Pancreaticopleural fistula: a rare complication of chronic pancreatitis

Simon Wakefield, Benjamin Tuttty, Julian Britton

Summary
Pancreaticopleural fistula is an uncommon sequelae of pancreatitis. The condition is often elusive, as respiratory rather than abdominal symptoms usually predominate and the fistula can be difficult to demonstrate radiologically. Confirmation is by demonstrating a high amylase content in the pleural aspirate relative to the serum. About half the fistulae will close with conservative treatment but persistent or recurrent effusions, often associated with stenosis or disruption of the main pancreatic duct, are an indication for surgery. The long-term outcome is good in 80–95% of cases. We report five patients with pleural effusion of pancreatic origin due to pancreaticopleural fistula.

Keywords: pancreatitis, pancreaticopleural fistula, pleural effusion

The association between pancreatic disease and pleural effusions is well recognised.1-3 Small, transient, usually left-sided effusions, thought to be either lymphatic or sympathetic in origin, often occur with acute pancreatitis. By contrast, chronic pancreatic disease, with or without pseudocyst formation, may be associated with large, recurrent pleural effusions. A fistula connecting the pancreas and pleural cavity may be present or direct extension of a pseudocyst through the diaphragm may occur.4 The pleural aspirate usually has a higher amylase content when compared to serum levels. Patients usually present with symptoms attributable to the pleural effusion and the underlying pancreatic disease is often asymptomatic.4,5 Frequently, the diagnosis is delayed as the condition is rare and, in addition, the fistula is often difficult to demonstrate radiologically. We report five patients with pleural effusion of pancreatic origin due to a pancreaticopleural fistula.

Case reports
The case reports are summarised in the table.

Discussion
Pleuropulmonary complications in association with pancreatitis are common. Atelectasis, pneumonitis, adult respiratory distress syndrome and pancreatico-bronchial fistulae may occur.1,7 Small pleural effusions develop in 3–17% of patients with acute pancreatitis and have been suggested as a negative prognostic feature.8 The pancreatic enzyme activity of the pleural fluid is low and the effusion tends to resolve along with the pancreatitis. These effusions are probably due to sympathetic, chemically induced, diaphragmo-pleural inflammation. Large recurrent effusions are a hallmark of pancreaticopleural fistula, and were present in all our patients. Pancreaticopleural fistula is commonly associated with both disruption of the main pancreatic duct and pseudocyst formation. The fistula develops either by direct passage of a...
pseudocyst through a natural diaphragmatic hiatus (eg, oesophageal or aortic) or by direct fistulisation through the dome of the diaphragm. The resulting pleural fluid is amylase-rich. When a fistula is present it is often difficult to demonstrate. Radiological evidence of a fistula, using endoscopic retrograde cholangiopancreatography (ERCP), was only found in two of our patients (cases 2 and 3). The fistula was evident at surgery in a further two patients (cases 4 and 5) and in one patient no fistula could be demonstrated (case 1). In this case, however, the acute pleural effusion that developed at ERCP is strong evidence that a fistula was present. All patients had evidence of chronic pancreatitis with irregularities in the pancreatic duct and pseudocyst formation, but only in one patient was there a preceding history of pancreatic disease.

The diagnosis of pancreaticopleural fistula is often delayed as the condition is rare. The presence of a large, recurrent pleural effusion may be the only presentation as the pancreatic disease is often asymptomatic. It is the amylase activity of the pleural fluid, which is markedly elevated when compared to serum levels, that confirms the diagnosis. Computed tomography (CT) was not able to provide an accurate radiological delineation of the fistula in any of our patients, although a previous report had suggested that CT is the investigation of choice. ERCP is mandatory to delineate the pancreatic anatomy but will often fail to demonstrate the fistula. Selective duct cannulation or even an operative pancreateogram (figure) may be required in the presence of tight strictureing.

Initial treatment should be directed towards control of the pleural effusion. Intercostal tube drainage, parenteral nutrition and somatostatin or its analogue octreotide achieved a 48% fistula closure rate over a 2–3 week period with conservative management. Persistence or recurrence of an effusion is an indication for pancreatic surgery and occurred in all our patients. Even when conservative treatment is successful, the underlying cause – chronic pancreatitis, may need addressing. If obstruction to the duct system is present, surgical decompression, with or without excision of the involved portion of the obstructed pancreas, may be required. It is unnecessary to repair a fistulous tract if pancreatic duct obstruction is dealt with adequately. Surgical procedures have an operative mortality of 3–5% and a good long-term outcome in 80–95%.

### Summary/learning points

- presentation of pancreaticopleural fistula is often with pleural effusion
- pancreatic disease is often asymptomatic
- pleural aspirate has a high amylase content while serum amylase is often normal
- radiological demonstration of the fistula is often difficult
- delineation of pancreatic duct anatomy by ERCP or intraoperative pancreateography is mandatory
- indications for surgery are persistent or recurrent effusions and stenosis or disruption of the main pancreatic duct

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