

## Solid Pseudopapillary Tumor of Pancreas in Elderly Female – A Rare Case Report

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**Abstract:** Solid pseudopapillary tumor (SPT) is a rare tumor of pancreas usually presenting in young females. Here we present a case of SPT in an elderly female. A 73 year old female presented with upper abdominal pain. Computed Tomography (CT) of abdomen showed heterogenous tumor arising from tail of pancreas. Distal pancreatectomy with splenectomy was done. Post op HPE was suggestive of acinic cell carcinoma or Neuroendocrine tumor and subsequent IHC study showed an elevated beta catenin level consistent with SPT. SPT is rare, even more rare in an elderly. Complete resection is the treatment of choice.

**Key words:** Case report, Frantz's tumor, Pancreatic tumor, Solid pseudopapillary neoplasm

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### I. Introduction

Solid pseudopapillary tumors also known as Frantz's tumor or Hamoudi tumor was first described by Frantz in 1959 and included in the World Health Organization classification in 1996. SPTs are rare tumors that account for 2.5% of resected pancreatic neoplasms. It typically affects young women with a mean age of 21.97 years. SPT of the pancreas should be considered in the differential diagnosis of any solid and partly cystic pancreatic or upper abdominal mass, particularly in young females. SPT possesses a low malignant potential and a complete surgical resection with clear margins is the treatment of choice. Following R0 resection, SPT has an excellent prognosis.

### II. Case Report

A 73 year old female presented to us with epigastric and left upper quadrant pain for 1 month. She had no history of trauma, weight loss and loss of appetite. She is a diabetic on insulin for 9 years. Abdominal examination revealed a firm mass of size 6x7 cm occupying the left hypochondrium and epigastrium. Complete hemogram, renal function and liver function tests were within normal limits. Abdominal ultrasound showed a well defined heteroechoic lesion of size 6.4x6.1 cm in the tail of pancreas. Contrast enhanced CT of the abdomen revealed a well defined large heterodense lesion of size 6.7x6.6 cm of the tail of pancreas with mild enhancement and thin rim of peripheral and multifocal patchy internal calcification [fig 1]. No liver/peritoneal/nodal metastasis was present. CA 19-9 and CEA levels were within normal limits. Endoscopic USG (EUS) guided FNAC was negative for malignancy. Upper GI endoscopy showed an extraneous impression in the posterior wall of stomach. With all the preoperative investigations, diagnosis of cystic neoplasm of the pancreas with high likelihood of mucinous cystadenoma was made. Intraoperatively 6x6 cm mass was found to be arising from tail of pancreas with involvement of splenic artery and vein and with no liver or peritoneal metastasis. A distal pancreatectomy with 1 cm margin, combined with splenectomy was performed [fig 2,3]. Histopathological examination (HPE) showed pancreatic acini, neoplasm arranged in sheets of nests separated by delicate fibrovascular septa and areas of necrosis with flocks of calcification [fig 4]. Differential diagnosis included acinic cell carcinoma and neuroendocrine tumor. IHC study reported as beta catenin – diffuse cytoplasmic and nuclear positivity, chromogranin – negative and synaptophysin – focally positive in tumor cells favouring SPT. Post operatively patient had an uneventful recovery and discharged on 10<sup>th</sup> postoperative day. Currently, 4 months postoperative, the patient is well without radiological evidence of disease recurrence.



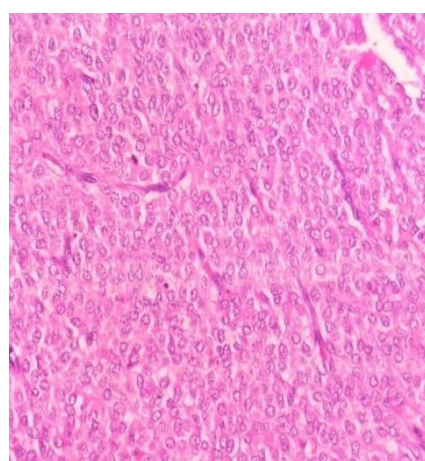
**Fig 1:**CECT abdomen showing heterodense lesion of size 6.7x6.6 cm of the tail of pancreas with mild enhancement and thin rim of peripheral and multifocal patchy internal calcification



**Fig 2:**Post operative specimen of Distal pancreatectomy with splenectomy with the tumor



**Fig 3:**Cut section of the tumor



**Fig 4:**HPE showing pancreatic acini ,neoplasm arranged in sheets of nests separated by delicate fibrovascular septa

### III. Discussion

About 10-15% of cystic lesion of pancreas are neoplastic. Pancreatic cystic neoplasms are a heterogeneous group ranging from benign to malignant. Solid pseudopapillary tumors, also known as Frantz's tumor or Hamoudi tumor accounts for 9.3% of cystic neoplasms and 2% of pancreatic tumors[1]

These tumors are most commonly reported in third decade. Females are 10 times more commonly affected. It is very rare in older age group. In our case, elderly woman was affected. Most common presenting symptom is pain 46.5%, followed by abdominal mass 34%. SPT is asymptomatic in 15% of cases. The most common location of the tumor was the tail of pancreas as in our case.

CT abdomen is the best imaging modality for diagnosing SPT. In CT, SPT appears as large well-encapsulated cystic tumour with variable solid and cystic components. The cystic components are centrally located, while the solid components are more peripheral. The solid components represent tumors which are hypoattenuating on both unenhanced and contrast-enhanced pancreatic and portal venous phases. The wall can be calcified in up to one third of cases. Buetow et al described peripheral calcification as a feature of SPTs. However, other patterns such as coarse central, stippled, and eggshell calcifications have also been reported[2,3]. In our case CT showed a well defined large heterodense lesion in tail of pancreas with mild enhancement with peripheral calcification. The solid components were not well appreciated.

The eggshell calcifications also occur in mucinous cystic neoplasm (MCN) in 30% of cases suggesting malignancy. In cases with predominant cystic component and peripheral eggshell calcification diagnostic dilemma exists between SPT and MCN. Endoscopic ultrasound guided FNA biopsy may be useful when routine imaging is inconclusive and diagnostic uncertainty exists. However, because of the tumor's largely

necrotic composition, FNA biopsy can be nondiagnostic in SPT. In our case also FNA was inconclusive. Characteristic cytologic features of SPTs include, branching papillary fronds with sheets and cords of cells arranged around a fibrovascular septa.

Since the definitive preoperative diagnosis is not arrived even after EUS FNA, we planned for surgical resection keeping MCN as differential diagnosis. Type of surgery depends on location of tumor and adjacent organ involvement. Tumors in head of pancreas are treated by Whipple's procedure, those located in body and tail are treated by distal pancreatectomy alone or along with splenectomy depending on vascular or splenic invasion. In our case tumor is located in body and tail of pancreas with infiltration of splenic vessels, hence we proceeded with distal pancreatectomy with splenectomy[4]

Grossly, these tumors appear tan or yellow and are well circumscribed and smooth. When opened, they show irregular cystic cavities with some areas of necrosis or hemorrhage. Most SPTs can be diagnosed with routine histologic evaluation by the presence of polygonal epithelial cells arranged in a discohesive pattern. In our case there are sheets of neoplastic cells separated by delicate fibrovascular septa raising the possibility of NET and acinic cell carcinoma. IHC study done showed abnormal  $\beta$ -catenin expression which is characteristic of SPT[5]. In our case definitive diagnosis of SPT was made only with IHC.

Given the excellent survival rates following surgical resection alone, adjuvant systemic therapy is not routinely utilized in case of SPT. For unresectable metastatic disease, there are anecdotal case reports of success with regimens including cisplatin, 5-fluorouracil, or gemcitabine. In addition, a single series has suggested that SPT may be radiosensitive, with a report of one patient with an unresectable tumour treated with 40 Gy of external beam radiation therapy who demonstrated tumour response and symptom control at 3 years[6,7]

In general the prognosis is good even in SPT with metastasis or invasion. More than 95% of patients with SPT limited to pancreas are treated by complete surgical excision. Local invasion, recurrence or limited metastasis are not contraindications for resection. Recurrence and metastasis usually occur within 4 years after surgery, thus at least 4-year follow-up is mandatory for all patients undergoing surgical resection. Due to the favorable prognosis and excellent long-term survival even in the presence of metastatic disease, predictive factors of survival are difficult to identify[8]

#### IV. Conclusion

SPT's are rare cystic tumours of pancreas usually occurring in young age. It is very rare in older age group. They usually present as lesion with solid and cystic components with central, stippled or peripheral eggshell calcification which is also seen in MCN. So the high index of suspicion of SPT should be kept in mind in cystic lesions of pancreas with peripheral calcification in older age group, for early diagnosis and appropriate management.

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