(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 6, Issue 2 , Page No: 910-915
March-April 2023

IJMSCR

Solid Pseudopapillary Tumour of Pancreas- A Report Of Two Cases

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Type of Publication: Case Report
Conflicts of Interest: Nil

Abstract

Solid pseudopapillary tumour of pancreas is a rare neoplasm with low grade malignant potential, constituting less than 2 percent of all exocrine pancreatic neoplasms. It predominantly affects young women in the second or third decade of life. Characteristic radiological findings combined with age and sex profile should suffice for a decision to operate. Distinctive histopathological features are sufficient to make a confirmatory diagnosis, alleviating the need for immunohistochemical confirmation. Although it is a relatively indolent entity with favourable prognosis, patients with malignant neoplasm should have careful follow-up. We present a series of two cases reported over a one-year period.

Keywords: Solid pseudopapillary tumour, Pancreas, Immunohistochemistry

Introduction

Solid pseudopapillary tumour of pancreas is a rare neoplasm of low-grade malignant potential, initially described by Frantz in 1959 as “papillary cystic tumor of the pancreas” in the Armed Forces Institute of Pathology band on tumors of the pancreas.[1–4] It was incorporated by World Health Organization in 1996 as “solid pseudopapillary tumor” in the histologic classification of exocrine tumor of the pancreas.[2] It accounts for 0.9-2.7% all exocrine pancreatic neoplasms and only 5% cystic neoplasms. It occurs predominantly in adolescent girls and young women in the second or third decade of life and shows no apparent ethnic predilection.[1,5] It is a relatively indolent entity with favorable prognosis; metastases rarely occur.[5] In this article, we present two cases which were reported in a period of one year.

Case Report

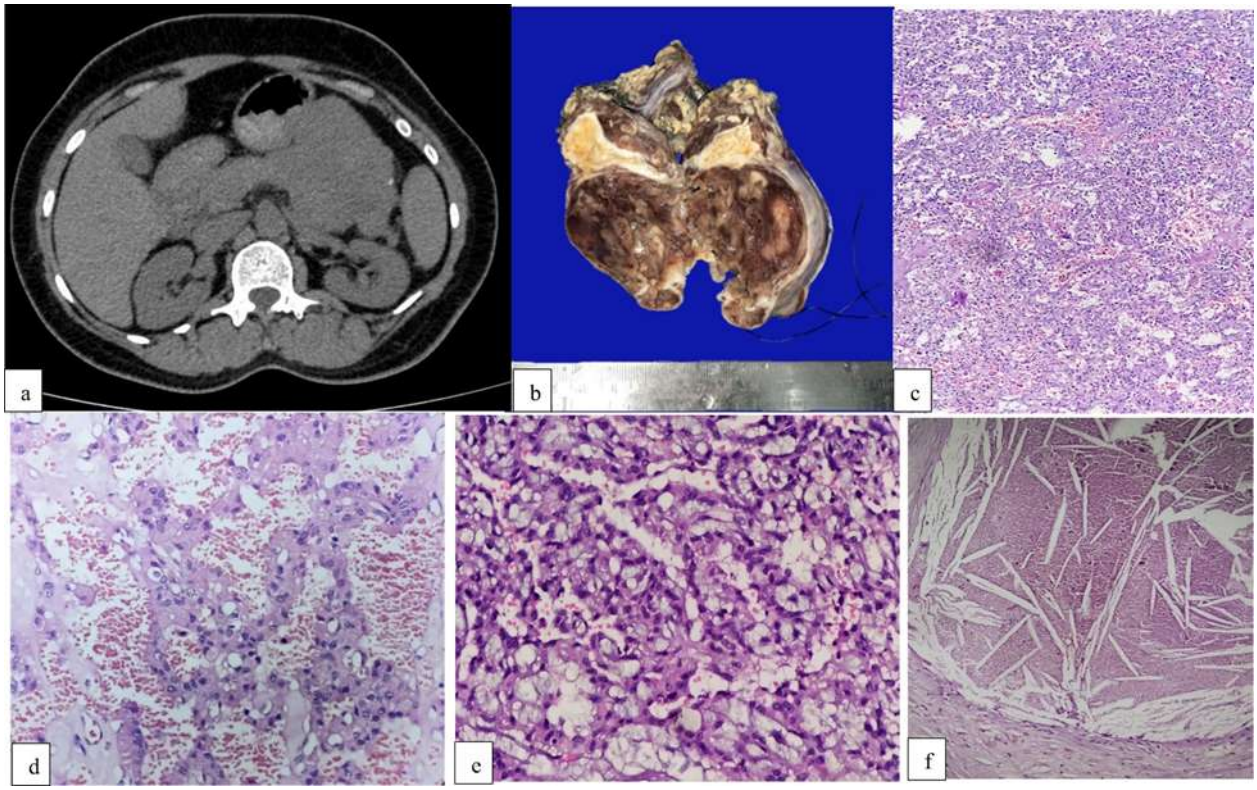
Case 1:

A 32-year-old female presented with complaints of pain in the left hypochondriac region for 10 days. There was no history of radiation of pain, vomiting or loss of weight. On examination, tenderness was present in the epigastrium, left hypochondrium and left renal angle. A 6x6 cm firm mass was palpable in left hypochondrium. CT contrast abdomen revealed a well-defined heterogeneously enhancing mass in the head and body of pancreas. (Figure 1a)

We received a partial pancreatectomy specimen measuring 9 x 8 x 4 cm with a tumour of size 7 x 6.5 x 5 cm in the body of pancreas. Parenchymal margins were uninvolved by 0.1 cm. Cut surface showed variegated appearance with necrotic and haemorrhagic areas. (Figure 1b)

Histopathological examination revealed a pseudopapillary pattern of polygonal to elongated tumour cells which was suggestive of Solid pseudopapillary tumor of pancreas, confined to the pancreas, with lymphovascular and perineural invasion. (Figures 1c, 1d and 1e).

Figure 1: a) CT contrast abdomen image



b) Gross image of partial pancreatectomy specimen

c,d,e) Pseudopapillary pattern of polygonal to elongated tumour cells with clear to eosinophilic cytoplasm and ovoid nuclei with longitudinal grooves and inconspicuous nucleoli. Adjacent areas of necrosis and hemorrhage are present. (x100, x200, x200, H&E)

f) Cholesterol crystals (x40, H&E)

Immunohistochemistry was performed with markers for confirmation of the diagnosis. Chromogranin A and synaptophysin showed negative cytoplasmic expressions, cytokeratin 7 showed negative cytoplasmic expression, and E-cadherin showed positive nuclear expression in the tumor cells. (Figure 2).

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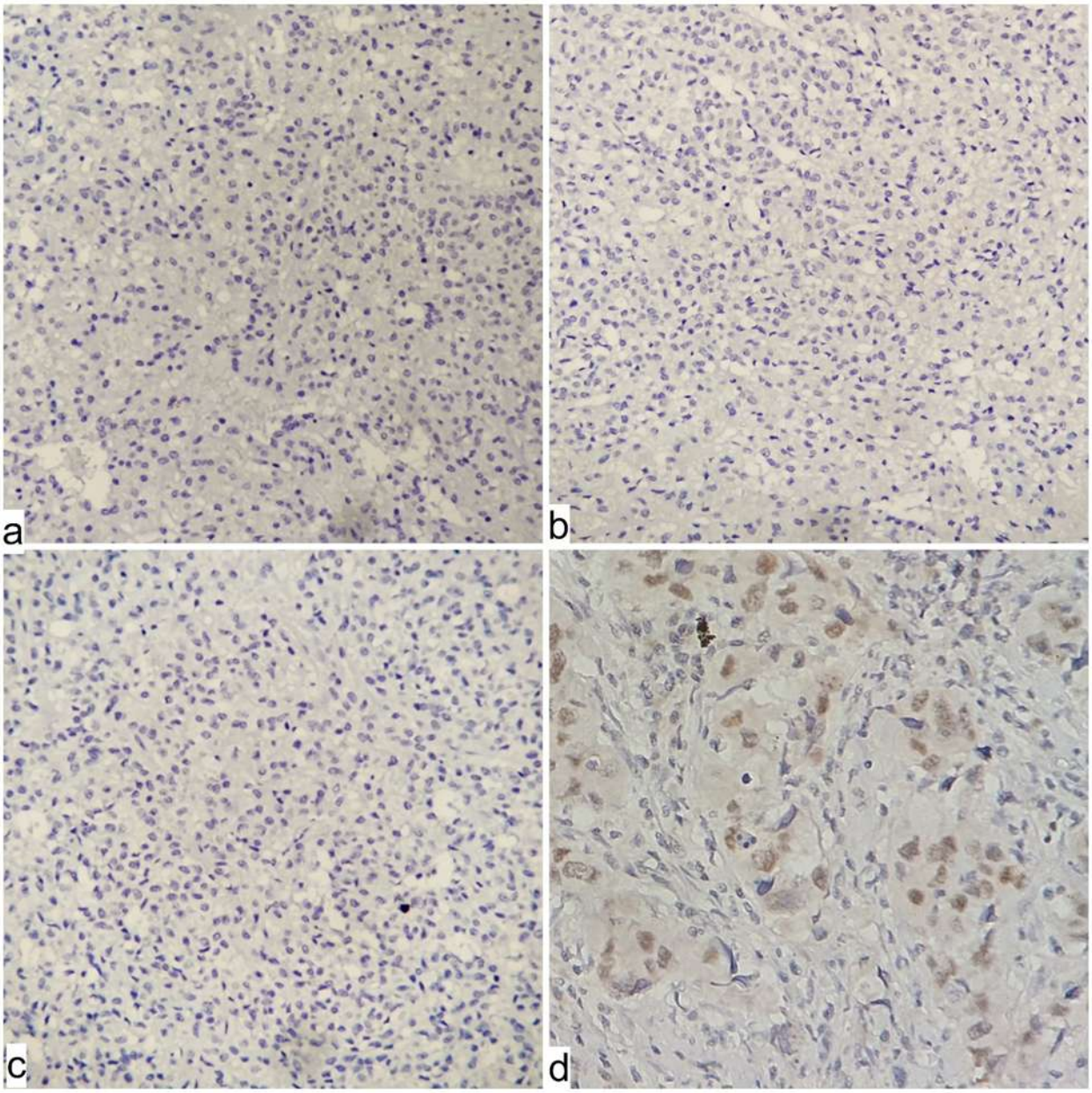
Figure 2: Immunohistochemical marker expressions-



1. Chromogranin A- negative
2. Synaptophysin- negative
3. Cytokeratin 7- negative
4. E-cadherin- positive

Case 2:

A 20-year-old female presented with complaints of abdominal pain for 1 month. No other significant history present. Examination revealed tenderness in the left hypochondriac region and a vague firm mass palpable in the epigastrium and left hypochondrium. CT-Abdomen contrast revealed a large well-defined heterogeneously enhancing solid-cystic lesion in the tail of pancreas, posteriorly compressing the splenic artery and vein. (Figure 3a)

We received a distal pancreatectomy specimen with a globular mass measuring 11 x 9.5 x 9 cm. Cut surface showed solid haemorrhagic and grey-brown areas, and cystic areas with haemorrhagic fluid. Maximum

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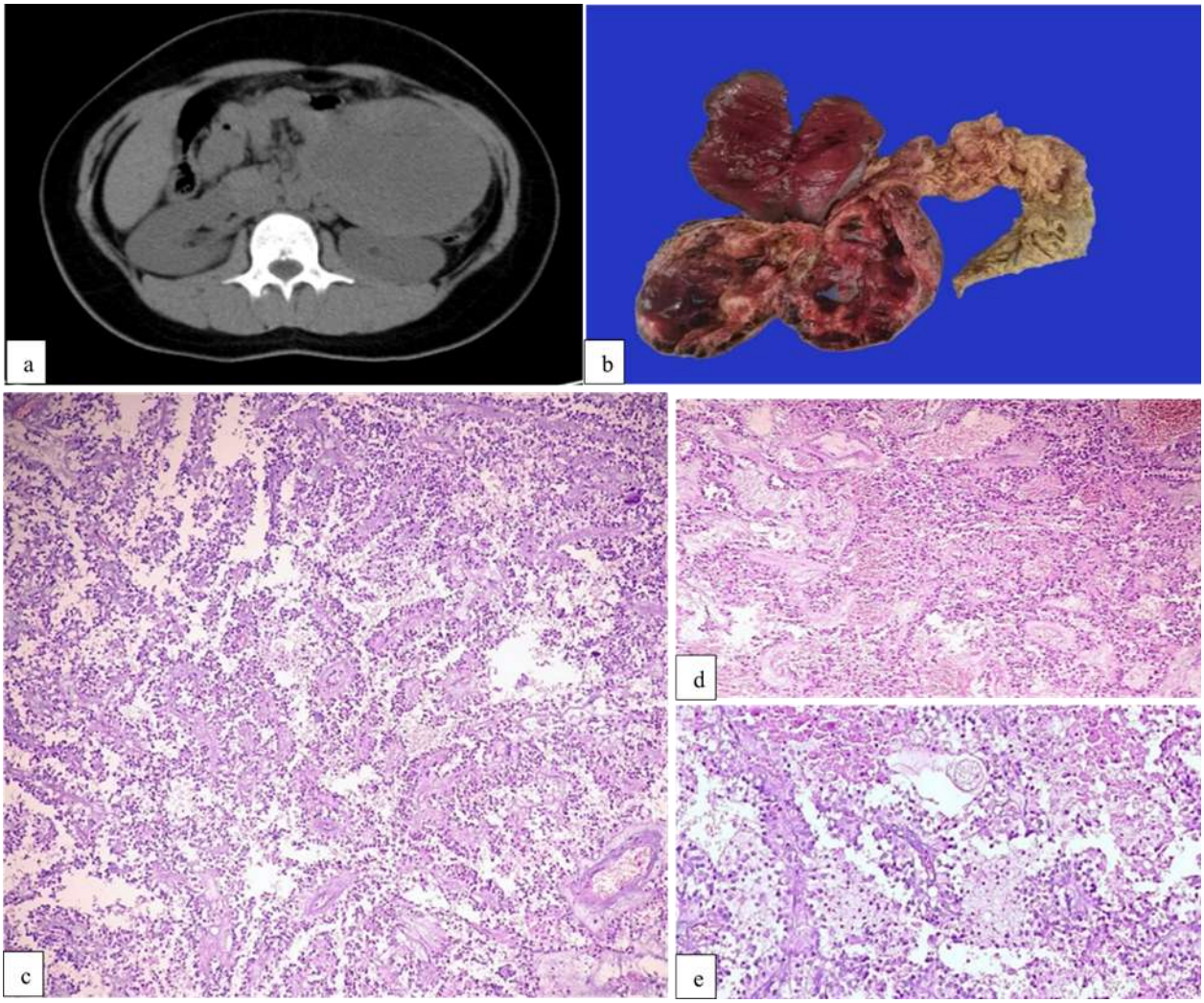
and minimum wall thickness were 2.8 cm and 0.2 cm respectively. (Figure 3b)

Histopathological examination showed sheets, nests, and cords of tumour cells around blood vessels along with areas of necrosis with pseudopapillary pattern of tumour cells, suggestive of Solid pseudopapillary tumor of pancreas, confined to the pancreas, with no infiltration into the adjacent parenchyma. (Figures 3c, 3d and 3e).

Figure 3: a) CT contrast abdomen image.



b) Gross image of distal pancreatectomy specimen.

c) Sheets, nests, and cords of tumour cells around blood vessels. (x100, H&E)

d) Areas of necrosis with pseudopapillary pattern of tumour cells, with moderate eosinophilic to vacuolated cytoplasm and uniform round to oval nuclei and inconspicuous nucleoli. (x100, H&E)

e) Sheets of foamy macrophages interspersed among tumour cells. (x200, H&E)

Discussion

SPPTs are rare neoplasms that are confined to the pancreas in 85% of patients and even the 10% to 15% of patients with liver or peritoneal metastases from SPTs commonly have long-term survival.[6] The presenting symptoms are usually either non-specific or absent, it is mostly diagnosed incidentally at ultrasound or CT imaging, They appear as a large

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mass well-circumscribed by a capsule, predominantly solid but sometimes with cystic degenerations, sometimes with calcifications.[7] It is difficult to make a precise diagnosis of SPPT based on the radiological images, especially with non-cystic masses.[7]

The molecular hallmark of SPNs is represented by point mutations in exon 3 of the CTNNB1 gene, which is involved in the Wnt/b-catenin signalling pathway, seen in more than 90% of cases.[8] In addition to its Wnt signalling role, β-catenin is a principal component of the adherens junction. E-cadherin is largely localized in the adherens junctions, mediating the adhesion between epithelial cells.[6] In contrast to conventional ductal adenocarcinoma of the pancreas SPTs are not associated with abnormalities in K-ras, p53, p16, or SMAD4/DPC4 genes.[6]

SPNs generally show a heterogeneous appearance, including various proportions of solid and pseudopapillary structures. The solid component is composed of uniform cells along with numerous delicate capillary-sized blood vessels. The pseudopapillary appearance is the result of neoplastic cells detaching from the capillary-sized blood vessels.[8]

Kim et Al[9] compared the clinicopathological features in pleomorphic SPPTs with conventional SPPTs. Except for the occurrence in older age group, the features of pleomorphic nuclei and a higher expression p53 protein expression, other features of pleomorphic SPNs, such as growth pattern, tumor size, infiltrative pattern, tumor extension, mitosis, and Ki-67 labeling index, were not different from those of conventional SPPTs. In addition, no difference in immunoreactivity for *β*-catenin and E-cadherin was seen.[9]

On immunohistochemistry, cells are consistently negative for mucin (ductal origin), enzymes (acinar origin) and hormones (endocrine origin), which supports the theory that SPPN arises from an embryonal pancreatic pluripotent cell.[10] Immunohistochemistry including acinar cell markers (trypsin, chymotrypsin, BCL10) or neuroendocrine markers (chromogranin and pancreatic hormones) is mandatory for the differential diagnosis.[8]

Conclusion

A diagnosis of SPPT pancreas should be highly suspected in young women with a pancreatic mass comprising of solid and cystic components. Although most commonly presenting with abdominal pain, it rarely can be an incidental imaging finding. Upto 15% of the cases present with extra-pancreatic metastases at the time of diagnosis. Since complete surgical resection offers a good prognosis, ruling out of other possible tumours and confirmation of diagnosis is absolutely necessary. Histopathology is sufficient for diagnosis of this tumor, but immunohistochemical markers E-cadherin or β-catenin can be used for confirmation when there are insufficient characteristic histopathological findings and/or clinical ambiguity or to rule out differential diagnoses. Although it is a relatively indolent entity with favourable prognosis, patients with malignant neoplasm should have careful follow-up.

Acknowledgements

I thank the faculty and technical staff of the Department of Pathology of Saveetha Medical College for all the help rendered by them.

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