Solid Pseudopapillary Tumour of Pancreas: A Report of 5 Cases

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Abstract
A solid pseudopapillary tumour of the pancreas (SPT) is a rare neoplasm accounting for less than 2% of exocrine pancreatic neoplasms. SPT occurs in adolescent young females and is mostly benign. It is a low-grade malignant tumour that may evolve years before symptoms start and has a favourable prognosis.1,3,5 In this report we present five cases (four females, one male, aged 16, 45, 23, 17 and 55 years, respectively) of SPT localised in the pancreas, and discuss the clinical, imaging and histologic findings with a review of the literature. We retrospectively reviewed these five patients with SPT who underwent surgical resection in our hospital with a definitive histologic diagnosis of SPT.

Introduction
SPT of the pancreas is an indolent exocrine pancreatic tumour which is well-known for its predilection for young women.1,2,4 This rare entity was first described by the author Dr. Frantz in 1959, and was called “papillary tumour of the pancreas- benign or malignant?”3 These neoplasms account for 1-2% of exocrine pancreatic neoplasms.3,6 In 1996, the WHO renamed this tumour as SPT for the international histological classification of tumours of the exocrine pancreas.1,3,6 We discuss diagnostic and differential diagnostic features of this unusual tumour and present five cases seen in our department.

Case Reports
We retrospectively reviewed the clinical data of five patients with histopathologically proved SPT at our hospital from August 2005 to August 2011. Among these, four were females-aged 16, 45, 23 and 17 years respectively and one was male-aged 55 years. Tumour specimens were confirmed on histopathology by at least two pathologists.

Clinical Features
The detailed clinical features of all these cases are as follows
The first case was a 16 year old female who presented to our hospital with the complaints of pain in the abdomen, nausea and vomiting. Her serum amylase was 2099 U/L (normal 22-80 U/L) and lipase 60 U/L (normal 21-67 U/L) preoperatively. The level of amylase fell to 449 U/L on first post operative day. On CT scan and ultrasound examination a 6 x 6 x 5 cm solid well defined round mass with a few cystic areas was visualised in distal body and tail of the pancreas and a radiological differential diagnosis of mucinous cystadenoma, islet cell tumour was kept. The ultrasound and C.T. scan pictures of this patient are given in figures 1 and 2. Distal pancreatectomy and splenectomy were performed.

The second case was a 45 year old female who again presented with abdominal pain and vomiting for 5 days. Her serum amylase and lipase levels were normal i.e. 56 U/L and 12 U/L respectively. On radiological examination a heterogeneous hypodense mass measuring 15 x10 cm was seen in the distal body of pancreas which was adherent to the spleen. Pancreatectomy and splenectomy were done.

The third patient was 23 year old female who came to our hospital with pain in the abdomen for 15 days. Her serum amylase levels were high i.e. 993 U/L while the lipase levels were within normal limits i.e. 47 U/L. On ultrasound and CT scan a large complex mass lesion with enhancing solid component was seen in the pancreas which measured 8 x 3 x 6.2 cm. No calcification was identified. Pancreatectomy and splenectomy were done.

The fourth case was a 17 year old female who again came with pain in the abdomen and vomiting for 5 days. Her serum amylase and lipase levels were exceptionally high i.e. 3859 U/L and 31903 U/L respectively. On CT scan a focal heterogenous mass was seen in the tail of pancreas measuring 11 x 8 x 10 cm. A distal pancreatectomy was done in this patient.

The fifth case was of a 55 year old male who presented with pain in the abdomen. His serum amylase levels preoperatively were 54792 U/L while the lipase levels were within normal limits. On ultrasound and CT scan a 5 x 5 x 4.6 cm mass was noted in the tail of pancreas. Distal pancreatectomy was done in this case.

On follow up of one year all the patients were alive with no recurrence or distant metastasis. Chest X ray and liver function tests were within normal limits.

**Histopathological Features**

**Macroscopic Features:** On gross examination, all the five cases were characterised with a well circumscribed mass (mean diameter 9 cm). The cut surface was found to be heterogeneous with multiple cysts measuring 0.5 - 2 cm in diameter, filled with brown fluid along with focal solid, necrotic and haemorrhagic areas. Microscopic features: The histological features of the tumour were similar in all five cases. The tumour was well encapsulated and composed of monomorphic cells forming solid and pseudopapillary structures. The papillae comprised of uniform polyhedral cells arranged around hyalinised fibrovascular stalks with small vessels. The solid areas showed similar cells in diffuse sheets having round-oval nuclei with finely dispersed chromatin. Areas of haemorrhage, cholesterol clefts and cystic change were seen. No mitotic activity, necrosis or capsular/vascular/perineural invasion were identified (Figures 3, 4 and 5). The peripancreatic lymph nodes resected in all the cases showed reactive hyperplasia.
only. In the two cases in which splenectomy was done; the spleen showed congestion only. No tumour infiltration was evident.

**Discussion**

SPT of the pancreas is an extremely rare pancreatic tumour which accounts for 1-2% of exocrine pancreatic tumours. It is usually asymptomatic with non-characteristic abdominal pain.\(^1,4,6\) It is encountered predominantly in young non-caucasian women during first three decades of life although rare cases have been reported in males and in children.\(^2,5,6\)

In our series, one patient was male and he was the oldest. The histogenesis of these tumours remains an enigma but they possibly originate from primordial cells.\(^1,3\) The majority of the tumours are located in the pancreatic body and tail.\(^2,6\) Grossly, the mass is usually large and well encapsulated with varying amounts of necrosis, haemorrhage and cystic change on cut section.\(^6\) Microscopically, both solid and cystic areas are seen. Cells are uniform, polygonal with round to oval grooved nuclei and fine chromatin.

Mitotic activity is not observed. Pseudopapillary structures are formed by the disintegration of tumour cells into pseudocystic cavities. Aggregates of foamy histiocytes, cholesterol clefts can be seen.\(^1-3,6\) Sonographic examination and magnetic resonance imaging also define this as hypervascular, well encapsulated round mass with mixed cystic and solid components.\(^2,3,6\) In the current study, a diagnostic immunohistochemistry (IHC) profile for SPT could not be demonstrated due to non availability of the facility. Case presentations in the literature have observed that histomorphologic features of this tumour are rather characteristic, and differential diagnosis via IHC is not helpful since it shows diversity in most cases.\(^1\) The differential diagnosis of SPT, includes pancreatic endocrine tumour, acinar cell carcinoma, ductal adenocarcinoma, neuroendocrine tumour, liver cyst or tumour, pseudocyst.\(^1,2\) Since, SPT is known for its good prognosis, differential diagnosis from ductal adenocarcinoma is essential. Complete surgical resection is usually curative and favours prolonged survival.\(^2\) The malignant pancreatic tumours are often older at presentation and have a male predilection. Microscopically, ductal and glandular structures form the tumour. They grow in an infiltrative manner.\(^3,4,6\) Acute pancreatitis can be differentiated from SPT by presence of raised serum lipase levels and absence of well defined mass on sonography.\(^7\) The reported rate of recurrence of SPT is 2-6% and for metastasis is 15%. An intensive followup is recommended for such cases.\(^1,2,3\) In our series, all the patients who underwent resection were disease free on a follow up of one year.

In conclusion, SPT is a rare tumour with a low malignant potential. A high index of clinical suspicion is necessary to suspect and diagnose SPT.\(^1,2\) Our results show that diagnosis depends on awareness of
clinical, macroscopic and microscopic features and on sufficient sampling of the tumour tissue.

References


Liraglutide-Induced Acute Pancreatitis

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Abstract

An obese lady of 51 year with Type 2 Diabetes Mellitus for 13 years was prescribed Liraglutide, a glucagon like peptide (GLP-1) analogue (Victoza) for glycaemic control and reduction of weight. She was on gliclazide and Insulin prior to initiation of Liraglutide. Eight weeks after initiation of GLP -1 analogue, she developed severe abdominal pain, nausea and vomiting. She was admitted to a private hospital and evaluated. Biochemical tests and CT scan revealed presence of pancreatitis and she was treated for acute pancreatitis. Liraglutide was withdrawn and symptoms subsided. Subsequent follow-up showed that pancreatic enzyme levels were normal.

Introduction

Liraglutide (Victoza) is a GLP-1 analogue approved by the FDA for the treatment of Type 2 diabetes mellitus (T2DM). It shares 97% structural homology with human GLP-1. However, unlike human GLP-1, which has a half-life of less than 5 minutes, Liraglutide is able to evade degradation by dipeptidyl peptidase IV, which confers on it a half-life of about 13 hours, administered subcutaneously once a day. In clinical studies, treatment with Liraglutide in patients with T2DM reduced haemoglobin A1c by 1% to 1.6%. Liraglutide has also been shown to produce dose-dependent weight loss in obese subjects. Its main side effects are nausea and vomiting. One of the safety concerns is a possible increased risk of pancreatitis. In the phase 2 and phase 3 trials of Liraglutide, there were 7 cases of pancreatitis reported among the 4257 patients treated with Liraglutide and only one case in the 2381 patients in the comparator group. The small number of events makes it difficult to draw conclusions about causation.

This case is being reported as there are only few reports of acute pancreatitis due to Liraglutide in the literature.

Case Report

A 51-year-old obese female of high socioeconomic status reported to our hospital with complaints of uncontrolled and fluctuating blood glucose levels. She had (T2DM) for 13 years, and had obesity with hypertension.