

**CASE REPORT**

Pseudopapillary Tumor of Pancreas

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Abstract

Solid pseudopapillary tumor of the pancreas is a rare neoplasm usually seen in young females and is usually misdiagnosed as pancreatic pseudocyst. Awareness of clinical, microscopic and macroscopic features alongwith sufficient sampling of tumor is necessary for correct diagnosis of SPPT. We present three rare cases of SPPT of the pancreas.

Key Words

Solid Pseudopapillary Tumor , Pancreas, Neoplasm

Introduction

Solid pseudopapillary tumor (SPPT) of the pancreas is a rare neoplasm accounting for 0.2% - 2.7% of primary pancreatic tumors with slow growth rate and excellent prognosis (1). SPPT is usually seen in young females and is usually misdiagnosed as pancreatic pseudocyst. Awareness of clinical, microscopic and macroscopic features alongwith sufficient sampling of tumor is necessary for correct diagnosis of SPPT. We present three rare cases of SPPT of the pancreas.

Case 1

A 30 year female presented to Department of Surgical Gastroenterology with complaint of upper abdominal pain for last two years. Ultrasound (USG) examination revealed a well defined, encapsulated mass lesion in pancreatic body. Contrast enhanced Computed Tomography (CT) scan revealed a well defined, non-calcified, nonenhancing solid pancreatic mass lesion in body region. Pancreatic duct was normal. Splenic vein was compressed by mass lesion. Spleen preserving distal pancreatectomy was done. Per-operative examination revealed a well defined 8 x 7 cm solid mass in pancreatic body region, extending medially 2 cm lateral to superior mesenteric vein (SMV). No evidence of peripancreatic lymphadenopathy was seen. Cut surface showed encapsulated fleshy tumor arising from body of pancreas with papillary excrescences with no cystic components or mucinous material. Histopathology of resected specimen revealed encapsulated tumor arranged in pseudopapillae, solid sheets, nests, trabeculae and microcystic areas alongwith areas of loose myxoid stroma and foci of stromal sclerosis. The tumor cells were monomorphic, oval to elongated and showed round to oval nuclei with dispersed chromatin. Occasional nuclear grooves were

seen. Fair number of extracellular as well as intracellular eosinophilic globules were seen. With these histological features, final diagnosis of SPPT of pancreas was made.

Case 2

A 14 year female presented to Department of Pediatric Gastroenterology with complaint of upper abdominal swelling for last three years. On clinical examination, swelling was solitary, smooth, non-tender, movable in side by side direction, non-pulsatile and non-fluctuant. USG examination revealed a well defined mass lesion in pancreatic body region. CT scan revealed a well defined, non-calcified, solid-cystic pancreatic mass lesion in body region. Pancreatic duct was normal and spleen was enlarged in size. Distal pancreatectomy alongwith splenectomy was done. Per-operative examination revealed a well defined 12 x 9 cm solid-cystic mass in pancreatic body region. No evidence of peripancreatic lymphadenopathy was seen. Cut section showed growth had granular appearance with embedded areas of haemorrhage and necrosis. Histopathology of resected specimen revealed partially encapsulated tumor arranged in sheets and trabeculae of small round cells. The cells had round to oval nuclei with finely dispersed chromatin, inconspicuous nuclei and eosinophilic cytoplasm. Occasionally cells showed nuclear grooving. Pseudorosetting alongwith presence of foam cells was also seen. Areas of hemorrhage and necrosis was also seen. With these histological features, final diagnosis of SPPT of pancreas was made.

Case 3

A 14 year female presented to Department of Pediatric Gastroenterology with complaint of upper abdominal swelling for last one year. On clinical examination,

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Fig 1. CT scan image showing a well defined, non-calcified, nonenhancing solid pancreatic mass lesion in body region.



Fig 2. Resected surgical specimen showing a fleshy tumor arising from body of pancreas with papillary excrescences with no cystic components or mucinous material.

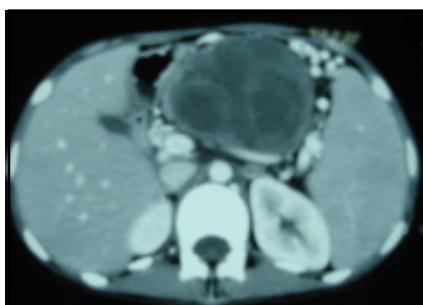


Fig 3. CT Scan Image - Well Defined, Non-Calcified, Solid-Cystic Pancreatic Mass Lesion in Body Region

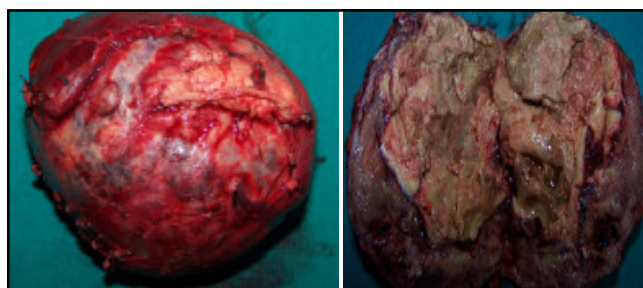


Fig 4 & 5. Resected Surgical Specimen Showing a Fleshy Tumor with Granular Appearance Arising From Body of Pancreas with Embedded Areas of Haemorrhage & Necrosis

swelling was solitary, smooth, non-tender, movable in side by side direction, non-pulsatile and non-fluctuant. USG examination revealed a well defined mass lesion in pancreatic body and tail region. CT scan revealed a well defined, capsulated, non-calcified, solid-cystic pancreatic mass lesion in body region. Pancreatic duct was normal. Splenic vein was compressed by mass lesion. Biopsy from mass lesion revealed findings suggestive of SPPT of pancreas. Patient is planned for distal pancreatectomy with spleen preserving surgery.

Discussion

Cystic pancreatic neoplasms were first classified by Robson and Moynihan in 1903(2). In 1959, Frantz described a rare tumor known as papillary tumor of the pancreas, benign or malignant (3). This tumor was known by various names like Gruber-Frantz tumor, solid and cystic tumor, solid and papillary epithelial neoplasm, papillary cystic tumor and solid-cystic papillary epithelial neoplasm. In 1996, the World Health Organization (WHO) renamed this tumor as SPPT for the international histological classification of tumors of the exocrine pancreas (4).

SPPT is predominantly seen in young women between the 2nd and 3rd decades of life (5). SPPT has predilection for persons of Asian origin and is probably the most common tumor in Asian children. Approximately 20% of cases of SPPT have been reported in children (6). In our

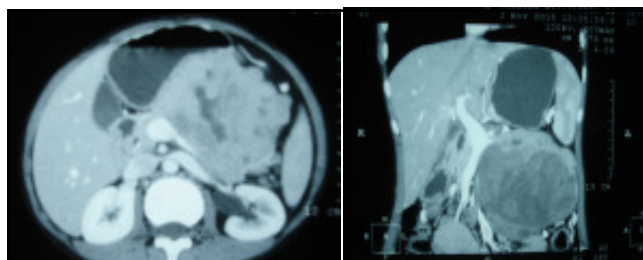


Fig 6 & 7. Axial and Coronal CT scan Image Showing a Well Defined, Capsulated, Non-Calcified, Solid-Cystic Pancreatic Mass Lesion in Body Region. Splenic Vein is Compressed by Mass Lesion

case series, all three were female and two were children. Rare cases of SPPT have also been reported in men (7,8). The presence of progesterone receptors and predilection for females suggests that SPPT is a hormone dependent tumor (9).

The cell of origin of SPPT is unknown (10). Some authors have reported SPPT as neoplasm of uncommitted cells with most cells similar to intercalated duct cells or centroacinar cells (11).

Pancreatic ductal and non-ductal neoplasms progress genetically though two primarily dichotomous pathways : K-ras, p 16, DPC4, and p53 gene alterations in the case



of ductal neoplasms, and varying rates of APC/?-catenin alterations in nonductal neoplasms, including virtually all SPPT (12).

The most frequent symptom of SPPT is upper abdominal pain seen in nearly half the patients. SPPT may occur anywhere in the pancreas but is most frequently found in the head or tail (13). In our series, all three were involving pancreatic body region.

The classic CT features of SPPT are a large well-encapsulated mass with varying solid and cystic components caused by hemorrhagic degeneration (14, 15). Calcifications and enhancing solid areas may be present at periphery of mass. Atypical appearances include : metastasis to liver, ductal obstruction, extra-capsular invasion, dense calcification and male patients (15). In our series, all three were well encapsulated, noncalcified mass lesions with no evidence of ductal obstruction, metastasis or extra-capsular spread.

Nishihara K and coworkers reported venous invasion, diffuse growth pattern, extensive tumor necrosis, significant nuclear atypia and high mitotic rate as indicators of malignant potential of SPPT (16). According to WHO classification, SPPT with clear criteria of malignancy are designated as solid-pseudopapillary carcinomas (4,13).

The differential diagnosis of SPPT includes pseudocyst of pancreas, neuroendocrine tumor and ductal adenocarcinoma of pancreas. Ductal adenocarcinoma is most commonly seen in elderly male and grows in a infiltrative manner. Neuroendocrine tumors present with a solid or microacinar pattern and nuclei are small, round, smoothly contoured with fine chromatin. Moreover, peripheral portions of neuroendocrine tumors show hypervascularity on arterial phase CT images. Absence of history of pancreatitis in presence of solid and cystic pancreatic mass in young age group patient must raise suspicion for SPPT (17,18).

Diagnosis of SPPT is usually made only after operative biopsy. SPPT has very good prognosis unlike ductal adenocarcinoma of pancreas. Tang LH and coworkers have described overall 5 year survival rate of 97%, even in the presence of disseminated disease (13). Even benign SPPT tumors have malignant potential and correct treatment by total surgical excision is curative. Our case series highlights importance of awareness of SPPT as a distinct class of pancreatic tumor in young females with excellent prognosis.

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