

Solid Pseudopapillary Neoplasm of the Pancreas: Case Series of a Rare Tumor

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ABSTRACT

Solid pseudopapillary neoplasm is an infrequent type of pancreatic tumor that primarily occurs in young females. This retrospective case series analyzed three cases of SPN at Civil Service Hospital from 2012-2022. Patient's median age was 18 years, and one of the patients presented with painless lump and other two had abdominal pain and dullness at presentation. Pre-operative investigations like CT/MRI and fine-needle aspiration cytology confirmed SPN in all cases. All patients received resection treatment, and no adjuvant therapy was administered. During follow-ups lasting 4-7 years, none of the patients experienced recurrence, indicating that surgical resection is the mainstay of treatment with excellent prognosis.

Keywords

Solid pseudopapillary neoplasm; pancreatic tumor; case series; Whipple's procedure

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INTRODUCTION

Solid pseudopapillary neoplasm (SPN) of the pancreas accounts for 1-2% of pancreatic neoplasms.¹ The tumor has undergone several name changes over the years, but the World Health Organization proposed the name 'solid-pseudopapillary tumor' in 1996.² Recent trends have shown rise in the number of reported cases of SPN, with 87.8% reported after 2000.³ Case reports and series have been reported from Nepal.^{4,5} Most individuals with this condition typically do not exhibit noticeable symptoms, but they may experience a slowly growing abdominal mass or experience nonspecific abdominal discomfort.³

Diagnosis is challenging due to the rare occurrence and non-specific symptoms, but characteristic findings of solid and cystic components with hemorrhage on CT/MRI help in the diagnosis.⁶ Surgical resection is the primary management, resulting in good prognosis, with reported survival at 5-year around 95%.⁷ Here, we present a retrospective analysis of three cases of SPN managed at our institute.

CASE 1

A 22-year female presented with a painless and non-progressive lump in the abdomen for 10 days. On abdominal examination, she had a firm, painless mass over the epigastric region with ill-defined margins, measuring 15 cm x 10 cm. Laboratory investigations showed elevated lipase (605 U/L) and amylase 4904 IU/L while liver functions, hematology, and renal function tests were normal.

Contrast-enhanced CT abdomen showed a multi-lobulated cystic lesion of size 13x9 cm in the retroperitoneum on the left side with few enhancing septations and minimal peripheral solid component abutting the pancreatic tail and upper pole of the right kidney- likely complex retroperitoneal cyst of the tail of pancreas D/D mucinous cystadenoma (Figure 1).

The patient underwent distal pancreaticotectomy and tumor excision, which revealed a large single

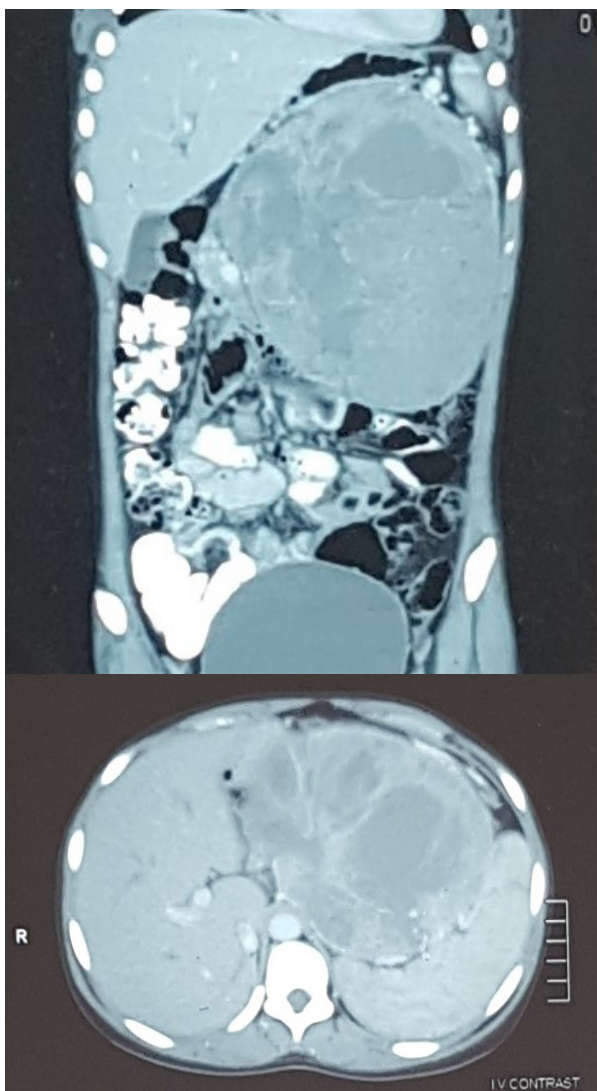


Figure 1. CT scan showing a huge mass in the retroperitoneum

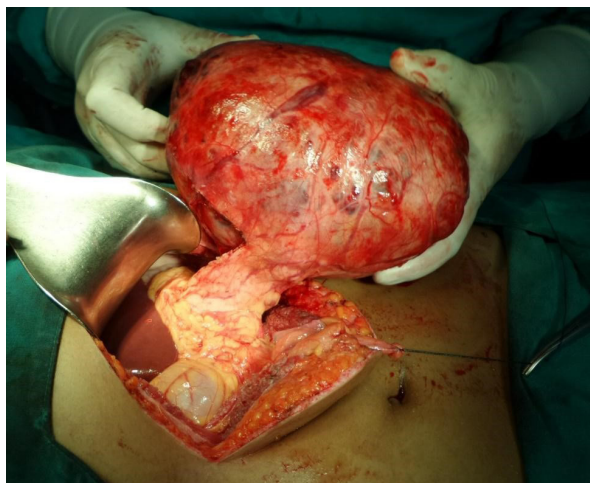


Figure 2. Intraoperative picture showing mass with its origin from pancreas

15x10 cm cystic tumor originating from the inferolateral surface of the pancreas near the tail. (Figure 2).

On histopathological examination, proliferation of tumor cells in pseudopapillary pattern, solid sheets and varying cysts was seen. The cells were monomorphic with a round to oval nuclei with vesicular chromatin and scant eosinophilic cytoplasm. Mitosis was not evident. Periphery was surrounded by a variable thin and thick capsule. Overall histological findings were suggestive of SPN. (Figure 3).

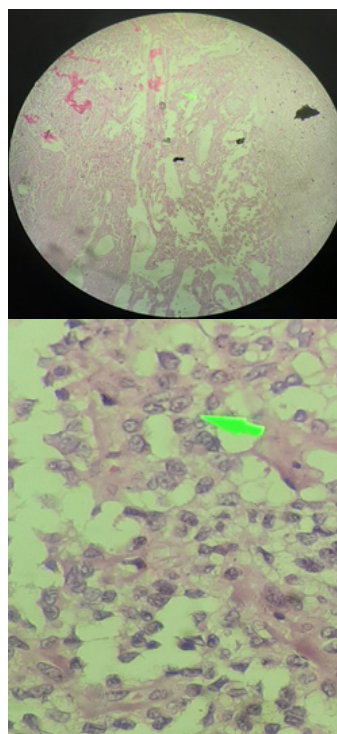


Figure 3. Photomicrograph showing features of SPN

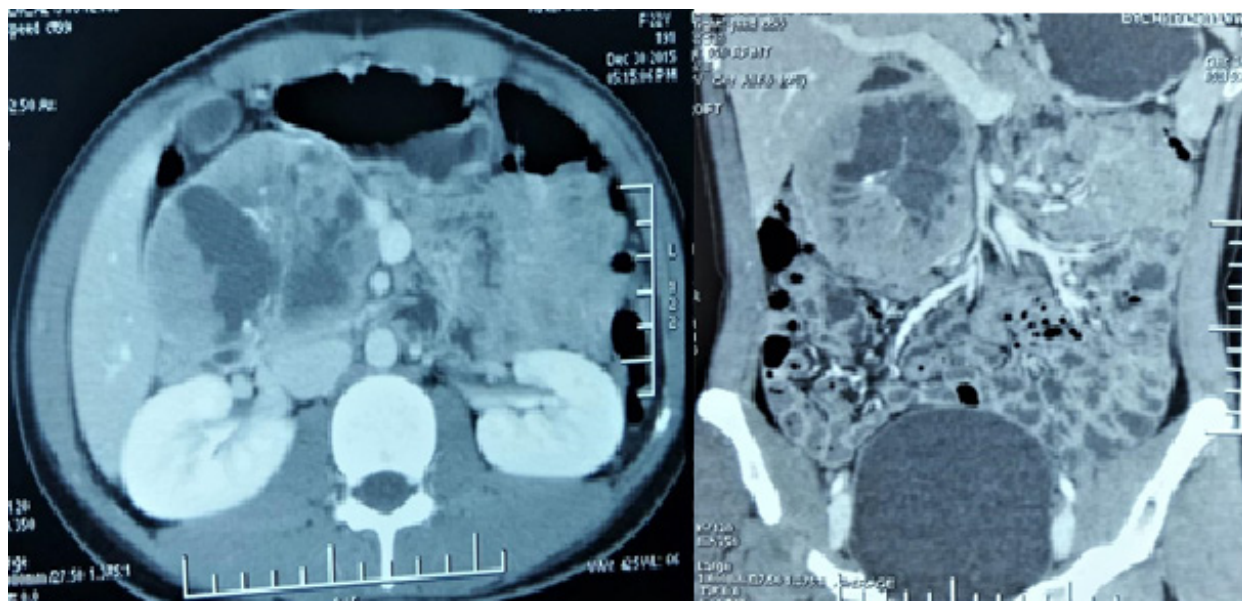


Figure 4. CT scan showing a mass in the pancreatic head with multiple cystic areas

The patient is on a 7-year follow-up with no recurrence or symptoms.

CASE 2

A 22-year female presented with dull aching upper abdominal pain for one month. Abdominal examination revealed a soft, pulsatile mass around 8x6cm over the epigastric region. Laboratory investigations showed elevated lipase (329 U/L) and amylase (30 IU/L). CT scans revealed a large solid hypoechoic mass of 94 x 79 x 75 mm at the pancreatic head with multiple cystic areas and calcifications, suggestive of a SPN of the pancreas. (Figure 4).

Fine needle aspiration cytology (FNAC) showed pseudo papillae, pseudo-rosettes, and clusters of small tumor cells in the background of metachromatic fibro-vascular stalks consistent with SPN. The lady underwent a Whipple's procedure, which revealed an encapsulated tumor measuring 8 x 6 x 2 cm at the pancreatic head with dense adhesions to third part of the duodenum.

Histopathological examination showed a pseudopapillary pattern of neoplastic cell proliferation with areas of sclerosis, hemorrhage, and foci of cholesterol clefts and calcifications. Cells appear monomorphic, having round to oval nuclei with nuclear indentations, occasional grooving, vesicular chromatin, and variable nucleoli. Cytoplasm was moderate in amount, granular, and eosinophilic to clear. Mitotic figures were not appreciated. Histological findings were suggestive of an SPN. The resection margins were all clear and none of the seven lymph nodes dissected were involved. (Figure 5).

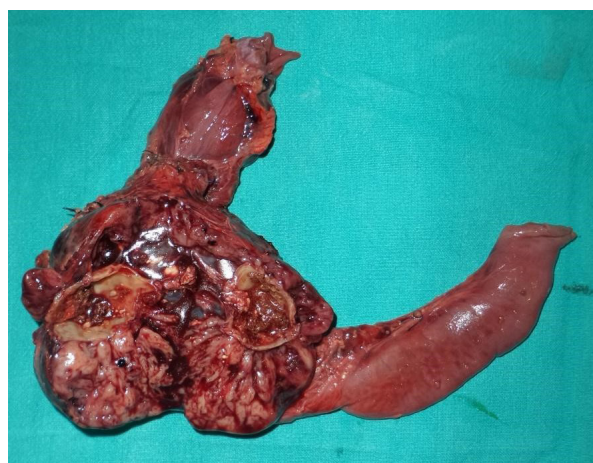


Figure 5. Gross appearance of tumor after removal

The patient was discharged on 10th post-operative day and is currently on 6th-year follow-up with no symptoms and recurrence.

CASE 3

A 12-year girl presented with complains of severe periumbilical pain and vomiting for a week. On examination she had dehydration and a large painful, pulsatile mass in the epigastric region. Laboratory investigations showed elevated alkaline phosphatase (240 IU/L) and lipase (380u/l).

MRI abdomen revealed well defined heterogeneous peripherally enhancing lesion at the head of the pancreas measuring 8.3 X 7.2cm. It was heterogeneously hypointense on T1 and heterogeneously hyperintense on FAT-saturated and T2-weighted images. In post-contrast study,

there was an enhancement of the wall. Overall radiological features were suggestive of SPN FNAC features were consistent with the SPN.

Whipple's procedure was performed. The operative findings showed a 10 cm x 7cm x 7 cm mass at the pancreatic head and dense adhesion with surrounding vessels and transverse colon. On histopathological examination, an encapsulated tumor showing extensive necrosis with minimal viable tissue was identified. The tumor was characterized by a distinct pattern wherein uniform polyhedral cells were arranged on delicate fibrovascular cores. The cells exhibited a uniform, round morphology with cytoplasm ranging from eosinophilic to clear. The overall histological was characteristics of SPN. No evidence of tumor involvement in resection margins, perineural, or lymphovascular invasion. The patient had an uneventful recovery. She is currently on a 4-year follow-up with no symptoms or recurrence.

DISCUSSION

SPN is a rare tumor typically affecting adolescent girl.¹ It may present with a variety of symptoms, but common are abdominal pain and a painless lump. In our study, one patient presented with a painless lump, and two presented with abdominal pain. While SPN can occur in any part of the pancreas, it is more common in the body or tail (61%).³ Two out of our three patients were of the pancreatic head.

Around 80% of patients typically display characteristic imaging features on CT or magnetic resonance imaging (MRI), revealing solid and cystic or solid tumors with uneven density within the capsule.⁸ Two of three patients in our series have imaging highly suggestive of SPN. Literature has shown elevated amylase in 22.6%, and lipase in 29.3%.⁹ All three of our patients have elevated lipase with two of three patients having slightly elevated lipase and one having marked elevation. Only one patient of ours has elevated amylase.

The cytomorphology can be highly accurate in distinguishing SPN from other solid or cystic pancreatic tumors.¹⁰ In our series two out of three patients underwent FNAC pre-operatively, and in both cases diagnosis of SPN could be clinched.

Surgery is the Primary treatment for SPN, with complete resection being the goal. The choice of surgical procedure is contingent upon the tumor's specific location. Despite the large size of many SPNs, complete resection can often be achieved due to the well-defined capsule and clear boundary of the tumor.¹¹ Complete resection could be achieved in all three of our patients.

Histopathologically, the tumor exhibits a combination of solid, pseudo-papillary, and cystic regions. The pseudo-papillary region consists of

structures resembling papillae, surrounding the tumor cells' central small vessels. Cystic areas often display hemorrhage and necrosis. Hematoxylin and eosin staining frequently reveals tumor cells with eosinophilic cytoplasm and round and centered nuclei. Nuclear mitosis and atypia are uncommon.¹¹ Our series also showed similar histopathological features and definite diagnosis of SPN was clinched on histopathology.

Immunohistochemical (IHC) markers are usually not helpful because of lack of specific markers for SPN, with different IHC used with variable results.¹² As there are no definite IHC for SPN, and histomorphological appearance were almost diagnostic none of our patients underwent further IHC.

Typically, SPN demonstrate a favorable prognosis following surgical intervention, with a low recurrence rate of approximately 10-15% and a mortality rate of only 2%. The overall 5-year survival rate for individuals diagnosed with SPN is approximately 95%.¹³ None of the patients in our series have recurrence with a follow up of 5-7 years.

CONCLUSION

SPN is a uncommon neoplasm primarily affecting young and adolescent female. Proper pre-operative imaging and cytology can accurately diagnose the tumor preoperatively. Surgery is usually feasible even in large tumors and mostly curative. However, it is recommended to closely monitor patients to promptly detect any signs of local recurrence or distant metastasis.

CONSENT

Written informed consent was obtained from the patient for publication of this case report.

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CONFLICT OF INTEREST

The author(s) declare that they do not have any conflicts of interest with respect to the research, authorship, and/or publication of this article.

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