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PAPILLARY-CYSTIC TUMOUR OF THE PANCREAS

Case Report

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Abstract. A papillary-cystic pancreatic tumour was excised in a 19-year-old girl, who was apparentely free from recurrence 20 months later. Despite morphologic indications of malignancy, the prognosis after surgical removal of this uncommon tumour is good. Its origin probably is the epithelial cells of the small pancreatic ducts. The case is described and the literature is reviewed.

Key words: pancreas, papillary-cystic neoplasm.

Cancer of the pancreas, wich was first described in 1836, ranks fourth among fatal cancers in men and fifth in women (14). Ductal adenocarcinoma is by far the most common form, accounting for more than 80% of tumours arising in the pancreas. The peak incidence of pancreatic adenocarcinoma is in the fifth and sixth decades of life, and the 5-year survival rate is less than 5%.

In contrast to this common neoplasm, a malignant pancreatic tumour with much more favourable prognosis has recently been described. It has been termed 'papillary-cystic' or 'solid and papillary', and approximately 80 cases have been reported. We now present a case of this uncommon ans as yet not widely known pancreatic neoplasm.

CASE REPORT

A 19-year-old white girl was admitted to this hospital in July 1985. She had been in good health until one month previously, when a tender mass developed in the right upper quadrant of the abdomen. The mass became so conspicuous that it deformed the right costal arch. The patient denied weight loss, fever, jaundice, trauma, dyspepsia, abdominal pain or symptoms indicative of hypoglycaemia. All laboratory findings were normal.

Abdominal sonography and a computed tomographic scan (Fig. 1) showed a large cystic mass in the body of the pancreas. The mass displaced the stomach forwards and upwards, causing partial obstruction of the splenomesenteric vessels. At laparotomy a well-encapsulated, highly vascularized tumour was found in the pancreatic body. The liver appeared free from metastases and there was no sign of local invasion or regional lymph-node involvement. The tumour was enucleated in toto from the pancreatic parenchyma.

Macroscopically the tumour was a large (15x9 cm) cyst, weight 250 g, with a well-defined fibrous capsule. Some vascular ectasia was clearly visible on the outer surface of the capsule. The cut surface showed cystic degeneration, necrosis and areas of haemorrhage with abundant blood clots. The appearance thus was variegated, with reddish-brown as the predominant colour.

Microscopy showed the tumour to be composed of papillary and solid areas intermingled with microcystic structures which were separated by fibrous tissue stalks. In addition there were large haemorrhagic and a few necrotic areas. The papillary areas consisted of fibrovascular stalks with epithelial covering comprising a few layers of small and uniform cells, ranging from columnar to cuboidal. The nuclei, as a rule located at the cell base, were vesicular, oval or round. The nucleoli were prominent. The cytoplasm was eosinophilic, sometimes granular. Many cells showed degeneration and their cytoplasm was clear and hence inhomogeneous. The solid areas were characterized by the same type of cells, which were arranged like pseudorosettes around thin vascular cores. The microcystic areas consisted of slightly basophilic substance of mucoid appearance. Mitotic figures were rare. The surrounding pancreatic tissue displayed compression and was not infiltrated by tumour cells (Figs. 2, 3).

The patient is well 20 months postoperatively, and without evidence of recurrence at ultrasonography, computed tomography, laboratory tests (carcinoembryonic antigen) and physical examination. She has received no other treatment.

CONCLUSIONS

Papillary-cystic tumour of the pancreas was first described by Frantz in 1959 (7). The number of hitherto reported cases is around 80 (13). The tumour is most common in girls and young women

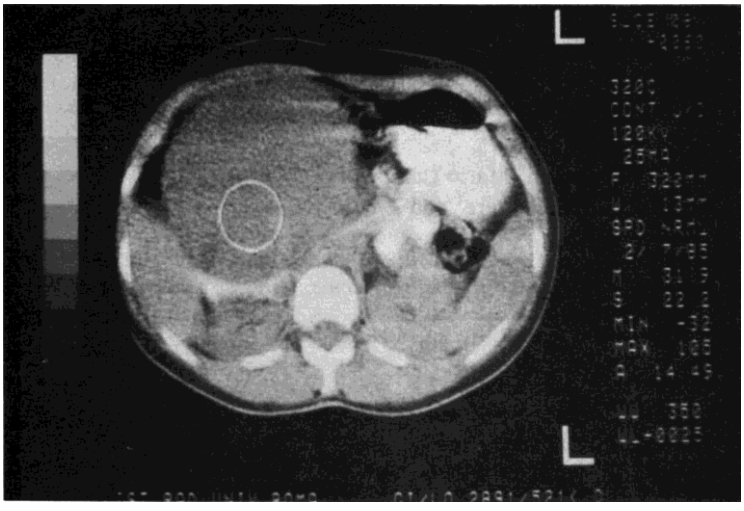

Fig. 1. CT scan showing a large cystic mass in the body of the pancreas.

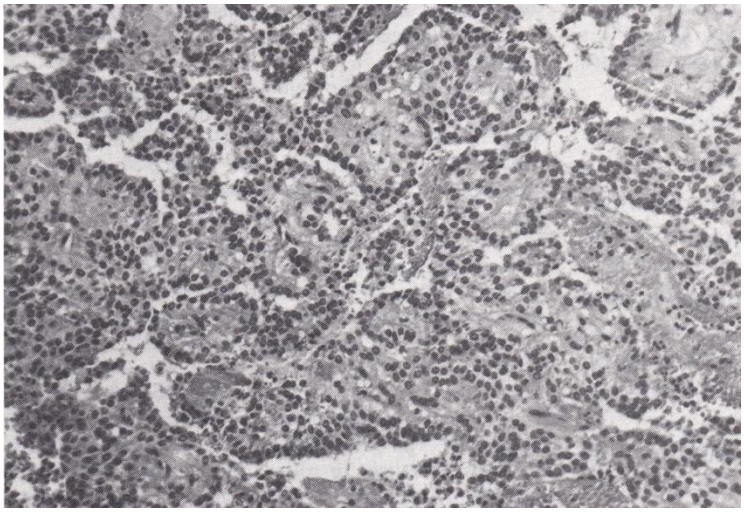

Fig. 2. High-magnification detail of the papillary structures with some foci of necrosis.

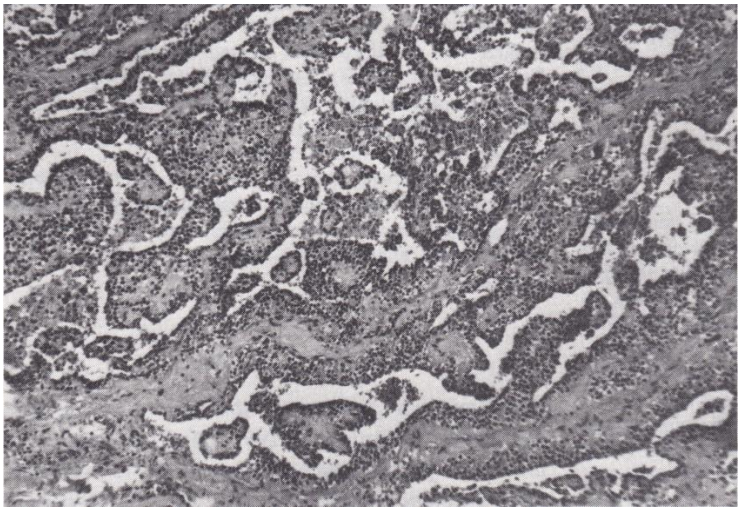

Fig. 3. Papillary-cystic configuration of the neoplasm.

(13, 15), particularly black females (4), aged 12 to 33 years (mean age in reported cases 24 years). They are usually taller and heavier than their contemporaries, which may suggest involvement of hormonal factors in the pathogenesis (5, 6).

Symptomatology is shifting. The patients complain of epigastric discomfort (6) which progresses to dull pain, with dyspepsia and weight loss in rare cases (13). Generally, however, the presenting manifestation is an abdominal mass, as a rule in the left upper quadrant, which moves minimally with breathing. The tail of the pancreas is the usual site in about c.90% (14).

When the tumour presents as an emergency, the cause is believed to be severe bleeding due to its high vascularity (3).

Although opinion concerning the origin of the papillary-cystic tumour is not unanimous, it is mostly accepted to be in the epithelial cells of the small pancreatic ducts. Distinction of this neoplasm from endocrine pancreatic tumours, pancreatoblastoma and cystadenocarcinoma is very important. Immunohistochemical studies showed no staining indicative of pancreatic hormones, thus excluding an endocrine origin (15). The absence of squamous epithelial nests and sarcomatous components distinguishes the papillary-cystic neoplasm from pancreatoblastoma or infantile pancreatic carcinoma (5, 6, 7). Finally, papillary-cystic tumour of the pancreas differs from cystadenocarcinoma, which is characterized by high cellular pleomorphism and usually occurs at older ages (11).

This papillary-cystic tumour is considered to be of low-grade malignancy (14). The rarity of metastatic spread was illustrated by a series of 52 cases (4) in which there was only one death from liver metastases, while the other patients were alive and without clinical recurrence 7 years postoperatively.

The treatment of papillary-cystic tumour of the pancreas must be surgical, using a Whipple procedure if the location is the head of the gland, or pancreatic resection if it is in the body or tail. The prognosis appears to be excellent.

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