

# Central Pancreatectomy for Solid Pseudopapillary Epithelial Neoplasm (SPEN): Reporting a Case

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#### Abstract

Solid pseudopapillary epithelial neoplasm (SPEN) is a rare pancreatic tumor having low but definite malignant potential. We present a case of 43 years old female who has undergone central pancreatectomy for SPEN situated in the body of pancreas. SPEN is having non-specific clinical presentation. Radiological imaging plays a central role in diagnosis. Endoluminal ultrasound guided fine needle aspiration biopsy (EUS-FNAB) is useful in confirming diagnosis in doubtful cases. Surgical resection is the treatment of choice. Central pancreatectomy is feasible for SPEN situated in body of pancreas.

Keywords: Solid Pseudopapillary Epithelial Neoplasm; Central Pancreatectomy

#### Introduction

Solid pseudopapillary epithelial neoplasm is a seldomly found pancreatic tumor with known malignant potential. It predominantly affects females, commonly occurring in second to forth decade of life. This entity was described by Virginia frantz in 1959 and in it was reclassified by WHO in 1996 [1,2]. It comprises very small proportion of all pancreatic tumours. It is also known as Frantz's tumor, solid and papillary tumor, solid-cystic tumor, papillary cystic tumor as well as solid and papillary epithelial neoplasm. It is of low malignant potential; however some cases may be locally aggressive and infiltrative, with metastases to lung, liver and skin [3]. Surgical resection is the treatment of choice and prognosis is excellent. They are more commonly found in tail of pancreas. Here, we report a case of central pancreatectomy for SPEN of body of pancreas.

#### **Case Report**

43 years old female was referred for persistent vague upper abdominal pain. She was having history of diabetes and hypothyroidism for 2 years. Patient has undergone left retroperitoneoscopic donor nephrectomy 8 years back. Patient has undergone excision of pituitary adenoma 2 years back. Her biochemical investigations were within normal range except blood sugar. CA 19-9 level was 13.94 U/ml. Abdominal examination was unremarkable. Ultrasonography of abdomen was suggestive of 18 x 20 mm sized hypoechoic lesion in body of

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pancreas. Her contrast enhanced computed tomography (CT) scan was suggestive of 20 x 24 x 25 mm sized well defined hypo to isodense lesion in proximal part of body of pancreas. The lesion shows patchy enhancement in post contrast study. EUS was suggestive of 12 x 8 x 9.6 mm well defined lesion microcystic areas. FNAB and immunohistochemistry (IHC) of lesion was suggestive of solid pseudopapillary epithelial neoplasm. After optimizing blood sugar levels, patient was subjected to central pancreatectomy. Pancreas transected at the level of neck and distally 1 cm away from lesion. Proximal stump of pancreas was closed with prolene 3/0 in double layer. Drainage of distal pancreatic duct was carried out by end to side pancreato-jejunostomy with Roux En Y loop of jejunum. Pancreatic duct to mucosa anastomosis was carried out with prolene 5/0 interrupted suture and serosa to pancreatic parenchyma anastomosis was carried out by prolene 3/0 interrupted suture. Operative time was 180 minutes. No blood transfusion was required during surgery. Then end to side jejuno-jejunostomy was carried with layers. Her postoperative course was uneventful. Patient was allowed oral liquid from first postoperative day. Antibiotic coverage was given for 3 days. Patient was discharged on fifth postoperative day. She has developed incisional hernia eight months after surgery. She has undergone incisional hernia repair one year after central pancreatectomy. Patient is doing good at 18 months follow up with normal ultrasonographic findings.



Figure 1: Operative specimen.



Figure 2: Solid and cystic areas with pseudorosettes and pseudopapillae.

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Figure 3: Beta-catenin positive.



Figure 4: Ki67.

# Discussion

SPEN is relatively rare tumor, affecting mainly women in second or third decade of life. (Median age of presentation is 26 years) [4,5]. Cases occurring in first decade of life are rare, and less than 10% of SPEN cases have been reported in patients older than 40 years. SPEN in men is a rare occurrence accounting less than 10% of cases. Exact origin of these tumors has not been identified. Probable sites of origin are ductal epithelial cells, neuroendocrine cells, multipotent primordial cells, or even angle of extra genital ridge [6].

Clinical presentations are very nonspecific. Patients with such tumors present with vague abdominal pain or abdominal dullness as observed in our case. With increased availability of CT and MRI, more and more cases are identified incidentally. Tumor markers are not helpful in diagnosis as well as in prognosis of SPEN [7]. SPEN can occur in every part of pancreas but they are slightly more common in tail of pancreas. My patient was having this neoplasm in body of pancreas. Morphologically, it is a sizeable tumor with distinct capsule. Invasion into nearby organ like duodenum, omentum, spleen or adrenal is extremely rare. Radiological imaging of abdomen in the form of abdominal ultrasound, CT scan of abdomen, and MRI usually shows a mass with distinct capsule. Abdominal imaging may demonstrate

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solid as well cystic components within mass. Periphery of lesion may show calcification. Intravenous contrast enhancement, inside the mass, is evident whenever there is hemorrhagic necrosis [8]. MR imaging may be helpful when CT scan cannot differentiate such tumors from other space occupying lesions (SOLs) of pancreas. Still, confirmatory preoperative diagnosis is difficult to arrive in all patients because of the similarity of findings among SOLs of pancreas. EUS guided FNAB may helpful in arriving definitive diagnosis. As in my case, patient was subjected to EUS FNAB and confirmatory diagnosis of was established by immunohistochemical staining of biopsy specimen. However, role of EUS-FNAB is controversial as it is associated with a risk of tumor cells implantation along the tract of biopsy thereby allowing tumor spread [9]. In addition, it is associated with complications like hemorrhage, acute pancreatitis, and duodenal perforation. However, risk of complications is less than 1% with EUS-FNAB [10-16]. So, it should be utilized for definitive preoperative diagnosis of SPEN when abdominal imaging is not sufficient to this tumor from other SOLs of pancreas.

Since SPEN is associated with very minimal risk of local invasion, amount of pancreatic parenchyma to be resected can be minimized. Tumours situated in body of pancreas can be managed by central pancreatectomy which will preserve the function of distal pancreas. Another advantage with CP is preservation of spleen, Utilisation of central pancreatectomy is limited by risk of development of pancreatic fistula from anastomosis site of pancreato-jejunostomy. However, with meticulous surgical technique such risk can be reduced. Surgical loop magnification should be useful in carrying out duct to mucosa anastomosis. CP is can be performed safely with risk acceptable morbidity and no normality.

## Conclusion

Central pancreatectomy is a feasible and relatively safe for SPEN situated in body of pancreas.

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