Tracheal paraganglioma

from either supraglottic and subglottic laryngeal paraganglia is not disputed.

In one of the previously reported cases of tracheal paraganglioma massive bleeding accompanied surgical removal and the patient died perioperatively. Although this was a large tumour extending from vocal cord to carina, there was no evidence of malignancy. The three other cases have behaved in a benign fashion. Evidence has recently been collected that the presence of sustentacular cells positive for S100 and GFAP is a marker for a benign lesion and, specifically, that they are not found in malignant lesions. It has also been shown that GFAP positive cells are found only in parasympathetic paragangliomas.

The immunohistochemical findings in this case confirmed that the tumour was a paraganglioma derived from parasympathetic paraganglia and suggest that benign clinical behaviour is to be expected.


Pleural effusions associated with pancreaticopleural fistula

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Abstract

Two cases of pancreaticopleural fistula are reported. The delay in diagnosis and extensive investigations performed highlight the need for pleural fluid amylase estimation at an early stage.

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Pancreaticopleural fistula is an uncommon cause of large, recurrent, predominantly left sided pleural effusions. When the underlying pancreatic disease is asymptomatic the diagnosis may not be considered and, as a consequence, extensive unnecessary investigations may ensue. We report two cases in which long delays in diagnosis (four and 13 months) of the pancreaticopleural fistula occurred. These cases emphasise the importance of considering the diagnosis and of estimating the pleural amylase content.

Case 1

A 50 year old man presented to another hospital with exertional dyspnoea and a four month history of weight loss. There was no previous history to suggest pancreatic disease but he drank 40 units of alcohol per week.

Examination indicated a large left sided pleural effusion which was confirmed by chest radiography. The pleural fluid had a protein content of 87 g/l and both cytological and microbiological investigations were negative. The pleural biopsy specimen was normal, as were the results of bronchoscopic examination. A computed tomographic (CT) scan of the chest revealed thickening of the left hemidiaphragm, and a CT scan of the abdomen revealed left para-aortic lymphadenopathy. Ultrasound guided biopsy of the lymph nodes showed reactive changes only. A bone marrow aspirate and trephine sample were normal.

Endoscopic retrograde pancreatogram showing a dilated pancreatic duct (black arrow) distal to a mid duct stricture. A clear leak of contrast from the pancreas can be seen arising from the strictured area tracking towards the pleura (white arrow). Selective cannulation of the pancreatic duct to the level of the stricture was required to show these findings.
Over a 12 month follow up period he had a small persistent effusion. Further investigations were initiated following the development of a right pleural effusion. The pleural fluid was dark and straw-coloured and was assessed for amylase content. Pancreatic hyperamylasaemia (20,000 units/l) was confirmed. Concurrent serum amylase estimation was normal (159 units/l). An abdominal radiograph, upper abdominal ultrasonographic scan, and a CT scan of the abdomen were normal. He was then transferred to this hospital where endoscopic retrograde pancreatography confirmed a pancreaticopleural fistula with stricture of the pancreatic duct (fig). A distal pancreactectomy was performed and the patient recovered uneventfully.

Case 2
A 38 year old man was admitted to another hospital with right sided chest pain and dyspnoea. There was a history of alcohol excess and three proven episodes of pancreatitis over eight years. At the time of presentation there were no symptoms referable to the pancreas. Examination revealed a right pleural effusion which was confirmed by chest radiography.

He had a normochromic normocytic anaemia with normal liver function tests and serum amylase. The pleural fluid was clear and straw-coloured, its protein content was 47 g/l, and both cytological and microbiological investigations were normal. Examination of pleural biopsy samples revealed reactive changes only. A CT scan of the chest and upper abdominal ultrasonography were normal. He was transferred to another hospital for further investigation where pleural fluid hyperamylasaemia (>20,000 units/l pancreatic amylase) was confirmed. Concurrent serum amylase estimation was normal (140 units/l). Abdominal radiography, repeat ultrasonography, and a CT scan of the abdomen confirmed features of chronic pancreatitis. He was referred to this hospital where endoscopic retrograde pancreatography performed four months after the initial presentation confirmed a pancreaticopleural fistula with a proximal pancreatic duct stricture.

Having failed to respond to medical treatment (total parenteral nutrition and octreotide), he underwent pancreaticojugal anastomosis and he remains well without recurrence of the effusion.

Discussion
The two cases illustrate the delay and unnecessary investigations that may occur when the diagnosis of pancreaticopleural fistula is not considered.

Chest radiographic abnormalities occur quite frequently in the course of upper abdominal disease, particularly subphrenic abscess.

Acute pancreatitis may lead to atelectasis, pneumonitis, the adult respiratory distress syndrome, and pleural effusions. Effusions arising from acute pancreatitis are usually small, left sided and self limiting. The incidence of pleural effusions in acute pancreatitis is between 3% and 17%.1

In chronic pancreatitis, as a consequence of fistula and pancreatic pseudocyst formation, extremely large effusions may be seen which frequently recur following drainage. These result from a fistulous tract between the pancreas and the pleural space or by direct extension of a pancreatic pseudocyst through the mediastinum.

Pleural effusions caused by chronic pancreatitis are usually left sided but may be right sided or bilateral. The amylase content tends to be significantly elevated and the effusion is usually an exudate. The pleural amylase level may be elevated in chronic pleural effusions that are associated with an oesophageal perforation or primary lung neoplasms, but in these instances the amylase is of the S (salivary) type rather than the pancreatic (P) type associated with pancreatic disease.

Some 40–50% of patients with chronic pancreatitis and large effusions respond to conservative management with intercostal drainage and total parenteral nutrition, somatostatin or its analogue octreotide.2 Persistence or recurrence of the effusion(s) is an indication for pancreatic surgery. This is dependent upon delineation of pancreatic anatomy, so endoscopic retrograde pancreatography is mandatory prior to surgery. Adequate pancreatography may require selective duct cannulation, particularly in the presence of tight stricture. About 80–95% will have a satisfactory outcome from surgical intervention with a mortality rate of 3%.3 The overall mortality rate from pancreaticopleural fistula is about 5%.2

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