

# Solid pseudopapillary tumour of the pancreas: Report of five cases

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## Abstract

**Abstract:** Solid pseudopapillary tumor (SPT) is a rare, distinct variant of pancreatic neoplasm that has elicited great interest in recent times. The tumor is characterised by the young age of female patients, uniformly good prognosis, and curative surgical excision. SPT is a tumor of low malignant potential. Since it has a characteristic morphology, diagnosis is usually straightforward. The immunohistochemical profile is of interest, with features of both exocrine and endocrine differentiation. From the years 2002 to 2008, we encountered five patients with SPT of pancreas. All were young females with a median age of 24.5 years. All presented with upper gastrointestinal symptoms and mass abdomen. Immunohistochemical analysis was performed on all cases. Four patients are alive and well, and one patient was lost to follow up.

Solid pseudopapillary tumor (SPT) of pancreas is a rare, cystic neoplasm of low malignant potential that occur in adolescent girls and young women. Though a few cases of local recurrence and metastases are documented, the prognosis is good. Complete surgical resection (including metastases) is the treatment of choice. During the period 2002 to 2008, we encountered five patients with SPT of pancreas. Of the five cases, three showed infiltration into either mesenteric or portal vein and one showed infiltration into adjacent normal pancreatic tissue. This report highlights the infiltrative nature of this tumour and the need for a radical treatment for a better patient survival.

## CASE DETAILS

## Figure 1

Table 1: Data of five patients with SPT of pancreas

| Case | Age | Sex | Symptoms                       | Location | Size            | Metastases /<br>Invasion                                       | CA19-9<br>marker | Treatment  | Follow up         |
|------|-----|-----|--------------------------------|----------|-----------------|--|------------------|--|-------------------|
| 1.   | 17y | F   | Abdominal<br>pain, vomiting    | Head     | 13x6x8<br>cms   | SMV†<br>infiltration,<br>no<br>metastases                      | WNL*             | Palliative<br>and<br>followed<br>by<br>CT- & surgery | GJ‡<br>JJ**<br>up |
| 2.   | 22y | F   | Abdomen<br>swelling,<br>nausea | Tail     | 22x21x15<br>cms | None   | WNL              | Excision with<br>splenectomy                         | Uneventful        |
| 3.   | 11y | F   | Nausea,<br>abdominal<br>pain   | Head     | 10x10x6<br>cms  | SMV<br>infiltration,<br>no<br>metastases                       | WNL              | Excision of<br>tumour                                | Uneventful        |
| 4.   | 31y | F   | Abdominal<br>pain              | Neck     | 3.5x3x2<br>cms  | Portal vein<br>confluence<br>infiltration,<br>no<br>metastases | Not<br>Done      | Distal<br>pancreatectomy<br>and<br>splenectomy       | Uneventful        |
| 5.   | 17y | F   | Abdominal<br>pain              | Tail     | 5x4x3.5<br>cms  | Infiltration<br>into tail of<br>pancreas,<br>no<br>metastases  | Not<br>done      | Distal<br>pancreatectomy<br>and<br>splenectomy       | Uneventful        |

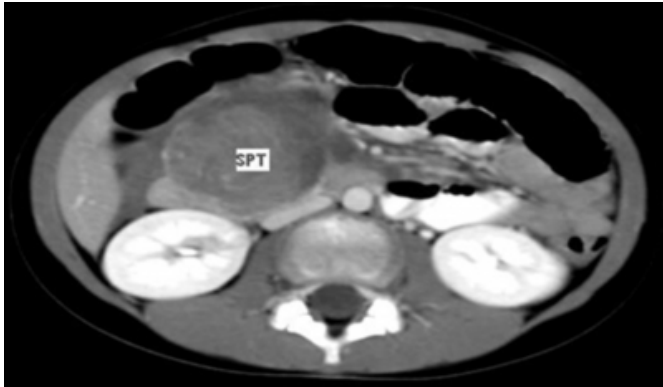
\* Within normal limits, † Superior mesenteric vein, ‡Gastrojejunostomy,

\*\*jejunostomy, ++ Chemotherapy

CT scan of the abdomen was done for all which showed heterogeneously dense mass lesion arising from the pancreas, with solid and cystic component (Fig.1).

**Figure 2**

Figure 1: Contrast CT abdomen showing SPT as a heterogenously dense mass arising from the head of pancreas



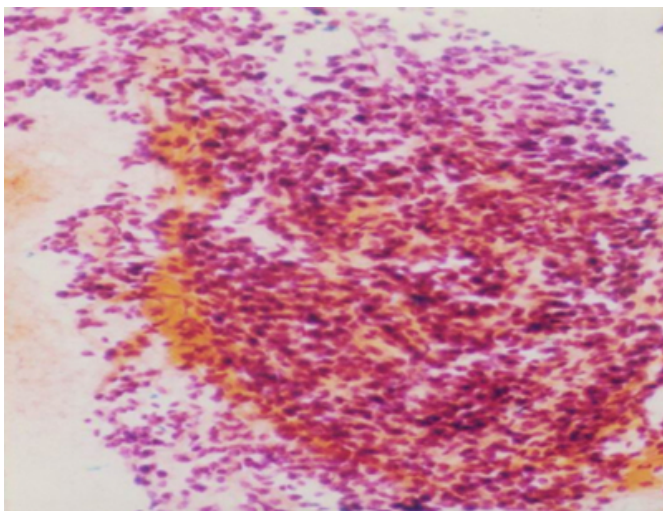
CT guided fine needle aspiration cytology (FNAC) was done for two cases and reported as papillary cystic tumour of pancreas. Post-operative histopathology reports were consistent with SPT of pancreas.

### **PATHOLOGICAL FINDINGS**

**Cytology:** Smears are highly cellular with tumour cells arranged in branching papillae as well as dispersed singly. The cells have scant to moderate amount of cytoplasm and oval, central to eccentrically located nucleus with finely granular chromatin. Few of the cells have longitudinal nuclear grooves (Fig.2).

**Figure 3**

Figure 2: High power (40x) view of hypercellular smears showing papillary as well as singly dispersed tumour cells.



**Gross examination:** Encapsulated and the cut surface reveals lobulated solid and cystic areas filled with necrotic debris. (Fig.3)

**Figure 4**

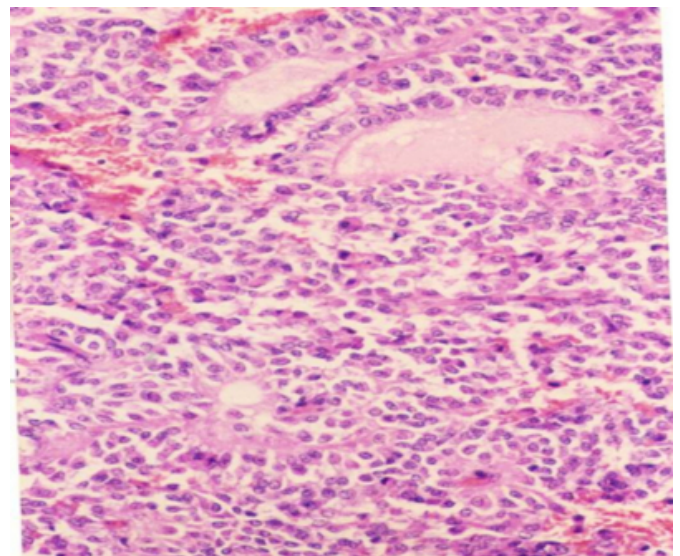
Figure 3: Gross picture of SPT of pancreas showing encapsulation with cystic and solid areas



**Histopathology:** The tumour is composed of cells arranged in the form of solid sheets, microcysts and pseudopapillae (Fig.4). Pseudopapillae are formed by central thin walled blood vessel surrounded by several layers of mildly pleomorphic epithelial cells. The cells have moderate amount of eosinophilic to vacuolated cytoplasm. The nuclei are ovoid and folded with indistinct nucleoli and few mitoses. Areas of haemorrhage and necrosis are seen.

**Figure 5**

Figure 4: Histomorphology (40x) of SPT of pancreas showing pseudopapillary, microcystic and solid areas.



**Figure 6**

Table 2: Immunohistochemistry

| Case | Vimentin          | Cytokeratin      | Progesterone Receptor | Synaptophysin    |
|------|-------------------|------------------|-----------------------|------------------|
| 1.   | Strongly Positive | Focally positive | Focally positive      | Negative         |
| 2.   | Strongly Positive | Focally positive | Positive              | Positive         |
| 3.   | Strongly Positive | Focally positive | Focally Positive      | Focally positive |
| 4.   | Weakly Positive   | Negative         | Focally positive      | Negative         |
| 5.   | Strongly Positive | Negative         | Focally positive      | Positive         |

## DISCUSSION

Solid pseudopapillary tumour of pancreas (SPT) was first described by Frantz in 1959.<sup>1</sup> Approximately 718 cases have been reported in the literature.<sup>2</sup> It accounts for 1-2% of all non-endocrine tumours of the pancreas.<sup>3</sup> Its origin and histogenesis have always been an enigma for pathologists. Characteristic gross and histomorphology is usually sufficient to make a diagnosis of SPT.

Immunohistochemistry is done to exclude other tumours of the pancreas.

SPT has many synonyms like Gruber – Frantz tumour, solid and cystic tumour, solid and papillary neoplasm and papillary epithelial neoplasm.<sup>24</sup> It is often asymptomatic until it is large and generally presents with mild abdominal pain, palpable epigastric mass and upper gastrointestinal symptoms.

Local recurrences and metastases are unusual. The liver is the most common site for metastases and rare cases of lymph node and peritoneal spread have been reported.<sup>5</sup> Rarely, this tumour arises from an extra-pancreatic site such as retroperitoneum, mesocolon or liver.<sup>6</sup>

Prolonged natural history and favourable prognosis in spite of stable metastatic disease, makes it difficult to propose pathological prognostic criteria.

Differential diagnosis on gross examination include congenital pancreatic cyst, pseudocysts, pancreatic hydatid cyst, serous cystadenoma or carcinoma, mucinous neoplasms, cystic islet cell tumour and mucinous duct

ectasia.

Differential diagnosis on histomorphological grounds include islet cell tumour, acinic cell carcinoma, mucinous cystic neoplasms, microcystic adenoma and pancreatoblastoma.

Grossly, it is encapsulated and cut surface reveals lobulated solid and cystic areas filled with necrotic debris.

The light microscopic features are quite characteristic and generally do not present diagnostic problems.

Immunohistochemistry shows a distinctive pattern that corresponds neither to exocrine nor to endocrine pancreatic cell types but may aid in diagnosis in problematic cases. SPT of pancreas is positive for  $\beta_1$ -anti-trypsin,  $\beta_1$ -anti-chymotrypsin, neuron specific enolase, vimentin, synaptophysin and progesterone receptor. It is usually negative for chromogranin, S-100, CEA and estrogen receptor. Staining for pancytokeratin is variable. Ki-67 staining will show a low proliferative index (below 1%).

The origin and histogenesis of this tumor is controversial. Koshmahl et al proposed that even though SPT of pancreas had a complex immune profile, it was found to be related to the genital ridge or ovarian anlage cells which were attached to the pancreatic tissue during early embryogenesis.<sup>7</sup>

In conclusion, SPT of pancreas is a cystic tumour with low malignant potential mostly seen in young women. It rarely metastasizes and has a good prognosis. Complete surgical excision is the choice of treatment. It usually does not present any diagnostic problems on light microscopy making immunohistochemical analysis superfluous in most cases. Its origin and histogenesis are a continuing enigma to pathologists.

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