

# Rare Tumour of Pancreas-Solid Pseudopapillary Tumour: A Case Series.

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## ABSTRACT

Solid pseudopapillary tumour (SPN), also known as Frantz tumour is a peculiar tumour of unknown origin. It accounts for 2.5 % of resected pancreatic neoplasms. It is a low-grade malignant potential tumour. In completely resected tumours, it has good survival rate. We report a case series of 4 cases of solid-pseudopapillary tumour of the pancreas. In all 4 cases, the tumour was completely resected with enucleation done in one patient and histopathology showed solid pseudopapillary tumour. Patients are in follow-up with no metastasis and in good health. Due to the rarity of the tumour and good survival with complete resection, studies are required for its deeper understanding.

**Keywords:** SPN, pancreas, surgical resection.

## INTRODUCTION

Solid pseudopapillary tumour is a rare neoplasm of pancreas. They account for upto 2.5% of resected pancreatic neoplasm.<sup>[1,2]</sup> They were classified as solid-pseudopapillary tumour of the pancreas by World Health Organization (WHO) IN 1996.<sup>[3]</sup> SPN occur in young females of 20-30 years. It is a well capsulated, slow growing tumour but few highly aggressive SPN are reported that exhibit high-grade malignant transformation.<sup>[4]</sup> The survival rate is good with complete resection. The rare occurrence of tumours mandates further studies for its deeper understanding. Hereby we report a series of 4 cases of SPN managed at our institute.

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## CASE SERIES

### Case Report 1:

A 15-year female patient presented with vague abdominal pain with vomiting since 5 days. On examination, tenderness was present in the epigastric region. All routine blood investigations were normal. Ultrasonography of abdomen revealed a 5.5\*5\*6.5 cm well-defined heterogeneous mass arising from the body of pancreas with mild vascularity without calcifications. CT abdomen showed similar findings suggestive of pancreatic neoplasm possibly solid pseudopapillary tumour. The patient was planned for

enucleation of tumour via Whipple's procedure. Intraoperatively [Figure 1], 6\*6\*7 cm hard mass was seen arising from the body of pancreas with extension to neck, free from surrounding structures. The pancreatic mass was enucleated with roux-en-y pancreaticojejunostomy done and specimen was sent for histopathological examination (HPE).

Postoperative course was uneventful and patient was discharged on 14th postoperative day. HPE report [Figure 2] was suggestive of SPN with tumour free margins.

### Case Report 2:

A 25-year female patient came to our institution with complaints of vague intermittent pain in abdomen since 2 years. On examination. The 3\*4cm hard lump was palpable in left hypochondrium. Ultrasonography of abdomen showed 5\*4.5\*5cm well-defined mass arising from pancreatic tail. MRI abdomen showed 4.9\*4.3\*6.2cm well-defined round to oval lesion in pancreatic tail possibly SPN with splenic vein thrombosis with collaterals at splenic hilum and splenorenal region. The patient was planned for distal pancreatectomy with splenectomy. Intraoperatively [Figure 3], 8\*6\*6 cm hard mass arising from pancreatic tail. Distal pancreatectomy with splenectomy was done. Postoperative period was uneventful and patient was discharged on 10th postoperative day. HPE report showed SPN in pancreatic tail with tumour free margins [Figure 4].

### Case Report 3:

A 19-year-old female patient came with chief complaints of vague abdominal pain on and off since 6 months. On examination, a 4\*4 cm lump was

palpable mainly in epigastric region. Ultrasonography of abdomen showed a 5.7\*6.4 cm well defined solid-cystic lesion arising from head and uncinate process of pancreas with ill-defined 2mm wall possibly neoplastic. It maintained fat planes with surrounding structures. CT abdomen [Figure 5] showed similar findings with mass showing close proximity to superior mesenteric vessels. The patient was posted for pancreaticoduodenectomy.

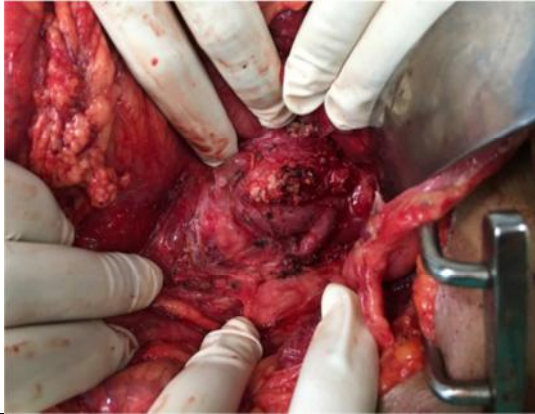


Figure 1: Hard Mass seen intra-operatively.

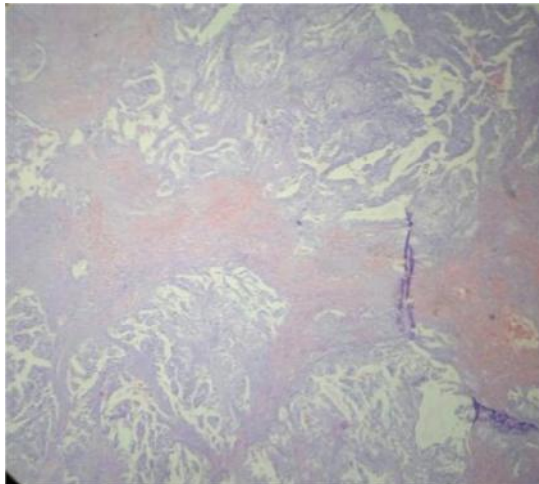


Figure 2: HPE report suggestive of SPN with tumour free margins.



Figure 3: Hard mass arising from pancreatic tail.

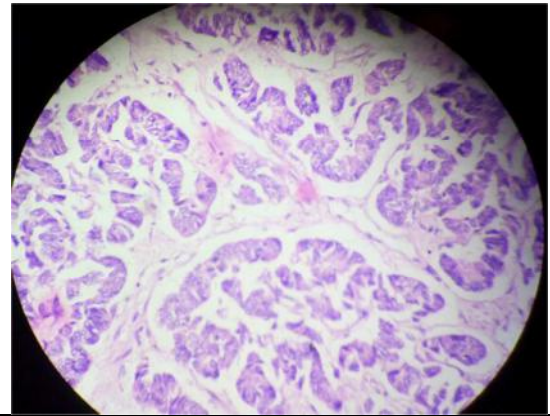


Figure 4: HPE report showed SPN in pancreatic tail with tumour free margins.

Intraoperatively [Figure 6], there was a 6\*6cm mass in head and uncinate process of pancreas abutting the inferior vena cava and superior mesenteric vessels. Whipples procedure was done. HPE which showed features of SPN with margins free of infiltration. The patient was discharged on postoperative day 10.

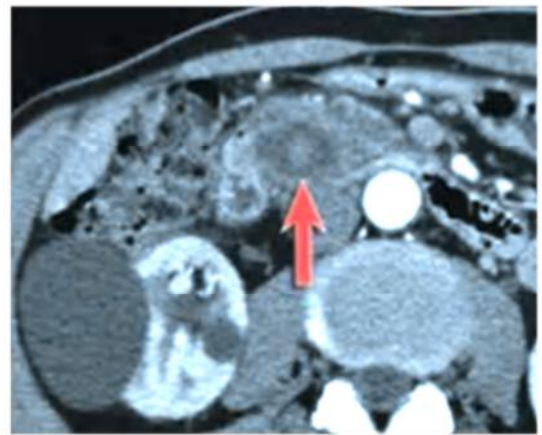


Figure 5: CT abdomen showed similar findings with mass showing close proximity to superior mesenteric vessels.

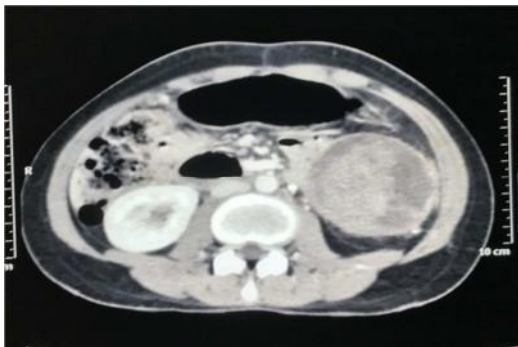


Figure 6: Hard mass in head and uncinate process of pancreas.

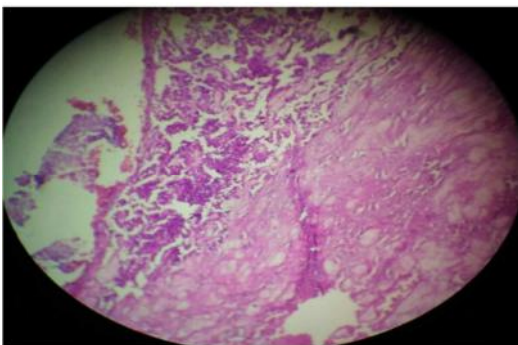


**Case Report 4:**

A 21-year female patient presented with complaints of intermittent abdominal pain since 15 days. Per abdomen examination and all routine examination was within normal limits. CT abdomen [Figure 7] was suggestive of 6.9\*6.1\*7.5 cm well defined peripheral enhancing solid-cystic lesion in pancreatic tail with mild enhancing solid component with 2.6 mm wall thickness with no calcifications, maintaining fat planes with surrounding structures. Splenic vein encircles the lesion superiorly with normal contrast opacification. The patient was posted for laparoscopic distal pancreatectomy with splenectomy. Intraoperatively, 6\*7\*7 cm hard mass was seen in pancreatic tail. On careful dissection, distal pancreatectomy was done, the spleen was saved. HPE [Figure 8] revealed SPN with immunohistochemistry positive for cytokeratin (focal), CD56, Synaptophysin, glypican 3, CD10, Beta-catenin and negative for EMA, Mic2, Chromogranin A. Patient was discharged on postoperative day 6. After 5 days of discharge, patient complained of vomiting which was resolved by oral antacids.



**Figure 7:** CT abdomen suggestive of well defined peripheral enhancing solid-cystic lesion in pancreatic tail.



**Figure 8:** HPE revealed SPN.

## DISCUSSION

Solid pseudopapillary tumour of pancreas is a peculiar tumour of unknown origin. Some favour the theory that it originates from multi-potent primordial cells, while others believe extra-pancreatic origin, from genital ridge angle-related cells.<sup>[5]</sup> SPN is also

called Frantz tumour after the pathologist who first described it in 1959.<sup>[6]</sup> Hamoudi et al<sup>[7]</sup> were first to describe its pathognomonic electron microscopic features.

SPN occurs in young females with male to female ratio of 1:9.8, mean age being 22 years. Its most common location is tail (36%) and head (34%) with metastasis seen in 20% cases.<sup>[8]</sup> SPN is mostly asymptomatic. However, it may present with vomiting and early satiety due to mass effect causing gastric outlet obstruction. 1% cases had jaundice according to Papavramidis series.<sup>[8]</sup> All of our patients had vague, intermittent abdominal pain.

Radiological modalities like ultrasonography, CT scan, MRI are useful in diagnosis of SPN which show well-defined lesion with necrosis, calcifications, haemorrhage, cystic degeneration with peripheral enhancement similar in density to nearby pancreatic tissue.<sup>[9]</sup> SPN differ from adenocarcinomas which are usually hypo attenuated on venous phase CT and from pancreatic neuroendocrine tumours that enhance on arterial phase CT. On HPE, SPN shows combination of solid and cystic components with gradual degenerative changes resulting in pseudopapillary formation.<sup>[4]</sup> On immuno-histochemistry, it is positive for vimentin, keratin, neuron-specific enolase, CD10 and progesterone.<sup>[10]</sup> In contrast to adenocarcinoma, abnormal beta-catenin expression is seen in SPN; while KRAS, TP53, SMAD4 appear unaltered.<sup>[11]</sup> Aggressive phenotypes of SPN are seen and histologically exhibit high mitotic rate, nuclear atypia, spindling of tumour cells and anaplastic giant cells consistent with sarcomatoid carcinoma.<sup>[4]</sup> Preoperative tissue diagnosis is not indicated.<sup>[12]</sup>

Surgical excision with clear margins provides good results in resectable tumours. Many series have demonstrated long-term survival after complete resection.<sup>[10, 13, 14]</sup> Surgery is the treatment of choice even in the cases of distant hepatic metastasis or local recurrence, which are not contraindications for surgical therapy.<sup>[15, 16]</sup> Lymphovascular invasion, local infiltration, nodal and metastasis predicts malignant behaviour with vascular invasion in most common cause of inability to resect the tumours. Recurrence, local invasion and limited metastasis are not necessarily contraindications for resection, and some patients with unresectable SPNs may also have a long survival time.<sup>[17]</sup> SPN are radiosensitive so radiotherapy can be an option for unresectable tumours.<sup>[8]</sup>

In our study, complete surgical resection with tumour free margins was achieved. All patients are in follow-up with no complaints, longest follow-up being for 2 years.

## CONCLUSION

Solid pseudopapillary tumour of pancreas is a rare exocrine tumour of pancreas found in young

females. They have potential for malignant transformation. Complete surgical excision of SPN has a 5-year survival rate of 95%.<sup>[8]</sup> Therefore, SPN should be considered as differential diagnosis of pancreatic mass in young females with vague abdominal pain and surgically treated even in advanced cases.

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