serious complications such as mediastinal pseudocyst and cardiac tamponade.

Conflict of Interest
None.

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