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Case Report

Frantz' Tumour: A Rare Pancreatic Neoplasm

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Abstract

Frantz' tumour of the pancreas is also known as solid pseudopapillary tumour (SPT) of the pancreas. It is a rare pancreatic neoplasm and represents about 3% of all the pancreatic cystic neoplasm. It occurs predominantly in young woman in 2nd to 3rd decade of life. These tumours exhibit indolent behaviour and very often reach considerable size before the first symptoms appear. Despite this presentation these tumours have low malignant potential and complete surgical resection render excellent prognosis. We reported a case of a 16-year-old girl who presented with upper abdominal mass with symptoms of gastric outlet obstruction for 7 months duration. Clinical examination revealed a huge epigastric mass measuring 10 x 12 cm in size. CT scan showed presence of mass arising from the body of the pancreas which was hypervascular, well-encapsulated with mixed cystic and solid components. She then underwent successful distal pancreatectomy and splenectomy and recovered uneventfully.

Keywords: Gastric outlet obstruction, neoplasm, pancreas, pancreatectomy, splenectomy

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Introduction

Solid Pseudopapillary Tumour (SPT) of the pancreas accounts for about 3% of the pancreatic cystic tumours (1). The first published description of pseudopapillary tumour was by Frantz in 1959 (2). This lesion is also named as Frantz tumours, solid and papillary tumours, papillary cystic tumours, solid cystic tumours or Hamoudi tumours. It usually occurs in female in 2nd and 3rd of life and the most common location of the tumour is at the head of pancreas (3). Unlike ductal adenocarcinoma of the pancreas, SPT rarely causes obstructive jaundice. The most common presentations are due to mass effect to the surrounding viscera such as early satiety and vomiting.

Case Report

A 16-year-old female presented with epigastric discomfort for the past 7 months associated with early satiety and postprandial emesis. She denied any symptoms of jaundice and complained of weight loss of 10 kg in 7 months duration. Physical examination revealed mild epigastric tenderness and a palpable mass about 10 x 12 cm in size.

Routine laboratory test results were unremarkable and tumour marker levels (carcinoembryogenic antigen, alpha-fetoprotein, CA 19-9) were within normal limits. In view of the symptoms of gastric outlet obstruction, oesophagogastroduodenoscopy (OGDS) was then

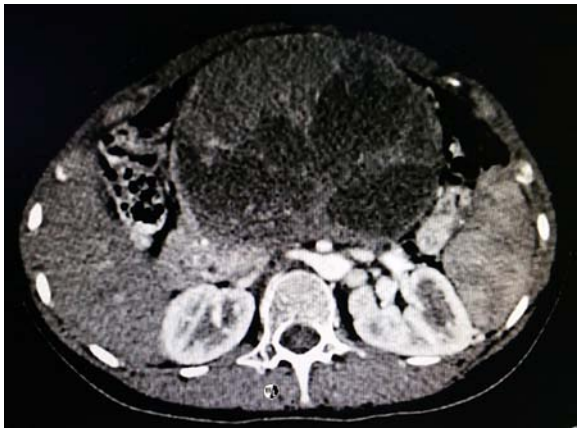

Figure 1: Computed tomography showed pancreatic lesion with mixed solid cystic component involving the body and tail of pancreas likely represent solid pseudopapillary tumour with left sided portal hypertension.

Figure 2: Resected specimen consisted of tumour of the distal pancreas and spleen

performed and revealed external compression to the body of stomach causing pylorus narrowing. CT scan showed a well encapsulated pancreatic body mass with mixed cystic and solid component causing compression to the surrounding structures with splenomegaly (Fig. 1). Celiac angiography revealed no invasion of vasculatures by the tumour.

She underwent distal pancreatectomy and splenectomy (Fig. 2). Laparotomy revealed a tail of pancreas mass with no surrounding infiltration except at the portal vein where tumour was shaved off .There was no evidence of intra-abdominal metastasis.

Her postoperative recovery was uneventful and she was discharged after 1 week of admission. Histopathology review of the pancreatic mass revealed a well-circumscribed mass with capsular invasion. Cut section shows a heterogenous, necrotic and haemorrhagic surface with central cavitation and clear resection margin. Histopathologically it showed encapsulated mass with sheets of tumour cells within

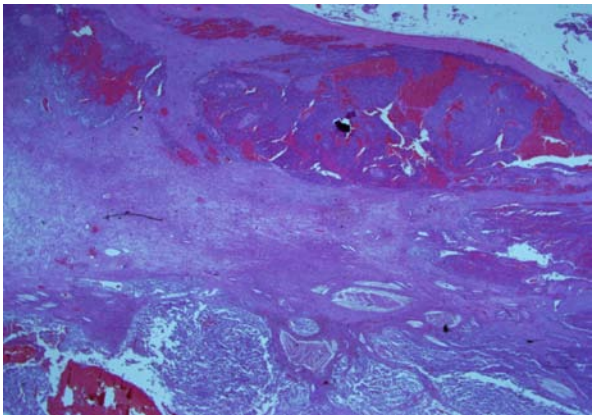

Figure 3: Histopathological picture shows encapsulated mass with sheets of tumour cells within.

(Fig. 3). The presence of pseudo papillary structure confirmed the diagnosis of solid pseudopapillary tumour. There were no pleomorphisms or mitotic activity seen to suggest benign behaviour. Immunohistochemistry test were positive for vimentin and CD10. The resected margins are clear except for portal vein site which is near margin less than 0.1 mm.

Discussion

Solid pseudopapillary tumour (SPT) is believed to arise from ductal or acinar origin. It is a tumour with low malignant potential. Most common sites were at the tail of pancreas (35.9%) as presented in our patient, followed by the head (34%) and other parts of pancreas (3). This rare SPT usually affects female patients in the second or third decades of life. However, our patient presented at earlier age which was 16 years old.

Lack of specific clinical symptoms is typical of this tumour as in the case presented. In the past, approximately 80% of patients with SPT presented late secondary to compressive symptoms. However, incidental detection of SPT is becoming more common with widespread use of cross sectional imaging, and it now accounts for up to 50% of cases. Patients commonly presented with nonspecific symptoms of abdominal pain, followed by nausea, vomiting, and weight loss (4). Other symptoms that occur less frequently include gastrointestinal obstruction, anemia, jaundice, and pancreatitis. Patients may also present with a palpable mass. Our patient had significant weight loss as the result of compression of the pancreatic mass to the stomach causing early satiety and reduce oral intake.

The finding of a mixed solid and cystic pancreatic lesion on cross sectional imaging in a young woman

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should raise the suspicion of a SPT. The lesions may appear as well demarcated solid tumors in Magnetic Resonance Imaging (MRI) (5). The tumour is usually found to be a vascular or hypovascular from arterial phase of CT, which contradicted with the finding in this case hence celiac arteriography was performed to delineate the vasculatures prior to surgical intervention (3). The arteriography was performed in this case to look for any neovascularization due to enlarged tumours, possible major vessels thrombosis like gastroduodenal artery or pancreaticoduodenal arteries causing formation of collateral vessels as well as to determine for any possibility of angioembolization to shrink the tumour before operation. In this case the angioembolization was not performed because there was no main feeding vessel to supply the tumour.

Although endoscopic ultrasound (EUS) with fine needle aspiration (FNA) can be used as an adjunct to achieve confirmatory diagnosis, we obviate the intention in this case as the clinical presentation correlates well with the radiological finding and histological diagnosis will not change the treatment option. Furthermore, nature of this low malignant lesion and potential of seedling during FNA should be weighted (6). According to the 2015 guideline from the American Gastroenterological Association (AGA), EUS is not recommended if the lesion is less than 3 cm with no evidence of ductal dilation or solid component within the cyst. The typical appearance of SPT on endoscopic ultrasound (EUS) is a well demarcated, hypoechogenic, solid cystic appearing mass and irregular calcifications occasionally present in up to 20 percent of cases (7).

The malignant potential of solid pseudopapillary neoplasms has not been well studied. However, tumour size $\geq$ 5 cm was associated with an increased risk of high grade malignancy. Given the lesion's malignant potential, combined with its good prognosis if resected, the finding of a pancreatic mixed solid and cystic lesion in a young woman on CT or MRI should lead to the suspicion of SPT until proven otherwise.

The type of operations depends mainly on the location of the tumour. Pylorus preserving pancreaticoduodenectomy or classical Whipple procedure are the operations for SPT at the head; and distal pancreatectomy with or without splenectomy for tumours distal to neck of the pancreas. Resection of only midportion of the pancreas leaving the head and the tail of pancreas can be done for tumours at the body of pancreas (3).

We performed distal pancreatectomy with splenectomy because this tumour has risk of malignant transformation thus we needed to ligate the splenic vessels to facilitate resection with possible free margin at the pancreatic bed. Furthermore, splenic preservation is technically difficult especially in long standing large tumours. The patient was preoperatively vaccinized and consented for splenectomy prior to the operation.

Unlike the criteria for resection of pancreatic adenocarcinoma, invasion to portal vein and superior mesenteric artery are not an absolute contraindication for resection as options like portal vein resection with reconstruction or tumour shavings from these vessels are acceptable (3).

We decided to shave the tumour from the portal vein rather than portal vein resection and reconstruction because resection of this vein carries higher morbidity especially ischeamic hepatitis or superior mesenteric vein thrombosis. Even patients with residual disease or metastases have been reported to have long-term survival following surgical treatment.

The role of chemoradiation therapy is very limited due to its poor response. It is only reserved to those unresectable tumours. Inpatients with liver metastasis, some reports advocated for Trans Arterial Chemo Embolization as palliative role of therapy (3).

The cut section of SPT macroscopically will show a well encapsulated tumour with solid and cystic compositions. Some areas of haemorrhages and central necrosis will be seen. The main character of the tumour is the presence of both solid and pseudopapillary structures.

Microscopically, the tumour exhibits characterictic branching papillae with myxoidstroma. Special stains, including vimentin, CD10 and betacatenin may be used to differentiate an SPT from a pancreatic neuroendocrine tumour (3).

SPT is regarded as benign tumour with low malignant potential and less than 5% risk of local recurrence (8). The tumour is usually operable at presentation unless it shows local vascular invasion or distant metastases, usually to the liver and peritoneum (3).

Conclusion

In summary, SPT of the pancreas should be treated surgically with curative intent as these tumour carries risk of malignant potential. Furthermore, the risk of recurrence after resection is very low and it carries good long term prognosis.

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