Pancreatic pseudocyst causing portal vein thrombosis and pancreatico-pleural fistula

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Abstract

Portal vein thrombosis and pancreaticopleural fistula are unusual complications of chronic pancreatitis. We describe a patient with chronic alcoholic pancreatitis in whom erosion of the splenic vein led to portal vein thrombosis and to the development of a pancreatico-pleural fistula. We suggest that fistula formation may occur over a considerable time period as the portal vein thrombosis was diagnosed three years before the amylaserich pleural effusions.

Pancreatico-pleural fistula¹⁻³ and portal vein thrombosis⁴ are rare, but well described, complications of chronic pancreatitis. Pancreatic fistulae are thought to be caused by disruption of the pancreatic duct in patients with chronic pancreatitis. Little is known of the natural history of pancreatic fistulae before clinical presentation and one of the puzzling features of the condition is that it may appear in the absence of symptoms or signs of pancreatic inflammation.¹ We present a patient in whom the fistulous track involved the portal venous system and probably caused portal vein thrombosis. As the portal vein thrombosis was noted three years before the patient presented with pancreatic pleural effusions we suggest that this lesion may

have a very long presymptomatic phase.

Case report

A 36 year old man, with a history of excessive alcohol intake, initially presented to another hospital in October 1984 with acute abdominal pain. Serum amylase was 1800 IU/l and a diagnosis of pancreatitis was made. Ultrasound showed an enlarged head of pancreas but no pseudocvst. Repeat ultrasound three months later showed similar findings. After discharge from hospital he reduced his alcohol intake but experienced similar, milder symptoms on two further occasions. These were treated at home with oral analgesics and a liquid diet. In August 1985 he was referred to our unit because of continuing abdominal discomfort. Serum amylase, in out-patients, was raised at 876 IU/l. An abdominal computed tomography scan revealed a cyst $(3 \times 2.5 \text{ cm})$ in the head of the pancreas (Fig 1) and a possible portal vein thrombosis. He was admitted to hospital where a splenic venogram confirmed portal vein thrombosis. An ERCP showed distortion of the pancreatic duct consistent with chronic pancreatitis, a small cyst in the head of pancreas communicating with the ductal system, and a normal biliary tree. In view of the small size of the cyst and the presence of portal hypertension, surgery to drain the cyst was not performed. In October 1987 he was admitted after a minor haematemesis. A diagnosis of bleeding from gastritis was made on endoscopy. Small white oesophageal varices were also noted. He was treated with H₂ blockers and discharged.

In January 1988 he was admitted with a one week history of gradually increasing shortness of breath and chest discomfort. He admitted to drinking approximately 284 ml of lager daily for the previous three years. On examination he was tachypnoeic, had a tachycardia and large bilateral pleural effusions. He was not jaundiced and there was not evidence of ascites. Serum amylase was 2663 IU/l and pleural fluid amylase 73000 IU/l. An abdominal computed tomography scan showed an irregular pancreas with calcification and a few tiny cystic areas, and confirmed the absence of ascites. He was treated initially with H₂ blockers and oral pancreatic supplements to suppress pancreatic secretion and with total parenteral nutrition and regular thoracentesis for symptom relief. Intravenous antibiotics were added subsequently when he became febrile. He required repeated large volume thoracentesis for relief of dyspnoea and bilateral large bore chest drains were inserted. As

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Figure 1: Abdominal computed tomography (CT) scan with oral and intravenous contrast showing a kidney shaped pseudocyst in the pancreatic head (arrowed).

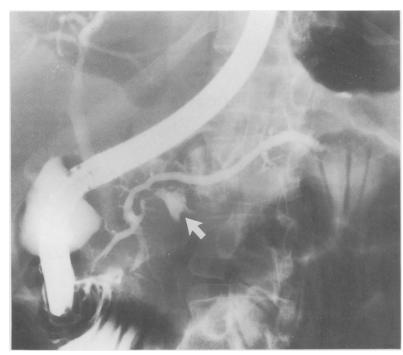
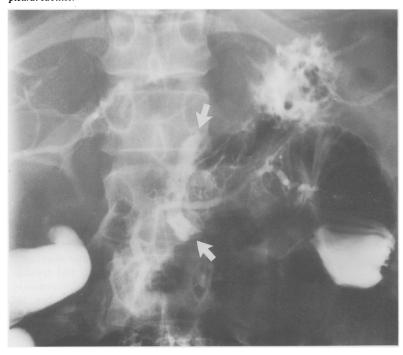


Figure 2: Endoscopic retrograde cholangiopancreatography (ERCP) showing a small cavity in the head of pancreas (arrowed) communicating with the pancreatic ductal system.

he was not responding after three weeks of conservative therapy a subtotal pancreatectomy was planned. Before surgery an ERCP was performed which showed a grossly distorted pancreatic ductal system with a small cyst in the head of the pancreas (Fig 2). Late films showed that contrast from this cystic lesion flowed upwards towards the mediastrinum in a fistulous track (Fig 3). Unfortunately, before surgery could be performed he developed respiratory failure and required ventilation. He subsequently developed severe intrathoracic sepsis with abscess formation, adult respiratory distress syndrome and renal failure. Despite ventilation, haemofiltration, intravenous antibiotics and numerous attempts to drain the intrathoracic abscesses he died nine weeks after admission.

Figure 3: Late film (20 mins) after the ERCP illustrated in Figure 2 showing contrast filling the inferior portion of the fistulous tract (arrowed) between the pancreas and the pleural cavities.



Post mortem examination confirmed the diagnosis of chronic pancreatitis with duct ectasia and ulceration of the wall of the distal part of the splenic vein. The ulcer extended to involve the proximal part of the portal vein and was continuous with the wall of a haemorrhagic cavity leading up into the posterior mediastinum through the oesophageal hiatus (Fig 4). Distally the portal vein showed evidence of previous thrombosis and recanalisation. Numerous loculated abscesses were present in the left pleural cavity.

Discussion

This patient had chronic alcoholic pancreatitis complicated by pseudocyst formation, portal vein thrombosis and a pancreatico-pleural fistula. We are not aware that a previous case with this combination of rare complications of chronic pancreatitis has been reported. At necropsy it appeared the pseudocyst initially eroded into the splenic vein causing portal vein thrombosis and subsequently progressed upwards into the mediastinum to rupture into the pleural cavity. The fistula may have eroded along the line of the left gastric (coronary) vein to reach the perioesophageal venous collaterals. These events occurred over a three year observation period which suggests that pancreatico-pleural fistulae may develop very slowly. It is possible that initially the portal vein thrombosis was unrelated directly to the pseudocyst and the fistula and that by chance the fistula eroded through the already thrombosed vein. This latter possibility appears less likely as in most cases of extrahepatic portal hypertension complicating pancreatitis the splenic vein alone is involved because of its close proximity to the pancreas.5

Pancreatic fistulae causing pancreatic ascites or pancreatic pleural effusions have been increasingly recognised in the last 15 years particularly in patients with alcoholic pancreatitis.13 They occur because of disruption of the duct in patients with chronic pancreatitis. The diagnosis may easily be missed if amylase is not measured in the pleural fluid as many patients have no abdominal symptoms or signs. Indeed one of the puzzling features of this condition has been the number of patients reported with no previous history of pancreatitis. Even in those with a history of acute pancreatitis the reported episode, in many cases, occurred months or years before the presentation of the fistula.¹ This discrepancy between the abdominal symptoms and the manifestation of the fistula may be explained if fistulae have a very long presymptomatic phase, as we suggest. In those patients with no history of an acute attack of pancreatitis a previous attack of abdominal pain may have been misdiagnosed, ignored, or forgotten.

Because of its rarity there are no controlled trials concerning management of pancreaticopleural fistula. Most authors suggest a trial of conservative therapy for two to three weeks,^{1-3 6-8} aimed at reducing pancreatic secretion and improving patient nutrition. If this fails excision or drainage of the damaged pancreas is recom-

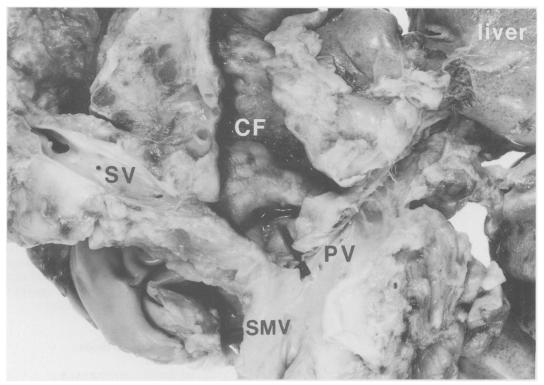


Figure 4: Posterior dissection of the portal venous system showing ulceration of the distal parts of the splenic vein (SV) and part of the cavity of the fistula (CF). Strands of connective tissue are seen in the lumen of the recanalised portal vein (PV). SMV =superior mesenteric vein

mended. It is also recommended that the fistula be visualised before surgery so that the appropriate procedure may be planned preoperatively. In many cases the fistula may be visualised by ERCP,⁷⁹ computed tomography scanning,² or the injection of lipiodol into the pleural cavity.¹⁰ There is one report of successful treatment using low dose radiotherapy.9 Conservative treatment failed in our patient and surgery was planned.

Drainage of the pseudocyst after its identification in 1985 might have averted the tragic sequelae in our patient. Most authors now recommend early drainage of pseudocysts.¹¹ Conservative treatment may be justified in patients with small pseudocysts, however,12 particularly if they occur in association with an acute attack of pancreatitis.13 The presence of portal hypertension⁴ and the location of the pseudocyst in the head of the pancreas would have added to the difficulties of surgery in our patient.

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