

Case Report

Solid pseudopapillary epithelial neoplasm of pancreas presenting as an abdominal lump: a case report

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ABSTRACT

Solid pseudopapillary epithelial neoplasm (SPEN) of the pancreas is a rare cystic exocrine tumour of the pancreas that predominantly affects women between 30 and 40 years of age. This case report aims to describe clinical presentation and management of SPEN. We report a case of 20 years old female who presented with palpable lump in upper abdomen and continuous dull aching pain. Preoperative contrast enhanced computed tomography (CECT) abdomen was suggestive of large solid mass of size 11×10×10 cm with liquified content in relation to tail of pancreas with a possibility of retroperitoneal tumour or gastrointestinal tumour (GIST). Spleen sparing distal pancreatectomy with segmental resection of transverse colon with colo-colic anastomosis was performed and the diagnosis of SPEN was subsequently confirmed on histopathology examination. SPEN is a slow-growing tumour with a low-grade malignant potential found incidentally in asymptomatic patients. The average tumour size is about 4 to 6 cm in diameter at presentation. Distal pancreatectomy with splenectomy is the procedure of choice as described in literature. SPEN should be kept as a differential diagnosis in a young female patient presenting with vague abdominal pain and retroperitoneal lump in relation to the pancreas. Complete surgical resection is the definite treatment with a 5-year survival rate of 95%.

Keywords: Solid pseudopapillary epithelial neoplasm, Distal pancreatectomy, Transverse colon, Case report

INTRODUCTION

Solid pseudopapillary epithelial neoplasm (SPEN) of the pancreas is a rare type of cystic exocrine pancreatic tumour that predominantly affects women between 30 and 40 years of age.¹ SPEN of pancreas was first described by V.K. Frantz in 1959.^{1,2} It is also known as a solid pseudopapillary tumour, papillary epithelial neoplasm, papillary cystic neoplasm, solid and papillary neoplasm, low-grade papillary neoplasm, and Frantz tumour.³ Women to men ratio is 10:1.⁴ Although rare, SPEN in men is more aggressive with local invasion.

SPEN has low-grade malignant potential with metastasis present in only 5% of cases. Most of the patients are asymptomatic at diagnosis and are incidentally detected while undergoing abdominal imaging for other complaints. Symptomatic patients present with dull aching

continuous abdominal pain or abdominal discomfort, awareness of abdominal lump (as in our case), nausea, vomiting, loss of appetite, early satiety, or weight loss.⁵

On abdominal examination there can be a palpable lump in epigastrium and left hypochondrium. Surgical resection is the definitive treatment and it usually has a good prognosis. Poor prognostic factors are male patients, tumour size greater than 5 cm, local invasion into adjacent structures, vascular invasion and necrosis or cellular atypia on histopathology. Neuroendocrine pancreatic tumour has similar clinical presentation and it should be considered as a differential diagnosis of SPEN.⁶

We herein report a case of 20 years old female who presented with lump in left hypochondrium and epigastrium with a contrast enhanced computed tomography (CECT) diagnosis of retroperitoneal tumour

or gastrointestinal tumour (GIST). Intra operatively spleen sparing distal pancreatectomy with segmental resection of transverse colon with colo-colic anastomosis was done and final diagnosis of SPEN was made on immunohistochemistry (IHC) report. The case report has been reported in line with the SCARE 2020 criteria.⁷

CASE REPORT

Patient information

A 20 years old female patient presented in outpatient department of surgery with history of awareness of lump in upper abdomen for last 1 year. Lump was gradually progressive in size over a period and became more prominent in last 3 months. It was associated with continuous dull aching pain in epigastric region without any aggravating or relieving factors. There was no history of fever, nausea, vomiting, abdominal distension, dyspepsia, and jaundice. Patient did not undergo any abdominal surgery previously and family history was insignificant.

Clinical findings

On abdominal examination there was visible fullness with a single palpable lump of size about 10×8 cm occupying epigastric and left hypochondrium region. It was firm in consistency with ill-defined margins, had irregular surface, was non-tender and did not move with respiration. Bowel sounds were normal on auscultation.

Lab investigations

Patient was anaemic with haemoglobin level of 9.8 gm/dl. Serum amylase and lipase levels were 55 and 197. Liver function test, renal function test and coagulation profile were within normal limits.

Diagnostic investigations

Ultrasound abdomen revealed a large well defined complex mass measuring 10.2×9.6×8.1 cm abutting the tail region of pancreas. The mass consisted of predominantly hyperechoic areas with few cystic components. Mild internal vascularity was also seen within the solid components. CECT abdomen demonstrated a large solid appearing mass lesion of size 11×10×10 cm with peripherally enhancing solid component which enhanced up to 56 HU and with liquified component. Mass was present in relation to tail of pancreas without any locoregional invasive changes. There was no daughter cyst configuration, calcifications, or fatty component (Figure 1).

Fat planes with the small and large bowel loops, liver, stomach, left adrenal and left kidney were distinctly seen. Possibility of a retroperitoneal tumour or GIST was kept. CECT thorax was done which was normal.

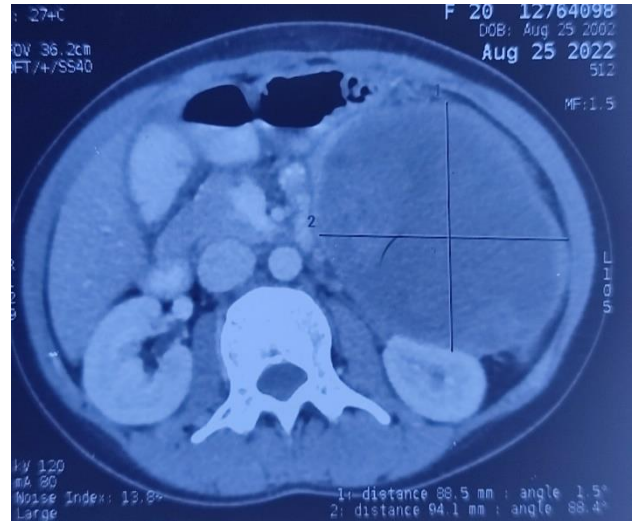


Figure 1: CECT abdomen photograph showing large solid mass in relation to the tail of pancreas.

Treatment

Patient was taken up for exploratory laparotomy. Intraoperatively on opening the lesser sac a solid mass of size 10×10 cm was present arising from the tail of pancreas (Figures 2 and 3). Mass was densely adherent to transverse colon with its mesentery was draped over the mass. It was not possible to ascertain whether the mass had infiltrated the colon or its mesentery (Figure 4). Spleen sparing distal pancreatectomy with segmental resection of transverse colon with colo-colic anastomosis was performed without compromising the resection margins (Figure 5).

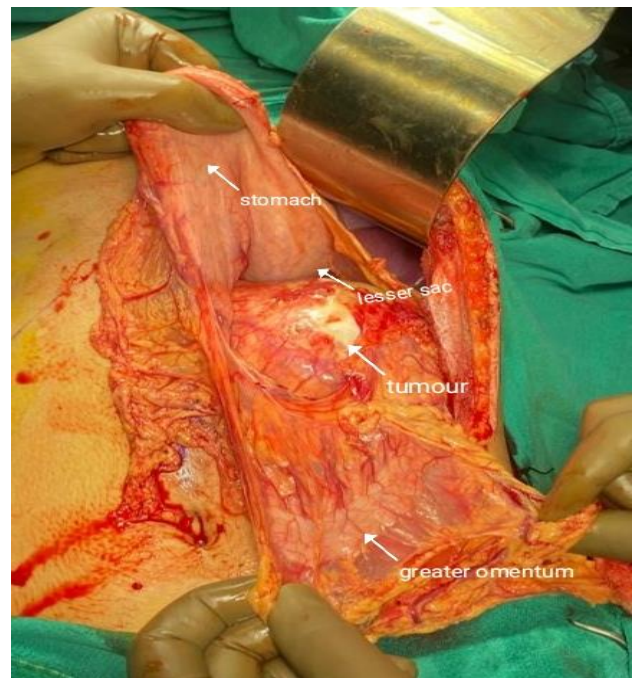


Figure 2: Intraoperative photograph showing location of tumour on opening the lesser sac.

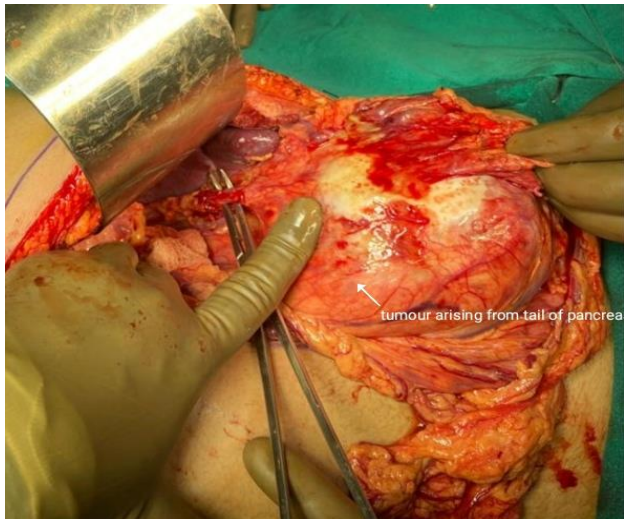


Figure 3: Intraoperative photograph showing mass arising from pancreas with feeding vessel.

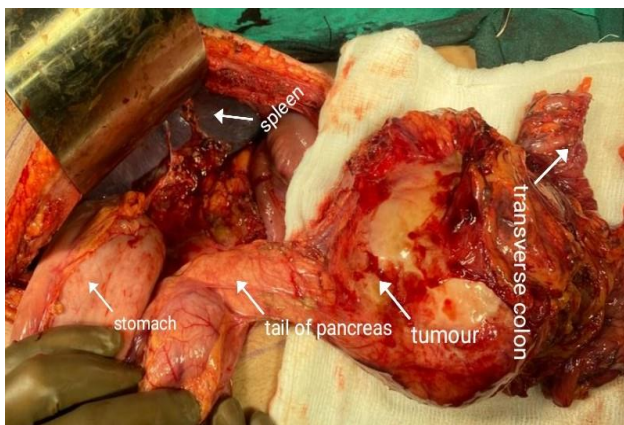


Figure 4: Intraoperative photograph showing large tumour arising from tail of pancreas with resected segment of transverse colon.



Figure 5: Photograph after resection showing tumour and resected segment of transverse colon.

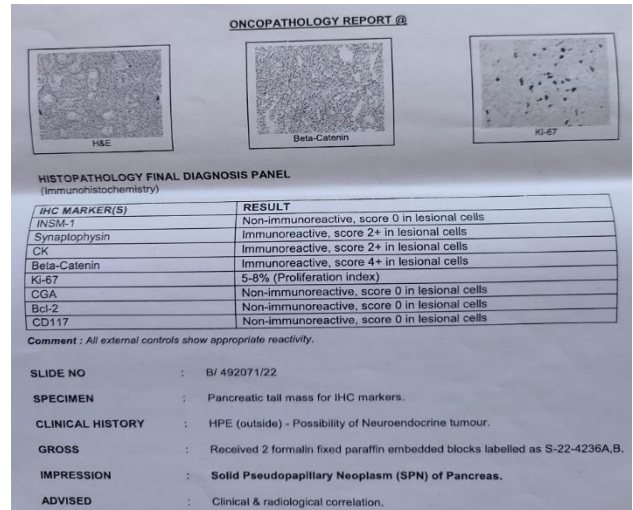


Figure 6: Photograph showing IHC report of the patient with impression of SPEN of pancreas.

Outcome and follow up

In post-operative period, drain and serum amylase levels were normal on POD-1 and POD-3. Drain was removed on 3rd day and patient was discharged on 6th POD in stable condition. Histopathological examination shows the possibility of neuroendocrine tumour with tumour free margins from the adjacent intestine. Final diagnosis of solid pseudo papillary epithelial neoplasm (SPEN) was confirmed in IHC report (Figure 6). Patient had been on a regular follow up for 6 months and no residual or recurrent mass has been detected.

DISCUSSION

SPEN was initially identified by Frantz in 1959 and reclassified by the World Health Organization in 1996.⁸ It is also known as a solid pseudopapillary tumour, papillary epithelial neoplasm, papillary cystic neoplasm, solid and papillary neoplasm, low-grade papillary neoplasm, and Frantz tumour. The tumour's cell of origin is still uncertain. It is a rare tumour usually affecting the women aged between 30 and 40 years. Although it may occur anywhere within the pancreas but the most common site of origin is pancreatic tail.

SPEN is a slow-growing tumour and have become symptomatic when the tumour becomes large enough to produce mass effect.⁹ Most of the patients are asymptomatic at diagnosis and are detected incidentally on imaging for other complaints. Symptomatic patients present with vague abdominal pain, abdominal discomfort, abdominal lump, loss of appetite, nausea, vomiting, weight loss.

No tumour marker is sensitive or specific enough to make a diagnosis with several studies reported normal levels for serum amylase and tumour markers such as CA 19-9, CEA, and CA125 in most patients.¹⁰ Liu et al reported a

biomarker (CA 72-4), which was elevated in only 8.6% of patients in their study.¹¹ Typically, SPEN are encapsulated, mixed solid and cystic tumours with haemorrhage.¹² CT shows a well-demarcated encapsulated pancreatic mass with variability ranging from predominantly solid, homogenous muscle density, mixed solid and cystic to thick-walled cyst with haemorrhage and necrosis.¹³ Calcifications and enhancing solid areas may be present at the periphery of the mass. Li et al. classified tumour into five types from purely solid to purely cystic on CT images based on solid-cystic ratio. Since the tumour is so rare so the preoperative diagnosis of SPEN is often missed as in our case.

The choice of surgical resection depends upon tumour's location, invasiveness, and presence of metastasis. The procedure for this tumour in the literature is a distal pancreatectomy with splenectomy however, we performed a spleen sparing distal pancreatectomy with segmental resection of transverse colon with colo-colic anastomosis for our patient. Following surgical resection, patients generally have an excellent prognosis, and several studies have reported a disease-free survival rate of >95%.¹⁴

We have followed-up the patient for 6 months without any evidence of recurrence. Long term follow-up is required as some patients may develop recurrence even after 7 years.¹⁵ Five-year survival rate is 95% if complete resection is done without compromising the resection margins. The indicators of poor prognosis are factors such as male patients, tumour size more than 5 cm, vascular or local invasion into adjacent structures, necrosis, or cellular atypia on histopathology, metastasis and unresectable tumours.

CONCLUSION

SPEN is rare neoplasms of pancreas with malignant potential. It should be kept as a differential diagnosis in a young female patient presenting with vague abdominal pain and palpable lump in relation to the pancreas. Complete surgical resection is the definite treatment with a 5-year survival rate of 95%. histopathological examination confirms the final diagnosis of SPEN. Long term follow-up is required to identify patients with recurrence however further studies are warranted to understand the pathogenesis of the disease, identify potential biomarkers and risk factors that would identify patients with high risk of recurrence.

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