Three cases of solid cystic pancreatic neoplasm

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Introduction

Three cases of a rare pancreatic tumour in young patients are presented and the management discussed. Compared to adenocarcinoma of the pancreas solid cystic neoplasms carry a good prognosis despite their size and local infiltration if aggressively resected surgically.

Case 1

A 24-year old woman was seen at the surgical clinic with abdominal pain and a large mass in the epigastrium. She gave a history of previous laparotomy 2 years ago but no further details were available. She had no gastrointestinal symptoms. A right subcostal scar was present and palpation showed an epigastric mass, which measured 15 × 6 cm. She had elevated direct serum bilirubin and alkaline phosphatase levels. The serum amylase was

normal. Ultrasound scan of the abdomen revealed a 17 × 7 cm mass in the head of the pancreas with solid and cystic components. The common bile duct was 1.5 cm in diameter and the liver was normal. At laparotomy, there was a large mass arising from the head and the body of the pancreas adherent to the under surface of the left lobe of the liver. The supraduodenal common bile duct was dilated. The stomach, small intestine and transverse colon were normal. A Whipple pancreatico-duodenectomy was carried out leaving a small part of the pancreas with the spleen. The retropancreatic portal vein was infiltrated by the mass and was removed with the pancreas.

Reconstruction of the portal vein was done using an end-to-end Gortex graft 10 mm in diameter with a fine Prolene (Ethicon) continuous suture. Biliary and gastric continuity was established with a roux-en-y jejunal loop. The remaining tail of the pancreas was left without reconstruction

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because there was no duct seen and it was oversewn with a fine absorbable suture. Blood loss was 2 litres. She was managed in the intensive care for 3 days.

Endocrine pancreatic function is normal at follow up. She is dependent on exogenous exocrine pancreatic supplementation. At six-monthly follow up she remains well with no evidence of recurrence on ultrasound scan. She came to our clinic 3 years after surgery for referral to the obstetricians for antenatal care and underwent uneventful elective caesarian section.

Case 2

A 26-year old man came to our clinic with an 8-month history of epigastric pain and discomfort. On examination he had a hard tender non-pulsatile, non-mobile multilobulated epigastric mass. The rest of the examination, including rectal examination, was normal. An ultrasound examination and a CT scan (Figure 1) showed a large cystic pancreatic mass with a solid component involving the body and tail sparing the head. At laparotomy there was a solid and cystic tumour of the distal half of the pancreas. This was attached to the spleen. A distal pancreatectomy with splenectomy was performed. The mass contained over four liters of cystic fluid. His post-operative recovery was uneventful. At follow up a year later he remains well. CT scan a year later showed no recurrent masses in the remnant pancreas.

Figure 1. Pre-operative CT scan of upper abdomen showing a large mass in the region of the tail of the pancreas.

Case 3

A 14-year old girl was seen as an emergency with clinical features of obstructive jaundice and a large mass in the epigastrium. She had a raised serum bilirubin, alkaline phosphatase and a prolonged prothrombin time. Ultrasound scan of the abdomen showed a 12 cm x 8 cm

mass in the head of the pancreas, with both solid and cystic components. The common bile duct was dilated. She was given systemic vitamin K and prepared for surgery. At laparotomy, a mass with solid and cystic components was seen in the head of the pancreas. Intra-operative fine needle aspiration was done and a report obtained immediately revealed a cellular smear consistent with papillary neoplasm. With difficulty a Whipple pancreatico-duodenectomy was performed. The tumour was shaved off the retro-pancreatic portal vein. Blood loss was 2.5 litres. Post-operatively she required 4 days of intensive care including ventilation. The rest of her stay was uneventful, and she was referred to the oncology unit where chemotherapy was administered using weekly 5-fluorouracil over a 12-week period. She has been followed up for 3 years with no clinical and ultrasound evidence of recurrence. She had one admission 2 years later for subacute intestinal obstruction, which settled in 2 days with conservative measures.

Macroscopically the neoplasm showed a solid tumour with several areas of cystic degeneration and haemorrhage. Histology showed a tumour composed of relatively uniform small cells with no cellular atypia. The cells were arranged in solid sheets and in a pseudopapillary pattern. Foci of necrosis and hemorrhage were evident. Appearances were characteristic of a solid-cystic papillary neoplasm of the pancreas.

Discussion

All three patients had large pancreatic masses consisting of both solid and cystic components. Two underwent pancreatico-duodenectomy with reconstruction. The third underwent distal pancreatectomy. All three operations were difficult and time consuming (>4 hours) with much blood loss (average of 2 litres). All three cases required intensive care management. The histology in all 3 patients showed a papillary cystic neoplasm. The margins appeared free of tumour and the lymph nodes showed no tumour deposits. All 3 patients attended for regular follow up and are free of recurrence. The patient with portal vein reconstruction showed no clinical evidence of portal hypertension, and none of the patients is diabetic. The patient with near-total pancreatectomy requires daily oral pancreatic supplements.

Papillary cystic neoplasm of the pancreas is a rare tumour (1). These tumours have been classified in the past as non-functioning islet cell tumour or carcinoma, acinar cell carcinoma, papillary cystadenocarcinoma, or pancreatoblastoma (1). Microscopically the tumour shows cystic and solid areas lined by mucinous cylindrical type of epithelium (2,3). These tumours are unpredictable and though histology may be innocuous, they can be locally aggressive. There is some evidence that these tumours are hormone dependent with progesterone receptor positivity being recorded (4). Immunochemical and electron microscopic studies suggest that this tumour arises from primordial or multipotential stem cells capable of differentiating into both exocrine and endocrine lines (1).

The typical presentation is a young female (5) with a large epigastric mass, abdominal pain and loss of weight (6). Obstructive jaundice is present when the tumour involves the head of pancreas and the retropancreatic common bile duct. These tumours are locally aggressive with a low grade malignancy, and are best treated by aggressive resection and debulking (6, 7). There is a favourable outcome after resection with long term survival of more than 5 years reported (2, 6, 7). One of our cases had an advanced tumour with involvement of the portal vein. Aggressive resection of the mass with portal vein reconstruction was possible. These patients are young and tolerate long complex surgery. The prognosis after resection is good compared to adenocarcinoma of the pancreas. In one series 11 of 12 patients survived a mean of 6.6 years (range 6 months to 15 years) (6).

Papillary cystic neoplasm should be considered in the differential diagnosis of large cystic pancreatic masses, especially in young females.

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