

Solid pseudopapillary neoplasm of the pancreas

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Abstract

Solid Pseudopapillary Neoplasm (SPN) of pancreas is atypical and uncommon pancreatic neoplasm which accounts for less than one percent of all pancreatic tumours. It is more common in females but has also been described in males. It has distinct radiological and histological features. Surgical resection is the mainstay of treatment.

We report two cases of pancreatic neoplasm which were revealed as pseudopapillary tumour on histopathology report and both of the patients are doing well in follow up.

Key words: Pancreatic Neoplasm, Pseudopapillary Tumour

INTRODUCTION

Solid Pseudopapillary Neoplasm (SPN), first described by Frantz in 1959¹, is a rare pancreatic neoplasm accounting for only one percent of pancreatic neoplasm². The rarity of SPN further contributes to lack of consensus in both diagnosis and treatment^{2, 3}. SPN typically occur in young women of age 20 to 30 years and are almost always benign in character. However, up to 20% may demonstrate a malignant character^{2, 4, 5}. Nearly 100% five year survival is achieved on patients with benign SPN, who lack lymph node invasion or distant metastasis. However, overall five year survival is estimated to be 95% including patients with malignant disease^{2, 4, 6}. The few patients that do not survive succumb to metastatic disease².

Here we present two cases of SPN, one in female and other in male and tumour located in head and tail of pancreas respectively.

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CASE REPORT

Case 1

A 44 years old female presented at Kathmandu Medical College Teaching hospital (KMCTH), Sinamangal with complains of intermittent upper abdominal pain for one month which was insidious in onset, dull aching and non-radiating, associated with intermittent non-projectile, bilious vomiting. Her pulse rate was 70/min and her blood pressure was 120/80 mmHg. There was no pallor and jaundice. The abdomen was soft but a firm; non tender immobile mass of 10 cm diameter was palpable in epigastrium. Laboratory data including haematology, blood chemistry, including amylase and lipase were within normal limits. Ultrasound abdomen showed a pancreatic tumour with solid and cystic components. Computed Tomography (CT) scan demonstrated a large predominantly cystic mass having irregular wall and containing nodular excrescence, with variable thickness septa arising from head of the pancreas causing splaying of pancreatic head parenchyma and extending anteriorly as an exophytic mass (Figure 1). Superior mesenteric vein (SMV) and SMV/splenic vein confluence were broadly abutted by the mass with flattening, probably due to infiltration.

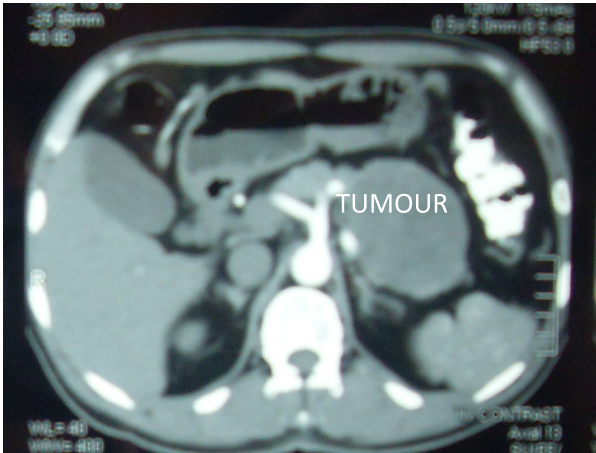


Figure 1: CT scan image of abdomen showing pancreatic head mass

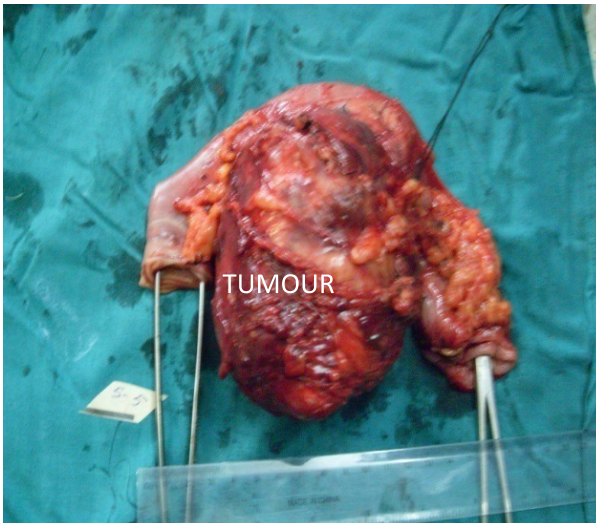


Figure 2: Specimen after pancreaticoduodenectomy

Pancreaticoduodenectomy was done on 2009 August 23. The resected tumour was 12 cm × 10 cm in size, firm in consistency, surrounded by pseudocapsule (Figure 2). Cut section showed cystic areas, with gelatinous substance, necrosis and haemorrhagic area. Histopathological examination was suggestive of solid pseudopapillary tumour of pancreas. Post operative period was uneventful and she was discharged on eighth post operative day. She is doing well in follow up of two years.

Case 2

A 43 years old gentleman presented to KMCTH, Sinamangal with complains of intermittent upper abdominal pain for two month which was insidious in

onset, dull aching. His pulse rate was 80/min and blood pressure was 110/70 mmHg. There was no pallor and jaundice. The abdomen was soft, no mass was palpable. Laboratory data including haematology, blood chemistry, including amylase and lipase were within normal limits. He had history of hospital admission for diabetic ketoacidosis two months back.

Ultrasound abdomen and CT scan revealed a mixed echogenic mass at the tail of pancreas of around 10 cm × 8 cm suggestive of a neuroendocrine tumour (Figure 3).

Distal Pancreatectomy with splenectomy was done on 2010 August 19. The resected tumour was 10 cm × 8 cm in size, soft in consistency, surrounded by pseudocapsule (Figure 4). Cut section showed cystic areas, with gelatinous substance, necrosis and haemorrhagic area. Histopathological examination was suggestive of solid pseudopapillary tumour of borderline malignancy of

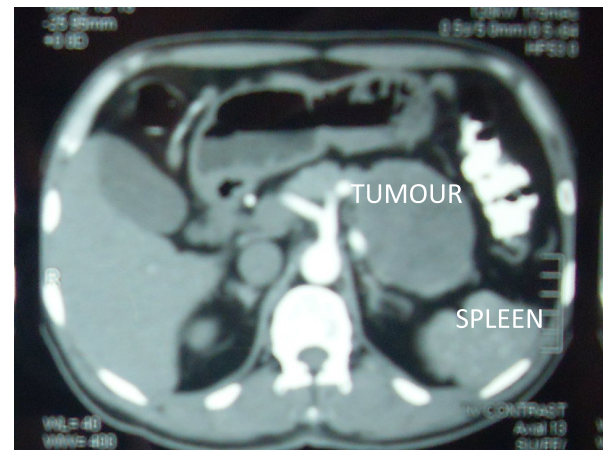


Figure 3: CT scan image of abdomen showing pancreatic tail mass (Case 2).

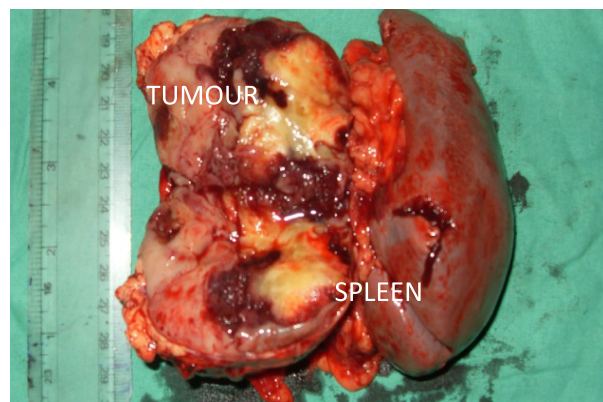


Figure 4: Specimen after Distal Pancreatectomy with splenectomy (Case 2).

pancreas. Post operative period was uneventful and he was discharged on 10th postoperative day. He is doing well in follow up of 18 months.

DISCUSSION

Solid Pseudopapillary Neoplasm is uncommon pancreatic neoplasm accounting for about one percent of pancreatic neoplasm^{2, 7}. SPN predominantly involve young female of age 20 to 30 years suggesting that hormonal factors contribute to the tumour growth⁹⁻¹⁰ but has also been described in males and children^{2, 4, 11}.

SPN may present as non specific abdominal complains, abdominal mass or fullness and incidental radiological findings¹². While asymptomatic presentation was prevalent in past, with wide spread use of ultrasonogram and other imaging modalities, incidental finding is becoming more common now¹³⁻¹⁵. Pain and abdominal mass or fullness is reported as most common symptoms¹³. A study reported abdominal pain as frequent as 67%, back pain 10%, weight loss 10%, nausea and vomiting 10% and diarrhoea five percent¹⁶. Less frequent symptoms include jaundice, gastrointestinal obstruction, anaemia and pancreatitis¹⁷.

A cross-sectional abdominal imaging is most important investigation which shows distinct characteristic large encapsulated mass with solid and cystic components, and intra-tumoural haemorrhage^{15, 18}. However laboratory studies including tumour markers are unhelpful¹⁵.

The pathological characteristic of SPN is also distinct which include larger size on presentation. However, despite large size, resection margins are negative with minimal rate of lymph node involvement and low malignant potential^{2, 4, 19}. Gross examination reveals a typical gelatinous structure mixed with solid component with variable amounts of necrosis, haemorrhage, and cystic changes⁷. However SPN in men tend to be solid¹¹. Microscopically, cystic parts are surrounded by solid tumor tissue with characteristic pseudopapillary structures which penetrate as finger like projection into these cystic areas with areas of haemorrhage and necrosis^{9, 20}. All these SPN demonstrate nuclear and cytoplasmic accumulation of Beta-catenin

protein in immunohistochemistry and 86% of them had activating mutation of beta-catenin gene¹¹. No pancreatic neuroendocrine tumours showed such immunohistochemical findings and genetic alteration. Hence, nuclear accumulation of beta-catenin appears to be useful markers of SPN¹¹.

The mainstay of treatment of SPN is surgical resection. Extent and type of surgery depends on the location of the tumour. Tumour located in head of pancreas needs pancreaticoduodenectomy, that in neck needs central resection and tumour in body and tail are dealt with distal resection^{12, 21}. Aggressive surgical approach towards local and distal metastasis is justified due to excellent long term prognosis even in presence of metastatic disease¹². Several studies have reported successful en-block resection of locally advanced SPN involving the portal vein, SMV and artery, spleen or duodenum^{2, 13, 14}. Even synchronous resection of liver metastasis is suggested because greater than five year survival has been reported after their removal^{2, 3, 22, 23}. Metastatic disease may occur in up to 20% of patients which are mostly confined to liver, mesentery and peritoneum^{2, 3, 22}.

Pancreatic fistula is the most common post-operative complication. Other reported complications include pancreatic abscess, intestinal obstruction, cholangitis, pneumonitis and wound infection^{13, 14, 23}.

The role of laparoscopic surgery has developed over years to include pancreatic surgery. However, its role for malignancy is reported to be less than 10% and long term results are still awaited²⁴. Laparoscopic surgery may be a reasonable option for SPN of pancreas due to their low malignant potential and younger population of patient^{16, 24}.

CONCLUSION

SPN of pancreas are rare and one of few malignant pancreatic tumors that can be treated effectively if diagnosed correctly. SPN is more common in females of reproductive age and present as abdominal mass or pain. It has distinct feature in imaging and histopathology. Surgical resection is the main stay of therapy. Laparoscopic surgery may be a reasonable option.

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