

## Case Report

# Solid pseudopapillary epithelial neoplasia of the pancreas causing sinistral portal hypertension: a case report

Akshat Mishra\*, Girish D. Bakhshi, Rajesh Yadav, Samprati Doddamalappa, Mahesh Chanap, Sachin Sholapur, Sumit Malgaonkar, Kunal Suradkar

Department of General Surgery, Grant Govt. Medical College and Sir J.J. Group of Hospitals, Mumbai, Maharashtra, India

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**\*Correspondence:**

Dr. Akshat Mishra,

E-mail: [drakimish@gmail.com](mailto:drakimish@gmail.com)

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### ABSTRACT

Solid pseudopapillary epithelial neoplasm (SPEN), also referred as Franz's tumour or Hamoudi's tumour is a low malignant potential epithelial neoplasm of the pancreas. It occurs at a much lower frequency than other cystic neoplasms of the pancreas. It occurs rarely and in young females. Although infrequently, large sized tumours are known to compress the splenic vein thereby resulting in extra hepatic portal venous obstruction (EHPVO) resulting in left sided portal hypertension (PHT). The resulting periportal collateral circulation poses an intraoperative challenge while approaching these tumors. Meticulous pre-operative planning can go a long way in managing such a case and providing complete surgical cure. We present a case of SPEN in a 37 years old female which was discovered incidentally and managed surgically to achieve a complete cure to both the tumour and the left sided PHT.

**Keywords:** SPEN, Left sided portal hypertension

### INTRODUCTION

Solid pseudopapillary neoplasm/tumour (SPEN) was first described by Franz in 1959, hence, it was previously referred to as Franz's tumour or Hamoudi's tumour.<sup>1</sup> In 1996, World Health Organisation (WHO) labelled it as SPEN.<sup>2</sup> SPEN is a rare neoplastic condition of the exocrine pancreas with a reported incidence ranging from 0.17% to 16% of all exocrine pancreatic neoplasms.<sup>3</sup> 90% of all tumours are reported in young females, usually in the 3<sup>rd</sup> decade of life with a median age of presentation ranging from 22 years to 28 years.<sup>4</sup> The usual presentation is nonspecific and most patients are asymptomatic at the time of diagnosis. The tumour is usually incidentally discovered on imaging done for other reasons. Rarely, it may show features of left sided portal hypertension. Ultrasonography (USG), contrast enhanced computed tomography (CECT) and magnetic resonance cholangio-pancreatography (MRCP) are essential for diagnosis and for its anatomical

considerations which can assist in further management. Endoscopic ultrasound (EUS) guided fine needle aspiration (FNA) is also an essential tool for diagnosis and can provide material for preoperative cytological diagnosis. Despite the usual size of the tumour at presentation and the possibility of both local and distant invasion, SPEN is regarded as a tumour with low malignant potential.<sup>5</sup> Rarely, the tumour may cause external compression of the splenic vein leading to left sided portal venous hypertension and subsequent development of a vivid collateral circulation. Currently the treatment of choice for SPEN is surgical resection with surgery providing 5-year survival rates of almost 95% of all cases.<sup>4</sup>

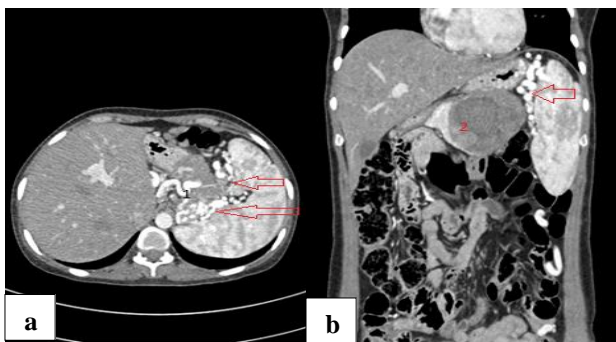
Depending upon the site of the tumour, distal pancreatectomy with splenectomy, central pancreatectomy or even pancreaticoduodenectomy are the various surgeries offered to the patients.<sup>6</sup>

## CASE REPORT

A 37-year-old female presented with complaints of vague abdominal discomfort localised primarily to the upper abdomen for 10 days. She was admitted at a secondary health care centre in her hometown for complaints of dizziness and generalised weakness where she was diagnosed as a case of iron deficiency anaemia and received blood transfusions for the same. During her work up for the same, a USG abdomen revealed a space occupying lesion in the tail of pancreas. She was then referred to our centre for further management. She had a history of a hypothyroidism for 1 year for which she was on 75 mcg thyroxine once daily. Clinical examination was unremarkable.

Her haemoglobin (Hb) on admission was 13.8 gm%, total bilirubin was 0.8 mg%, serum amylase was 19 U/l, serum lipase was 18.4 U/l and alkaline phosphatase was 62.2 U/l signifying no biochemical abnormality of the pancreato-biliary system. Her glycosylated haemoglobin (HbA1C) was 5.6% ruling out any pancreatic endocrine insufficiency.

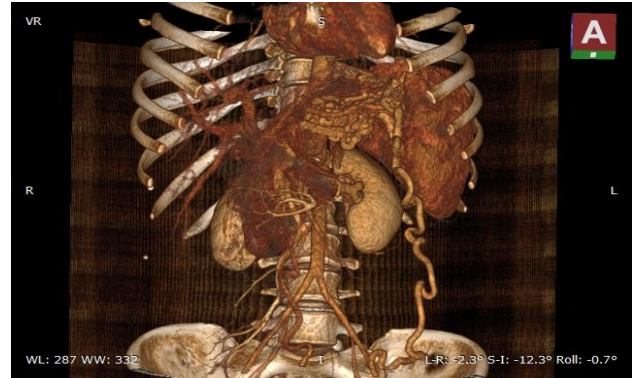
USG abdomen showed mild splenomegaly with a heterogenous, relatively well-defined solid mass with central necrotic areas is seen arising from anterior aspect of distal portion of body of pancreas measuring 6.1×4.5×5.8 cm (APXTRXCC) showing mild vascularity in the peripheral region with few tiny calcific foci. CECT abdomen revealed a well encapsulated lesion with varying solid and cystic components measuring 5.4×7.9×6.6 cm (APXTRXCC) arising from the body and tail of pancreas showing post contrast enhancement with solid components peripherally and central cystic areas and few calcifications. It was compressing the splenic vein resulting in multiple portal collaterals in the periportal, perisplenic, peripancreatic and perigastric region (Figure 1a and b).



**Figure 1: (a) Axial CECT showing splenic vein (1) and periportal and perigastric collaterals (arrow) and (b) coronal section showing SPEN (2) and perisplenic collaterals (arrow).**

Preoperative planning using a 3D reconstruction of CECT was done (Figure 2) to identify the vascular anatomy of portal collaterals and distal pancreatectomy with splenectomy was planned. It was decided to proceed with

an open surgical approach. During surgery, the collaterals were meticulously ligated in the gastrosplenic ligament to reach the splenic artery which was ligated next. Splenic vein was ligated 5mm away from its junction with superior mesenteric vein to achieve tumour free margin of pancreas. Pancreas was then divided after applying a vascular clamp.



**Figure 2: 3D reconstruction showing vivid portal collaterals.**

The cut edge of pancreas was underrun with absorbable sutures. The specimen was then delivered and sent for histopathology (Figure 3).



**Figure 3: Final specimen of distal pancreatectomy and splenectomy showing the tumour, specimen was sent for histopathology.**

The post-operative course was uneventful. Final histopathological report for the specimen was solid pseudopapillary neoplasm with no lymphatic or vascular emboli and no evidence of perineural invasion with no involvement by tumour. Distal margin was free of tumour while no regional lymph nodes were involved. Final AJCC TNM staging was p T3 p N0. Follow up of 2 months has shown her to be disease and symptom free.

## DISCUSSION

SPEN is a rare neoplasm of the exocrine pancreas with low malignant potential. Although varying in sizes, there seems to exist a cystic component secondary to degeneration in larger sized tumours.<sup>7</sup> Majority of this

tumour seems to occur in younger females with a median age of presentation of 28 years (range 7-79 years). SPEN are usually large tumours (>60 mm), predominantly solitary, well demarcated and may occur anywhere in the pancreas though there seems to exist a predilection for the pancreatic tail.<sup>8</sup> Present case was a 37 years old female with tumour occupying the body and tail of pancreas.

Majority of patients are asymptomatic and tumour is discovered incidentally during radiographic evaluation for some other condition. Sometimes there may be vague abdominal discomfort with or without a palpable non tender mass in the epigastrium. Large size tumours may present with mass effects symptoms, viz. early satiety, pain, nausea and or vomiting. Rarely the tumour may present with jaundice or hemoperitoneum secondary to rupture of tumour capsule.<sup>4,6</sup> In present case, patient was asymptomatic and tumour was found as incidental finding on imaging.

Radiologically, plain radiographs are valuable only if the tumour shows calcifications (peripheral calcifications may be seen in as much as 31% of all tumours), while on USG the lesion appears as a heterogenous solid, often cystic mass.<sup>9</sup> On CT and MRI, the tumour appears as well circumscribed, encapsulated and a heterogenous mass with solid and cystic components.<sup>9</sup> On MRI, areas of T2 hyperintensity correlate with areas of cystic degeneration while that of T1 hyperintensity correlate with that of haemorrhagic degeneration with the soft tissue component showing contrast enhancement.<sup>10,11</sup> In present case CECT helped not only in diagnosing tumour but also sinistral PHT which was a result of compression of the splenic vein by tumour. Left sided PHT is rarely associated with SPEN. Only 4 cases have been described in English literature.<sup>12-15</sup> Though splenic vein thrombosis has been reported with pancreatic neuroendocrine tumours<sup>16</sup>, a cases series showing SPEN causing left sided PHT has not been documented. Present case demonstrates use of 3D reconstruction of CECT to map the vascular anatomy of portal collaterals before surgery which helps in planning surgery and intraoperative steps.

At the time of surgery, conservative resection at the time of presentation with preservation of as much pancreatic tissue as possible is the treatment of choice. Extensive lymphatic dissection is not advisable when the disease is well localised while metastasis is not a contraindication to surgery Present case was managed with resection of pancreatic body and tail with splenectomy.

Grossly SPEN appear as large well circumscribed and encapsulated lesions that are well demarcated from rest of the pancreas and rarely show infiltration of the spleen or the duodenal wall.<sup>17</sup> In approximately 85% cases SPEN is limited to the pancreas whereas metastasis to the liver, regional lymph nodes, omentum, mesentery and peritoneum may be seen in 10-15% patients at the time of presentation.<sup>18</sup>

Angioinvasion, perineural invasion or deep invasion of the pancreatic parenchyma is defined as the criteria for differentiation of malignant SPEN despite its low malignant potential. Nuclear atypia, high mitotic index, Ki-67 on immunohistochemistry, extensive necrosis and sarcomatoid areas are other indicators associated with aggressive behaviour.<sup>19</sup> However, present case showed no such features of histopathology. Due to complete resectability, adjuvant therapy is rarely indicated in cases of SPEN, hence in present case no adjuvant therapy was given.

Despite the size of malignancy at presentation, SPEN is associated with a fairly high survival rate of 95% at 5 years of follow up after complete surgical resection. Even patients with metastatic disease at presentation have a high survival rate and in present case a favourable outcome is expected upon long term follow up.

## CONCLUSION

SPEN is a rare low-grade malignancy of the pancreas in young females which is usually detected incidentally on radiological studies. SPEN can exist as a cause of sinistral portal hypertension secondary to splenic vein compression due to mass effect. This usually results in the development of large portal collaterals. Preoperative 3D- reconstruction of CECT helps in vascular mapping and intraoperative approach to prevent bleeding and minimise blood loss. Though definite case series do not exist regarding SPEN causing sinistral PHT, but the varied case reports indicate the need for further research in this direction.

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