Acta Chirurgica Belgica

ISSN: 0001-5458 (Print) (Online) Journal homepage: https://www.tandfonline.com/loi/tacb20

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To cite this article: T. Darius, J. Brouwers, H. Van Dijck & P. Bernard (2006) Solid and Cystic Papillary Neoplasm of the Pancreas: A Rare Tumour in Young Women, Acta Chirurgica Belgica, 106:6, 726-729, DOI: 10.1080/00015458.2006.11679994

To link to this article: https://doi.org/10.1080/00015458.2006.11679994

Acta chir belg, 2006, **106**, 726-729

Solid and Cystic Papillary Neoplasm of the Pancreas : A Rare Tumour in Young Women

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Key words. Solid cystic papillary neoplasm ; pancreatic tumour ; Frantz's tumour.

Abstract. In this paper, we present an asymptomatic young patient with solid and cystic papillary neoplasm of the pancreas. It is an extremely rare tumour mostly seen in young females. It is often diagnosed incidentially or during investigations of gastrointestinal complaints. In differential diagnosis, any cystic and/or solid pancreatic disease process should be considered. Prognosis is excellent after radical resection and recurrence is rarely seen.

Introduction

Solid and cystic papillary neoplasm (SCPN) of the pancreas is an extremely rare paraneoplasm of the pancreas that normally occurs in young females. It was first described in 1959 by FRANTZ (1). Various synonyms include papillary cystic neoplasm, papillary epithelial neoplasm, papillary and cystic tumour, papillary and cystic epithelial carcinoma, papillary and solid neoplasm, solid and cystic acinar cell tumour and Gruber - Frantz's tumour (2). For this reason the real incidence of SCPN of the pancreas is diffucult to assess.

We present a young female in whom PSCN of the pancreas was incidentally found and a review of the current managament.

Case Report

A 19-year old woman had an acute onset of right fossa iliaca pain. There were no other symptoms. Clinical examination of the patient revealed mild abdominal tenderness in the right lower quadrant without palpable masses. Complete blood count, liver function tests and renal function tests were normal. CRP was 2,28 mg/dl.

Ultrasonography (US) of the abdomen showed a right ovarial cyst, but also a large mass localized at the tail of the pancreas, adherent to the spleen. Computer tomography (CT) revealed a $9 \times 6{,}5 \times 5{,}5$ cm mass involving the tail of the pancreas, with no evidence of invasion into surrounding tissues. Because there was no strong evidence for malignancy and the fact that she was doing her exams at the university, further investigation and possible surgical treatment was delayed. The right fossa iliaca pain disappeared spontaneously.

A computer tomographic reevaluation 2 months later was identical. A magnetic resonance scan was suggestive for a solid and cystic papillary neoplasm (SCPN) of the pancreatic tail. Again, there was no evidence of metastasis, no invasion into surrounding tissues, no pathological lymph nodes were visible (Fig. 1).

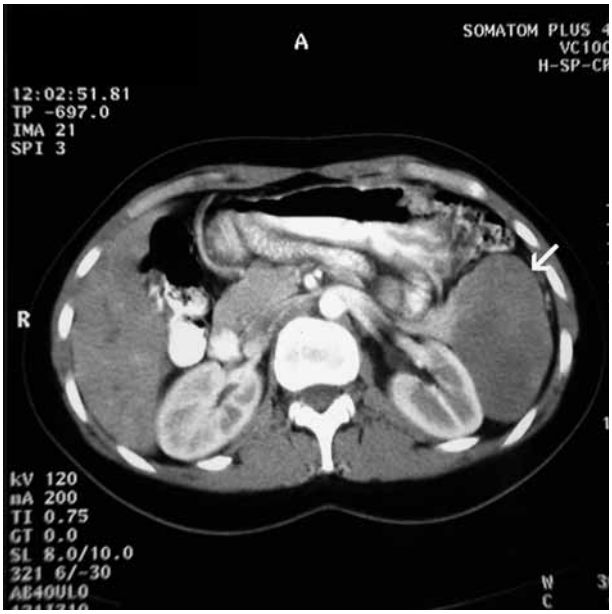

Fig. 1
CT scan showing the SCPN on the tail pf the pancreas (arrow)

A surgical intervention showed a well circumscribed mass involving the distal end of the pancreatic tail. The splenic vein was attracted into the mass but was easily dissected free from the tumour (Fig. 2). A limited distal pancreatectomy was performed by scalpel, with a surgical margin of 1 cm of the tumour. Afterwards, a manual suture of the ductus and parenchyma was done. A tube drain was left in place at the distal pancreatectomy loge.

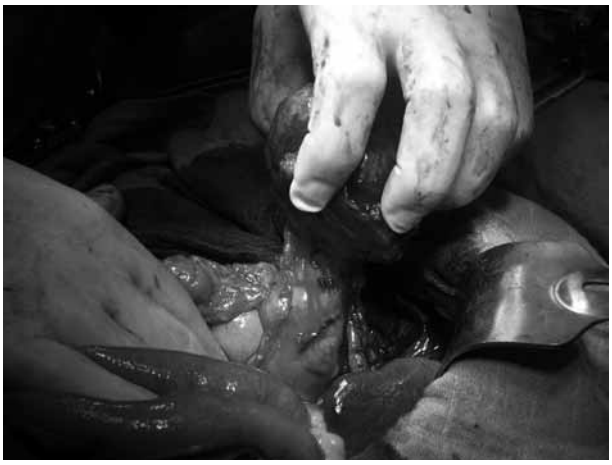

Fig. 2
Intraoperative finding of a well encapsulated mass originating from the head of the pancreas in which the splenic vein is attracted.

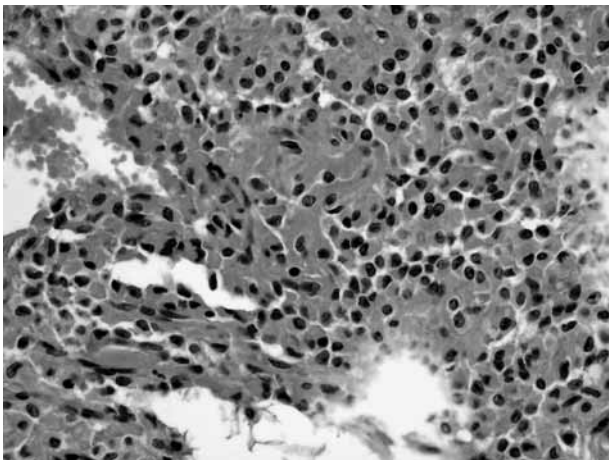

Fig. 3
A micrograph showing cystic degeneration with solid and pseudopapillary formations.

The tumour measured $10 \times 6 \times 5$ cm and was well encapsulated. Histopathological examination of the resected tumour was consistent with SCPN of the pancreas (Fig. 3).

On the seventh postoperative day, a minor pancreatic leak was diagnosed by the tube drain in the lesser sac, confirmed by biochemical evaluation of the fluid. There was a spontaneous resolution after treatment with long acting ocreotide (Somatuline, 90 mg SC), started immediately after diagnosis of the pancreatic leak. The patient was discharged on the thirteenth postoperative day.

Discussion

A solid and cystic papillary neoplasm of the pancreas is an extremely rare tumour of the pancreas. It is considered as a low-grade malignant tumour with an excellent prognosis (3-7). It accounts for only 1 to 2% of exocrine pancreatic tumours, and occurs predominantly in young females (8). In only 7%, men are affected, but generally they are 10 years older than women (6) and it is also exceptionally rare in children (6, 7, 9). The tumour can occur anywhere in the pancreas and frequently shows an exophytic growth (3). In approximately 15% of the cases metastases occurred (8). Metastases involved one or more of the following organs : lymph nodes, liver, spleen, colon, mesocolon and generalized carcinomatosis (10).

Clinically, the tumour causes few symptoms. SCPN of the pancreas are frequently diagnosed during investigation of gastrointestinal complaints such as abdominal pain, anorexia, weigth loss, nausea and vomiting or abdominal masses, in case of large tumours, or they are incidentally found in smaller tumours (50% of the cases) (1). Obstructive jaundice is seen in case of tumour location in the head of the pancreas (11). It may rarely present as an acute emergency due to rupture and haemoperitoneum. Even more unfrequently, it presents with an acute pancreatitis or as a posttraumatic pancreatic pseudocyst (12, 13).

Radiologically, US and CT scan can be effective means to suspect SCPN.

It is mostly heterogeneous on imaging, with varying proportion of solid and cystic areas. The cystic areas are due to the presence of haemorrhage and necrosis. The solid component of the lesion may show papillary projections in the cystic component, which may appear like septations. Calcifications are reported in some cases and may occasionally be seen on plain X-ray films (2, 14). On US, the neoplasm is isoechoic or hypoechoic relative to surrounding normal pancreatic parenchyma. Few small echo-free areas may be seen. On CT, the lesion is isodense or hypodense relative to normal pancreas on both non-contrast and contrast enhanced images. Few cystic aereas may be present. The thin capsule, which is difficult to evaluate on US, may be seen on CT scan as a hypodense, enhancing area. On MRI, it typically presents as a large, well-defined, encapsulated lesion with heterogeneous high or low signal intensity on T1, heterogeneous high signal intensity on T2-weighted, and early peripheral heterogeneous enhancement with progressive fill-in on gadolinium-enhanced dynamic MR imaging (15). Fine-needle aspiration can be used to obtain a preoperative cytological diagnosis, but is not necessary (10, 16).

In differential diagnosis, any cystic and/or solid pancreatic disease process should be considered. This includes inflammatory pseudocyst, mucinous cystic tumours, mucussecreting tumours, microcystic adenoma, islet cell tumour, acinar cell carcinoma, cystadenocarcinoma, pancreaticoblastoma, and vascular tumours

as haemangioma, lymphangioma and angiosarcoma. An inflammatory pseudocyst usually occurs after abdominal trauma or pancreatitis. Endocrine tumour occurs at a slightly older age than SCPN of the pancreas and is with no gender predilection. Pancreatoblastoma is a childhood malignant pancreatic neoplasm with poor prognosis and has a male predominance. Acinar cell tumours are always malignant and affect patients of both sexes in their sixth or seventh decades (1, 10).

Radical resection, where technically feasible, should be considered the therapy of choice as it is a safe and effective control of the disease. There is no role of non-operative management. The localization and presence of local invasion affect the surgical management (1). The various surgical procedures are Whipple's operation, pylorus preserving pancreaticoduodenectomy, distal pancreatectomy with or without splenectomy, enucleation and excision. Liver metastasis should be treated with resection (5). The role of neoadjuvant chemotherapy is described in only a few case reports. It was done in case of advanced disease with invasion of the superior mesenteric vein with good response (17-18).

Complete surgical removal of the tumour even in case of metastases or local invasion offers an excellent prognosis (4-5, 11, 19-20). Recurrence is rarely seen and should be treated by radical resection.

To the pathologist, SCPN represents a challenge. The light-microscopic features are characteristic and generally do not present diagnostic problems. Microscopically the tumour shows a mixture of papillary and solid patterns. The papillary structures are the fibrovascular stalks surrounded by tumour cells. The tumour cells are small, uniform cells with eosinophilic granular cytoplasm. The cystic areas are secondary and correspond to the abundant necrotic material, blood, cholesterol crystals and foam cells (1) (Fig. 3).

Although on gross examination degenerative cystic changes may lead to confusion with cystic neoplasms of the pancreas. SCPN are typically positive for vimentin, neuron-specific enolase (NSE), alfa1-antitrypsin, and alfa1-antichymotrypsin and are negative for chromogranin, epithelial membrane antigen, and cytokeratin.

Microscopic examination reveals a variety of patterns (solid, pseudopapillary, cystic, pseudomicrocystic, trabecular). Confusion may result because of the resemblance to other pancreatic neoplasms. The cells are polygonal to elongated with ovoid nuclei that are grooved or indented. Mitotic figures are rare. The cytoplasm ranges from clear to eosinophilic. Vimentin expression is common. Keratin is occasionally expressed in patchy fashion. Alfa1-antitrypsin, and alfa1-antichymotrypsin are frequently demonstrated in small, eosinophilic PAS-positive cytoplasmic globules (21).

Differentiation along endocrine cell lines has been postulated for this tumour, on the basis of NSE positivity, but the expression of vimentin and alfa1-antitrypsin does not support this interpretation. Because the pseudopapillary pattern is sometimes mistaken for a trabecular architecture and because the tumours are NSE positive, SCPN may be mistaken for pancreatic endocrine tumours as mentioned before (8). Recent reports suggest that SCPN originates from pluripotent stem cells (22).

In conclusion, SCPN is a rare low-grade malignant tumour usually seen in young women with few clinical symptoms. Radical resection is the treatment of choice and offers an excellent prognosis even in case of metastases or local invasion.

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