

Bilateral pancreaticopleural fistula in an infant: a rare complication of chronic relapsing pancreatitis

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An infantile case of chronic relapsing pancreatitis complicated by bilateral pancreaticopleural fistula and intractable pleural effusion is reported. Surgical drainage of the retroperitoneal pseudocyst proved effective, and the patient remains symptom free 3 years after surgery. Recognition of this entity and early surgical intervention may prevent long-term morbidity of chronic pancreatitis in children. A literature search was performed using electronic databases (PubMed, MEDLINE, and Embase) and the citation list of relevant publications. The latest searches were performed on April 3, 2013.

Key words: pancreaticopleural fistula, chronic pancreatitis, pleural effusion

Introduction

Pancreaticopleural fistula (PPF) is a rare complication of chronic pancreatitis which causes intractable pleural effusions.¹⁻⁷ Pleural effusion caused by PPF is distinguished from small, self-limiting pleural effusion associated with acute pancreatitis by a high level of amylase in the effusion.²⁻⁵ Most of the reported cases are adult males mainly with alcohol-related chronic pancreatitis.^{5,6,8,9} PPF in children is extremely rare, and only 8 cases have been reported in the literature to date.^{1-3,7} We herein report a new infantile case with bilateral PPF and a review of the literature.

Case report

A 6-month-old girl presented with a few days history of fever, vomiting, and diarrhea. An abdominal computed tomography (CT) showed enlarged pseudocysts of the head and tail of the pancreas with massive ascites. Despite medical treatment for acute pancreatitis including total parenteral nutrition, antibiotics, and octreotide, the pseudocysts enlarged and were complicated by bleeding, which required a retroperitoneal drainage as well as percutaneous cyst drainages. Hereditary pancreatitis was ruled out due to her family history, and there was no evidence of viral infection, tumor, biliary stone, or trauma. Endoscopic retrograde cholangiopancreatography

(ERCP) failed to show pancreaticobiliary malconnection or a pancreas duct obstruction as a cause of pancreatitis.

Acute symptoms subsided, but enteral nutrition caused exacerbation of pancreatitis and eventually bilateral intractable pleural effusion because of the increased secretion of digestive juices. At that time, the patient was transferred to the current institution, NCCHD (National Center for Child Health and Development).

Laboratory data showed elevated serum levels of pancreatic enzymes: amylase (192 IU/l), lipase (760 IU/l), trypsin (1,000 ng/ml), phospholipid A2 (4,860 ng/dl), and elastase 1 (3,100 ng/dl). The amylase level in the blood-stained pleural effusion was 35,450 IU/l. CT revealed a cystic lesion posterior to the head of the pancreas, portal vein thrombosis, cavernous transformation, and splenomegaly (Figure 1). The pancreatic duct of the tail was not dilated. Respiratory distress due to massive pleural effusion progressed despite bilateral pleural drainage and conservative therapy for 1 month. She, therefore, ultimately required ventilatory support when surgical therapy was introduced. The operative pancreaticogram showed bilateral fistulas extending from the pseudocyst into the mediastinum (Figure 2), and cystenterostomy was performed (Figure 3). She tolerated this surgery and has been symptom-free during 3 year's of follow-up. She is currently eating without restriction with normal exocrine and endocrine pancreatic function.

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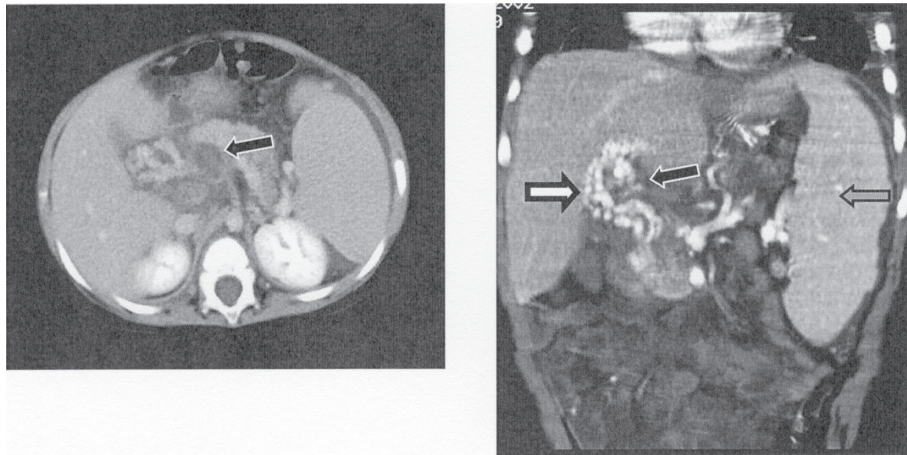
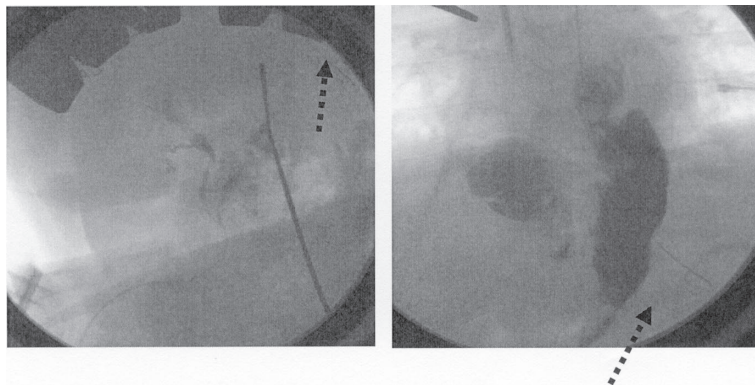
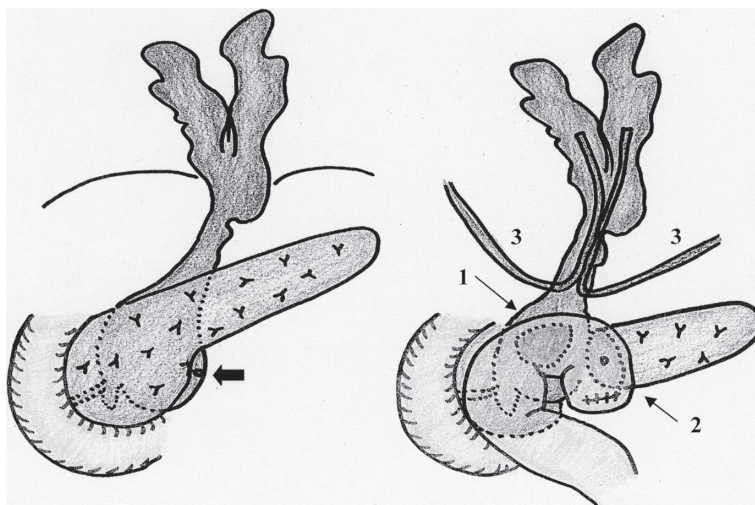


Figure 1. Abdominal CT scan showing the obstruction of the portal vein with the cystic lesion (black arrow), cavernous transformation (white arrow), and splenomegaly (gray arrow)



A. Cystography showing fluid collection from the pseudocyst going to the diaphragm (arrow) **B.** Extension of the fluid collection into the mediastinal space (arrow)

Figure 2



A. Pancreatic pseudocyst, pancreatico-pleural fistula, and opening of the pseudocyst (arrow) **B.** 1. cystenterostomy, 2. pancreaticoenterostomy, and 3. drainage of fistula

Figure 3

Discussion

PPF is a rare condition in which pancreatic fluid leaks directly into the pleural cavity, most commonly from a retroperitoneal pseudocyst through the diaphragmatic hiatus or directly through the diaphragm.^{1,3-6} It commonly results from disruption of the main pancreatic duct and pseudocyst formation in adult males with alcohol-related chronic pancreatitis.⁷ In 1 adult case, it was a rare report of bilateral pleural fluid storage.⁷ The hallmark of PPF is massive recurrent pleural effusion, which is often stained with blood and rich in amylase. This is in contrast to benign small pleural effusion associated with acute pancreatitis. Due to the rarity of chronic pancreatitis in children, only 8 cases of PPF have been reported in the pediatric population in the literature (Table 1).^{1-3,8} To our knowledge, the present case is the first pediatric case with bilateral PPF.

PPF must be suspected when the effusion is massive, recurrent, and rich in amylase. However, the radiological demonstration of PPF is often difficult even with ERCP, and the diagnosis can be delayed.^{1-3,8} A fistula was observed in 6 of 9 (67%) of the pediatric cases (Table 1). Although abdominal symptoms preceded the

development of respiratory symptoms in the present case, the underlying pancreatic disease can be asymptomatic, which may further delay the diagnosis.

Conservative management including chest tube drainage, parenteral nutrition, somatostatin analogues, or endoscopic pancreatic duct shunt placement can be successful in the adult population.^{4-6,9,10} However, this must be cautiously interpreted in children. All the reported pediatric patients (N = 9) were treated with surgical intervention. The aim of surgery is to achieve complete internal drainage of the main pancreatic duct.^{1-3,8,11} Longitudinal pancreaticojejunostomy known as the Peustow procedure is the standard procedure.^{1,3,4,8} However, in selected cases with a nondilated main pancreatic duct, cystenterostomy may be sufficient.^{5,8,10-13} It is unnecessary to repair the fistula as long as the pancreatic duct drainage is sufficient. Surgical procedures of PPF in the adult population have a good long-term outcome in 80%-95% of the cases.^{6,10,12} These results are likely to be reproduced in children according to the 9 cases reported to date in the literature. The recognition of this entity and early surgical intervention is necessary to prevent the progression of exocrine and endocrine insufficiency.

Table 1. Reported cases of pancreaticopleural fistula in children

Case	Age (yr)	Gender	Side of pleural effusion	Presentation	Serum amylase (IU/l)	Effusion amylase (IU/l)	Fistula demonstrated (method)	Pseudocyst	Treatment	Outcome	Etiology	Reference
1	12	M	Left	Abdominal pain	Lipase 1,974	Lipase 40,000	Yes (ERCP)	No	LPJ	Well	UK	Ranuh ¹
2	3	M	Right	Abdominal pain, dyspnea	4,935	>16,000	No	Yes	LPJ	Well	UK	Wakefield ²
3	4	M	Right	Anorexia, abdominal pain	329	9,737	Yes (ERCP)	Yes	LPJ	Well	UK	Wakefield ²
4	4	F	Right	Abdominal pain, dyspnea	751	12,170	Yes (ERCP)	Yes	LPJ	Well	UK	Bishop ³
5	3	M	NS	NS	NS	NS		NS	LPJ	UK	UK	Petersen ⁴
6	14	F	NS	NS	NS	NS		NS	Cystenterostomy	Well	UK	Petersen ⁴
7	4	M	NS	NS	NS	NS	2/4 (ERCP)	NS	LPJ	Well	UK	Petersen ⁴
8	7	M	NS	NS	NS	NS		NS	Distal pancreatectomy	Well	Hereditary	Petersen ⁴
Present case	0.5	F	Bilateral	Fever, vomiting, diarrhea	192	35,450	Yes (cystography)	Yes	LPJ, cystenterostomy	Well	UK	-

ERCP, endoscopic retrograde cholangiopancreatography; LPJ, longitudinal pancreaticojejunostomy; NS, not significant; UK, unknown

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