

# Papillary cystic neoplasm of the pancreas: presentation and natural history in two cases

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## Abstract

Two young women, one white and one Chinese, with the rare but increasingly recognised papillary and cystic neoplasm of the pancreas are reported. The initial symptom in both was non-specific abdominal pain which, after investigation, was found to be caused by a pancreatic tumour. One patient did not come to surgery until five years after the initial diagnosis when she developed jaundice. In the five year interval between diagnosis and the development of jaundice computed tomography showed no change in the size (20 cm) of the pancreatic mass. Histology after resection, however, showed signs of lymphatic invasion. Cystic neoplasm of the pancreas can thus be regarded as an indolent, very slow growing tumour with potential for local invasion and hence metastatic spread.

There has been increasing interest recently in the papillary and cystic neoplasm of the pancreas that occurs almost exclusively in young women and was originally described in 1959 by Frantz.<sup>1</sup> This tumour accounts for approximately 2% of exocrine neoplasms of the pancreas. Several authors have recently reported small series and have emphasised the typical features, which are as follows: presentation as a large asymptomatic abdominal mass; young women affected in >90% of cases; a preponderance of non-white patients; radiology showing a well encapsulated, hypovascular, solid and cystic tumour usually in the body or tail of the pancreas; characteristic immunohistochemical staining that can differentiate the tumour from other neoplasms; and a well recognised potential for local invasion and rarely (5% of cases) of metastatic spread.<sup>2-6</sup> The treatment of choice is radical resection.

## Case history 1

A 15 year old schoolgirl presented with a three year history of intermittent epigastric pain radiating to the back. The attacks of pain were of varying periodicity but often occurred post-prandially and lasted one or two hours. There was no history of weight loss or jaundice. Clinical examination showed minimal tenderness in the epigastrium only. Haematological parameters were normal and endoscopy showed no evidence of upper gastrointestinal tract abnormality. An ultrasound examination of the upper abdomen was performed to exclude cholelithiasis and surprisingly showed a 5 cm mass in the region of the body of the pancreas. Further investigations including computed tomography, magnetic resonance imaging (Fig 1), and selective angiography were undertaken. A well encapsulated

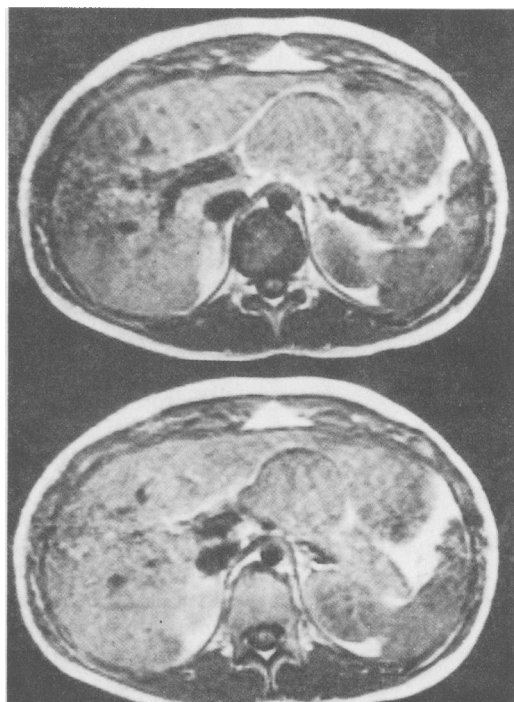


Figure 1: Magnetic resonance images (case 1) showing a well encapsulated mass in the region of the pancreatic tail. The intermediate to high signal from the mass on both the proton density image (above) and the T2 weighted image (below) suggest that it is composed of haematoma or haemorrhagic cystic fluid.

solid and cystic mass was shown. At operation the cystic mass was found to arise from the superior border of the neck of the pancreas with a tumour arising from the posterior abdominal wall. An extended distal pancreatectomy was performed with splenic conservation. The patient made an uneventful recovery and two years later remains asymptomatic.

## Case history

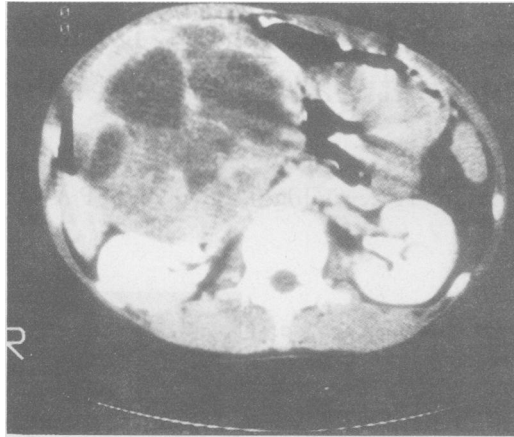
A 19 year old Chinese woman presented with a two week history of obstructive jaundice. Five years previously, at the age of 14, she had been investigated in Hong Kong for bouts of epigastric pain. Computed tomography had shown a mass in the head of the pancreas (Fig 2). Further investigation and treatment had been declined in spite of the presence of a large right upper quadrant mass known to the patient. Serum bilirubin at second presentation in 1990 was 471 mmol/l. Repeat computed tomography showed a well demarcated solid and cystic mass essentially unchanged in size compared with the scan undertaken five years previously (Fig 3). Microscopy of a biopsy needle core specimen showed papillary structures with a fibrovascular core overlaid by pseudostratified regular epithelium. Attempted transpapillary endo-

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*Figure 2: Computed tomogram from 1985 (case 2) showing a mass arising from the head of the pancreas. Mixed attenuation areas are shown, some of fluid density others of solid tissue.*



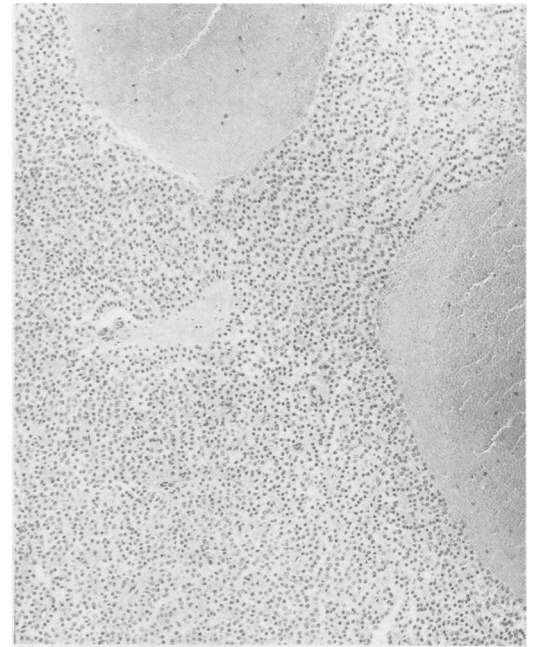
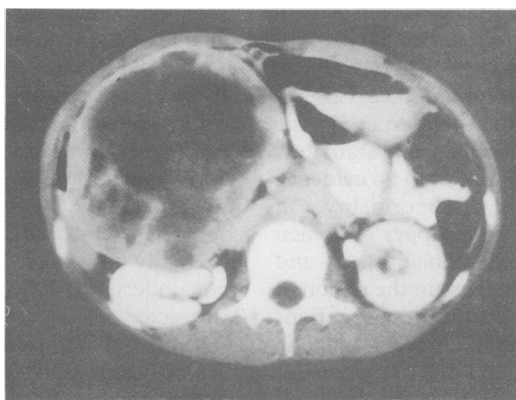
scopic biliary stenting to relieve the jaundice was unsuccessful, because of compression and distortion of the duodenum. Radical resection of the tumour was achieved by pylorus preserving pancreaticoduodenectomy and the patient made an uneventful recovery with rapid relief of jaundice.

Macroscopically, both tumours had a mottled appearance with areas of haemorrhage and cystic degeneration. Microscopy showed islands of monotonous cells with small round nuclei arranged in solid sheets and papillary fashion. The first case was originally classified as a non-functioning islet cell tumour but after wide consultation was shown to have a morphology identical to the solid variant of the papillary and cystic neoplasm described by Frantz (Fig 4). Apart from a very occasional cell expressing glucagon it was negative for hormonal and neuroendocrine markers. The second case was recognised as having the typical histological features of papillary and cystic neoplasm (Fig 5). In addition there was capsular infiltration, duodenal serosal infiltration, and lymphoid (lymphatic vessel) permeation, indicating a degree of local invasion. The cells of this neoplasm expressed neuron specific enolase but not S100 protein or protein gene product 9.5 (PGP9.5).

### Discussion

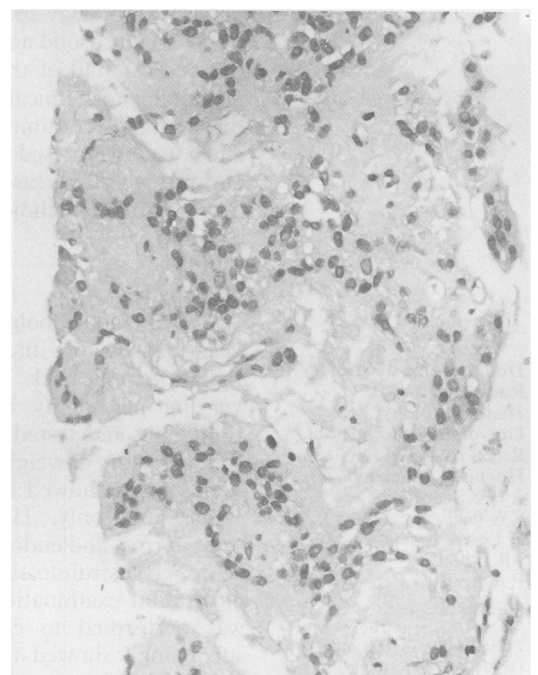
These two case reports add useful information to current knowledge of the natural history of papillary and cystic neoplasm and its optimal management. Although tumour growth may be arrested, apparently for several years as in the

*Figure 3: Computed tomogram taken in 1990 (case 2) showing essentially no change in tumour characteristics compared with computed tomogram from 1985.*



*Figure 4: Photomicrography (case 1) showing uniform cellular composition and large blood filled spaces. Haematoxylin and eosin stain, original magnification  $\times 150$ .*

second case, histological features of local invasion indicate a propensity to dedifferentiate with time and emphasise the necessity of carrying out radical resection. Histological distinction from other tumours of the pancreas is important because this neoplasm has a relatively good prognosis. The first tumour resembled an islet cell neoplasm but showed no evidence of hormone secretion or neuroendocrine differentiation. The second showed the more typical papillary structures of papillary and cystic neoplasm and was associated with lymphatic invasion and capsular permeation. The difficulty in dis-



*Figure 5 Photomicrograph (case2), needle biopsy core specimen showing trabecular arrangement and fibromyxoid stroma. Haematoxylin and eosin stain, original magnification  $\times 375$*

tinguishing between this neoplasm and islet cell tumours has been noted by other authors but can be overcome by identifying cells that express neuron specific enolase and no other neuroendocrine markers. Tissue characterisation studies by magnetic resonance imaging showed blood in the cystic areas, a highly diagnostic feature of papillary and cystic neoplasm and one not previously described.

Because the patients are usually young, operations should include recent modifications of pancreatic surgery which reduce post-operative morbidity, in particular, splenic conservation in distal pancreatectomy to maintain immunocompetence and pylorus preservation in pancreaticoduodenectomy to avoid dumping and diarrhoea associated with gastrectomy and enabling the patient to return to their weight before the illness.<sup>7 8</sup>

Papillary and cystic neoplasm is a rare and curable pancreatic tumour which requires sophisticated techniques for radiological and histological diagnosis and which should be treated by radical excision.

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