A classic case of Solitary Pseudopapillary Epithelial Neoplasm of Pancreas - Case report with Review of Literature

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ABSTRACT

We report a case of a solid pseudopapillary epithelial neoplasm of the pancreas in a 19-year-old female who presented with abdominal pain. Computed tomography of the abdomen showed a heterogenous density mass in the tail of the pancreas. The mass had fluid density in the central region and soft tissue density in the peripheral region. Post-contrast evaluation showed enhancement in the peripheral solid component. Based on the radiological investigation carried out at our institute, a diagnosis of solid pseudopapillary epithelial neoplasm of the pancreas was established which was confirmed on histopathology.

CASE REPORT

CASE REPORT

A 19-year-old woman presented to the surgery out-patient department in our institute with the chief complaints of left sided epigastric pain for a period of about 2 months. The pain was associated with nausea and fullness. She did not have any history of trauma or any past surgeries. On physical examination, tenderness was noted in the epigastric region. The patient had undergone ultrasound of the abdomen at a private clinic 3 days back which suggested pseudocyst of the pancreas. The woman was advised routine blood investigations which were within normal limits. The serum amylase (32 U/L) and lipase (39 U/L) levels were in the normal range. Patient was scheduled for a contrast enhanced computed tomography of the abdomen for further evaluation and to plan for surgery.

On plain computed tomography of the abdomen, a well-circumscribed almost round lesion measuring approximately 5.29 x 5.38 x 7.02 cm (AP x Trans x CC) i.e., approximately 100 cc was noted in the tail of the pancreas. This lesion showed heterogenous density with fluid density in the central region and soft tissue density in the peripheral region. Few tiny peripheral wall calcifications were noted in the lesion (Fig. 1a, 2a). The lesion showed beak sign with the tail of pancreas indicating pancreatic origin (Fig. 1b). On post-contrast evaluation, the peripheral solid component along with the wall showed significant enhancement whereas the central cystic component appeared to be non-enhancing (Fig. 1b, Fig. 2b). The main pancreatic duct and the common bile duct were of normal calibre. Thereafter, ultrasound of the abdomen was performed which revealed similar findings as that of computed tomography. A heterogenous lesion in the tail of pancreas with cystic component in the centre and solid component in the periphery was appreciated on ultrasound (Fig. 3a, 3b, 4a). The solid component demonstrated vascularity on colour doppler imaging (Fig. 4b). These classic imaging features suggested the

diagnosis of Solid Pseudopapillary Epithelial Neoplasm (SPEN)

The patient underwent a distal pancreatectomy. No evidence of tumour spread or lymphatic involvement was noted intraoperatively. On pathological examination, diagnosis of SPEN was confirmed. The patient's postoperative recovery was unremarkable, and she was discharged on postoperative day 5. No additional therapy was required.

DISCUSSION

Etiology & Demographics:

Solid pseudopapillary epithelial neoplasms, also known as Franz tumors or Hamoudi tumors are uncommon cystic tumors of the pancreas that have a distinctive histology and are essentially tumors that have good prognosis. These tumors account for 3% of the cystic neoplasms of pancreas [1]. SPENs typically occur in the second and third decades of life with the tumor occurring five times more often in females than in males and a predilection for women of Asian and African American race, while few occurrences have been reported in children and men. According to Mao et al, the mean age of occurrence is 23.9 years in females [2]. The patients present with nonspecific signs and symptoms with upper abdominal pain being the most common symptom [3]. SPEN is becoming increasingly regularly documented in the literature as its pathological aspects become better known [4].

Clinical & Imaging findings:

SPENs most commonly arise from the tail or head of pancreas, however, there have been few case reports where it was found in ectopic pancreatic tissue in the mesocolon [4]. These tumours usually tend to be large, well-encapsulated and thick walled with a slow rate of growth progression. Varied degrees of solid and cystic internal component with haemorrhage and cystic degeneration in the centre of the lesion is a typical feature in case of SPENs [5].

Ultrasound, computed tomography (CT), magnetic resonance imaging (MRI), and angiogram may be used to help characterize these pancreatic masses. On ultrasonographic evaluation, SPENs demonstrate a heterogenous mass with areas of internal hemorrhage and cystic degeneration with fluid-debris levels within and posterior acoustic enhancement owing to their cystic nature. CT demonstrates a heterogenous density mass with fluid density in the central region and soft tissue density in the peripheral region. Cystic portions suggesting hemorrhagic necrosis and calcification may be appreciated on CT [4]. Contrast evaluation demonstrates enhancement in the soft tissue component.

Despite the tumour growing to a considerably large size, there are very few incidences of the tumour causing dilatation of the pancreatic duct or the biliary system. Also, locoregional invasion is rarely seen in SPENs [6]. These tumours have low malignant potential and even if they metastasize, it is most commonly to the liver followed by regional lymph nodes, mesentery, omentin, and peritoneum [7]. Features that point towards malignancy include a focal breach or discontinuity in the tumour capsule, large size of the tumour (> 6 cm) and a pancreatic tail location. The malignant tumours often present in older age group and have a male predilection: they are designated solid pseudopapillary carcinomas [8]. As opposed to this, solid or near solid tumours and the presence of diffuse calcifications within the tumour may indicate benign nature of the tumour [9].

Cantisani et al. published a study in the year 2003 which concluded that MRI is superior to CT for differentiating certain tissue components in case of SPENs. The presence of a capsule, cystic degeneration and haemorrhagic component were better appreciated after gadolinium administration on dynamic MRI examination. A well-encapsulated lesion with heterogenous signal intensity on T1 and T2 weighted images which indicate that the mass has a complex nature are demonstrated on MRI evaluation. T2 weighted imaging shows a thick fibrous capsule, which is seen as a rim of low signal intensity. On gadolinium-enhanced dynamic MR imaging, early heterogenous enhancement of the fibrous capsule with progressive fill-in is the typical feature [10]. However, peripheral region of SPENs do not exhibit the hypervascularity typically seen in islet cell tumours [6].

Treatment & Prognosis:

Although radiological evaluation has certain characteristic features, they cannot conclude upon a definitive diagnosis alone, since there is significant overlap among the cystic pancreatic lesions. In such cases, histopathological evaluation is necessary since it is the gold standard. Endoscopic ultrasound guided FNAC of the pancreatic tumor followed by cytologic analysis plays an extremely important role to differentiate SPENs from other pancreatic lesions pre-operatively [11]. SPENs have a good prognosis and long-term survival rates, especially post-surgery. Even in case of metastasis, complete surgical resection of the tumor with chemotherapy is known to have good prognosis with a reported 5-year survival of 97%. Hence, complete resection is always the treatment of choice, not only because of the risk of malignancy, but also owing to the magnitude to which these tumors can grow [6]. For this reason, it is essential to arrive to a definitive diagnosis pre-operatively [12].

Differential Diagnoses:

The differential diagnosis of SPENs mainly include pseudocyst of pancreas, serous cystic neoplasm, mucinous cystic neoplasm, intraductal papillary mucinous neoplasm [13]. Characterization of the lesion based on imaging alone is far-fetched however, a definitive diagnosis is often attainable when the lesions demonstrate typical radiologic features [3].

1. Pancreatic pseudocyst

Characteristic features in case of pseudocyst of the pancreas include a unilocular cyst without the presence of solid components, central scar, or wall calcification. It shows mild wall enhancement on contrast evaluation. They may involve the head, body, tail or uncinate process of pancreas and in rare case reports, have been found within the abdomen and even in the chest. A clinical history of pancreatitis or abdominal trauma usually precedes the development of pseudocyst which is a major feature differentiating it from SPENs [3].

2. Serous cystic neoplasm

The old name for serous cystic neoplasm was serous cystic cystadenomas which accounts for 16% of the pancreatic neoplasms. They are classified based on macroscopic variation into microcystic and macro cystic subgroups [6]. These tumors are more common in females with a median age of 65 years which is a major factor that helps differentiate it from SPENs [5]. In the microcystic subgroup, a collection of multiple small cysts that range in size from a few millimeters to up to 2 cm are found within giving it the classic honeycomb appearance on CT or MR imaging. These lesions may appear solid on CT due to the compact arrangement of the cysts and so MRI or EUS may be needed for further characterization. Multiple septae that enhance on post-contrast evaluation are noted due to the hyper vascular septae which may also result in intra-lesional hemorrhage [6]. In case of serous cystic neoplasms, a fibrous central scar with or without a characteristic stellate calcification is considered to be pathognomonic [14]. The macro cystic variety also known as the oligocystic variety has fewer cysts within (1-2 in number) with the cysts generally being greater than 2 cm in size. The macro cystic variant is difficult to differentiate from MCN due to the large internal cysts [6].

3. Mucinous cystic neoplasm

These neoplasms account for 23% of pancreatic cystic lesions [13]. These tumors occur almost exclusively in middle-aged women [3]. Most of the Mucinous cystic neoplasms (MCNs) are solitary lesions and are multilocular with few large compartments. However, they can occur as unilocular lesions with a single compartment as well. The tumors usually have a smooth external contour with a round or lobular appearance and often have a tendency to grow up to a large size [3]. MCNs have a complex internal architecture consisting of papillary projections into the lumen, peripheral cysts, septae, mural nodule or solid component. Along with this, peripheral eggshell or septal calcification on computed tomography evaluation are specific for MCNs and may predict malignant potential of the tumor. The fraction of internal mucoid content and hemorrhagic fluid alters the internal Hounsfield density of the tumor [15].

4. Intraductal papillary mucinous neoplasm

These neoplasms affect both the sexes with a slight male preponderance, in the sixth to seventh decade of life. Anatomical classification of IPMNs include main duct type, branch duct type, and combined type [16]. The main duct type of Intraductal papillary mucinous neoplasm (IPMN) does not present as a cystic lesion but as segmental or diffuse ductal dilatation that is disproportionate to the degree of pancreatic parenchymal atrophy. This is due to the excessive mucin produced by the intraductal papillary growth of the columnar epithelium [17]. “Bulging duodenal papilla” sign is seen on CT and ERCP in case of IPMN where excessive mucin secretion results in bulging of major papilla into the duodenal lumen and is pathognomonic of main duct type IPMN [14]. In case of branch duct type IPMN, unilocular or multilocular cystic lesions are noted to communicate with the main pancreatic duct. The uncinate process is the classic location of the side branch type of IPMN [16]. Mixed type of IPMN is an advanced form of branch type IPMN in which has involved the main pancreatic duct as well [14]. Mural nodules, thick septae, wall calcifications and segmental or diffuse dilation of the main pancreatic duct indicate increased likelihood of malignancy [17]. However, the best indicator of malignant potential is the anatomical location of the tumour, with the main duct type having a 70% and the branch duct type having a 25% likelihood of harbouring malignancy [18].

TEACHING POINT

Solid pseudopapillary epithelial neoplasm is an uncommon condition that can be treated successfully with total surgical excision in young adults and adolescents. Recognition of the classical imaging features would help us to arrive at the prompt and accurate diagnosis of SPENs. It is critical to have a proper diagnosis of this tumour since a well-planned excision usually results in a complete cure.

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FIGURES

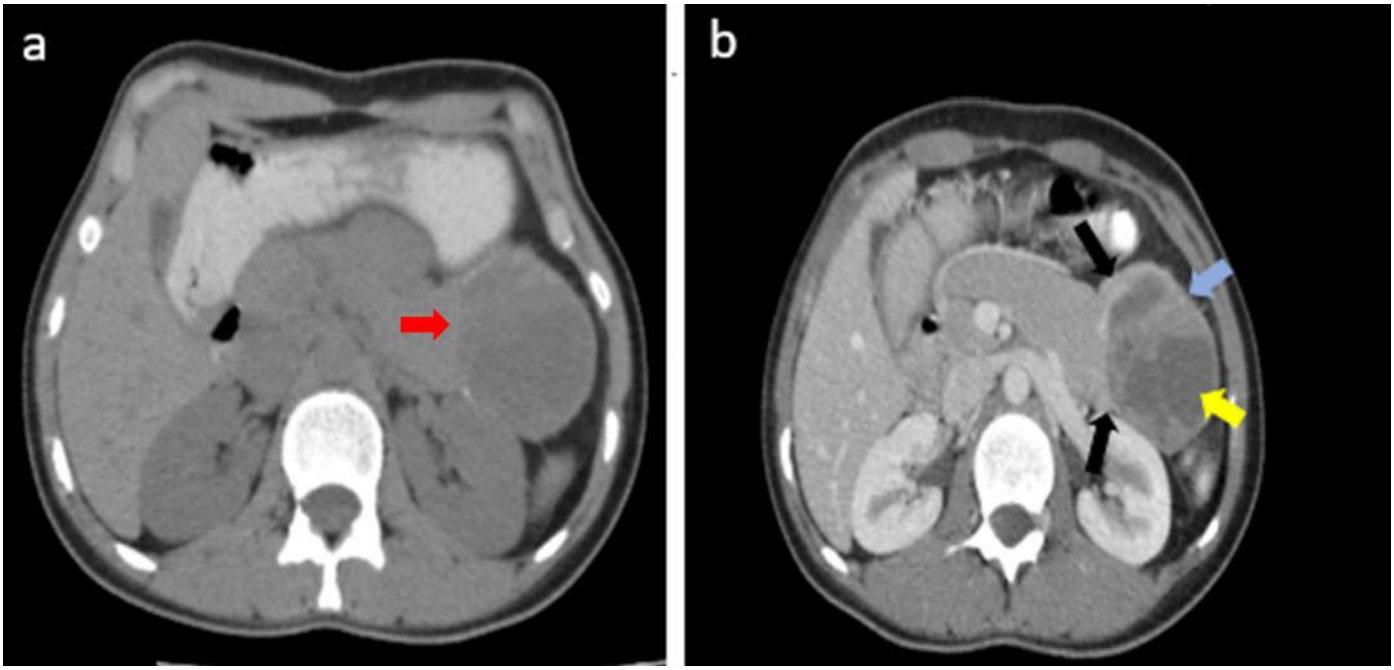

Figure 1: 19-year-old female with Solid Pseudopapillary Epithelial Neoplasm of the pancreas.

Findings: Axial section in plain non-contrast CT scan showing solid cystic lesion in the tail of pancreas (red arrow) in image measuring approximately 5.29 x 5.38 x 7.02 cm (AP x Trans x CC) i.e., approximately 100 cc (1a). Axial post-contrast image showing peripheral enhancing solid component (blue arrows) and central cystic components (yellow arrows) in image (1b). Beak sign with tail of pancreas (black arrows) in image (1b).

Technique: Axial multi-detector non-contrast CT of the abdomen, mA 50-200; kV 120; 2 mm slice thickness in image (1a). Axial Multi-detector CT, Arterial phase (100 ml Contrapaque), mA 50-200; kV 120; 1 mm slice thickness in image (1b).

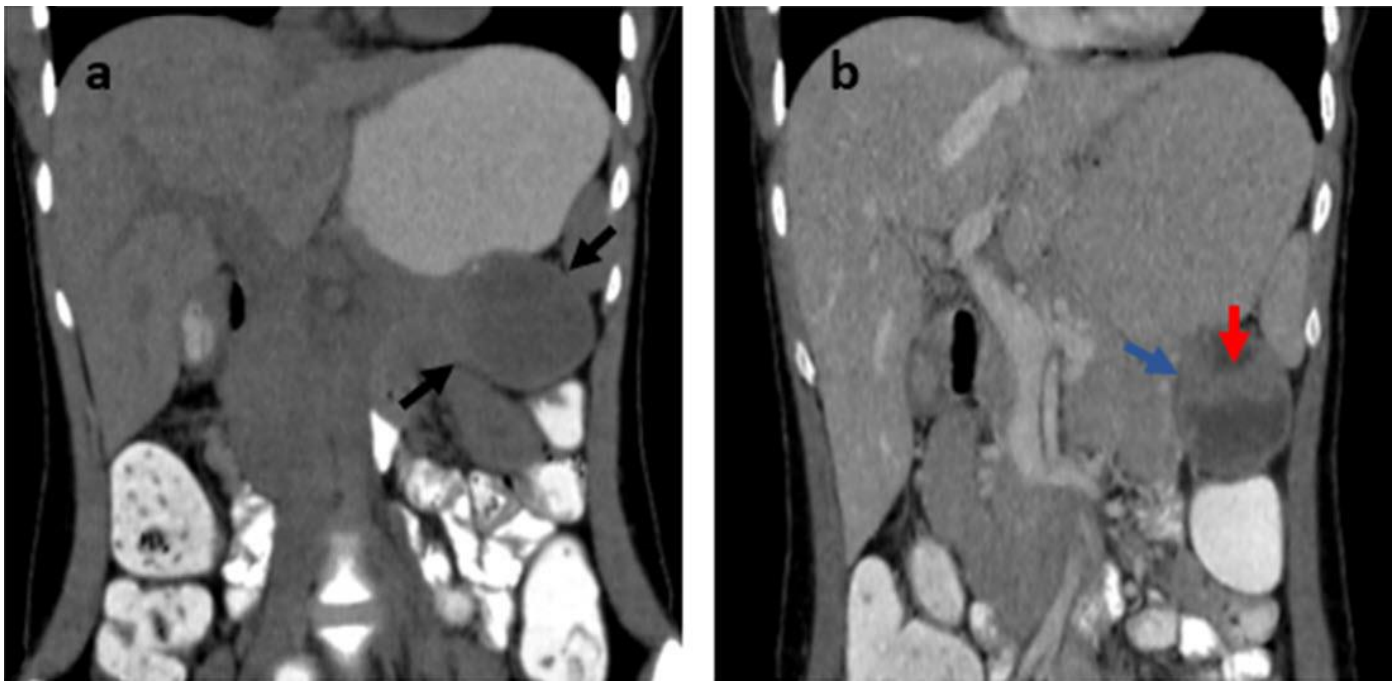

Figure 2: 19-year-old female with Solid Pseudopapillary Epithelial Neoplasm of the pancreas.

Findings: Coronal section in non-contrast CT scan showing solid cystic lesion (black arrows) in image measuring approximately 5.29 x 5.38 x 7.02 cm (AP x Trans x CC) i.e., approximately 100 cc (2a). Coronal post-contrast image showing peripherally enhancing solid component (blue arrows) and few centrally enhancing solid components (red arrows) in image (2b).

Technique: Non-contrast multi-detector CT of the abdomen, mA 50-200; kV 120; 2 mm slice thickness in image (2a). Multi-detector CT, Arterial phase (100 ml Contrapaque), mA 50-200; kV 120; 2 mm slice thickness in image (2b).

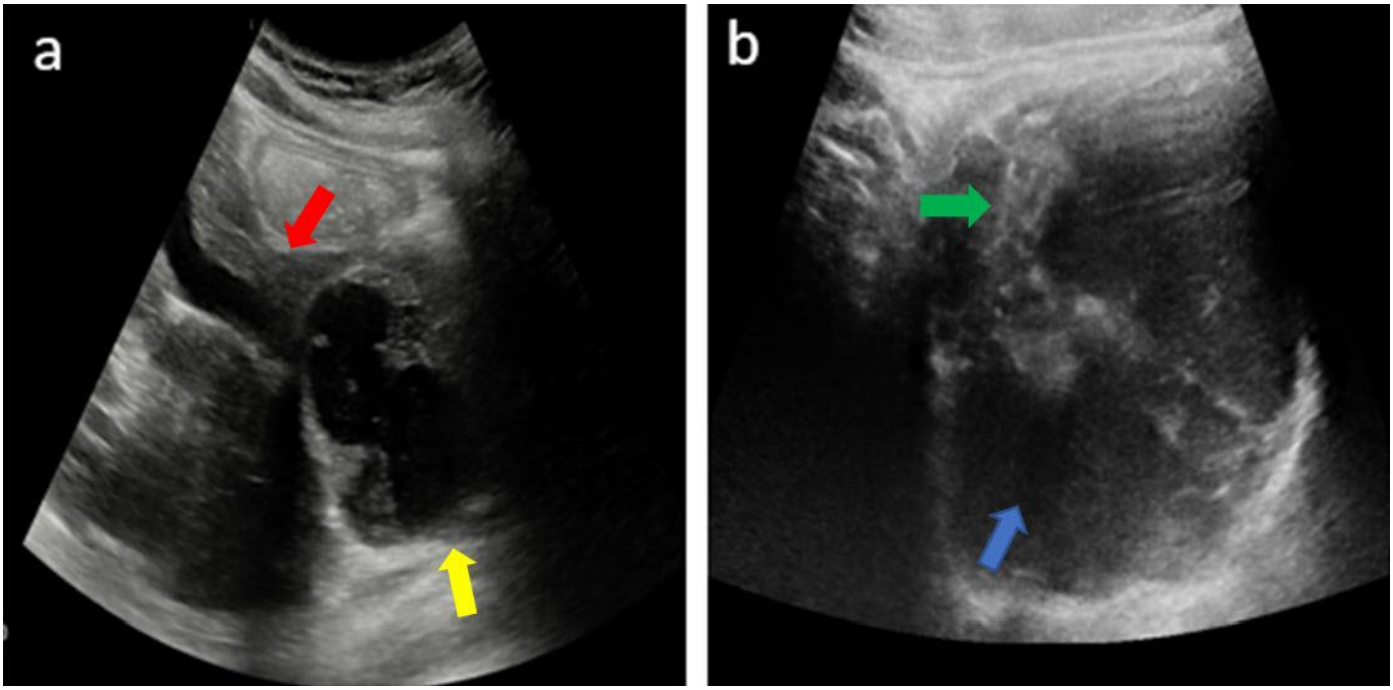

Figure 3: 19-year-old female with Solid Pseudopapillary Epithelial Neoplasm of the pancreas.

Findings - B-Mode image showing solid cystic lesion (yellow arrow) arising from the tail of pancreas (red arrow) in image (3a). Heterogenous lesion showing solid (green arrow) and cystic component (blue arrow) in image (3b).

Technique: Ultrasound of the epigastric region of abdomen, using a 1-7 MHz convex array transducer (Samsung RS80A), in axial plane in image (3a). Ultrasound of the epigastric region of abdomen, using a 3-12 MHz linear array transducer (Samsung RS80A), in axial plane in image (3b).

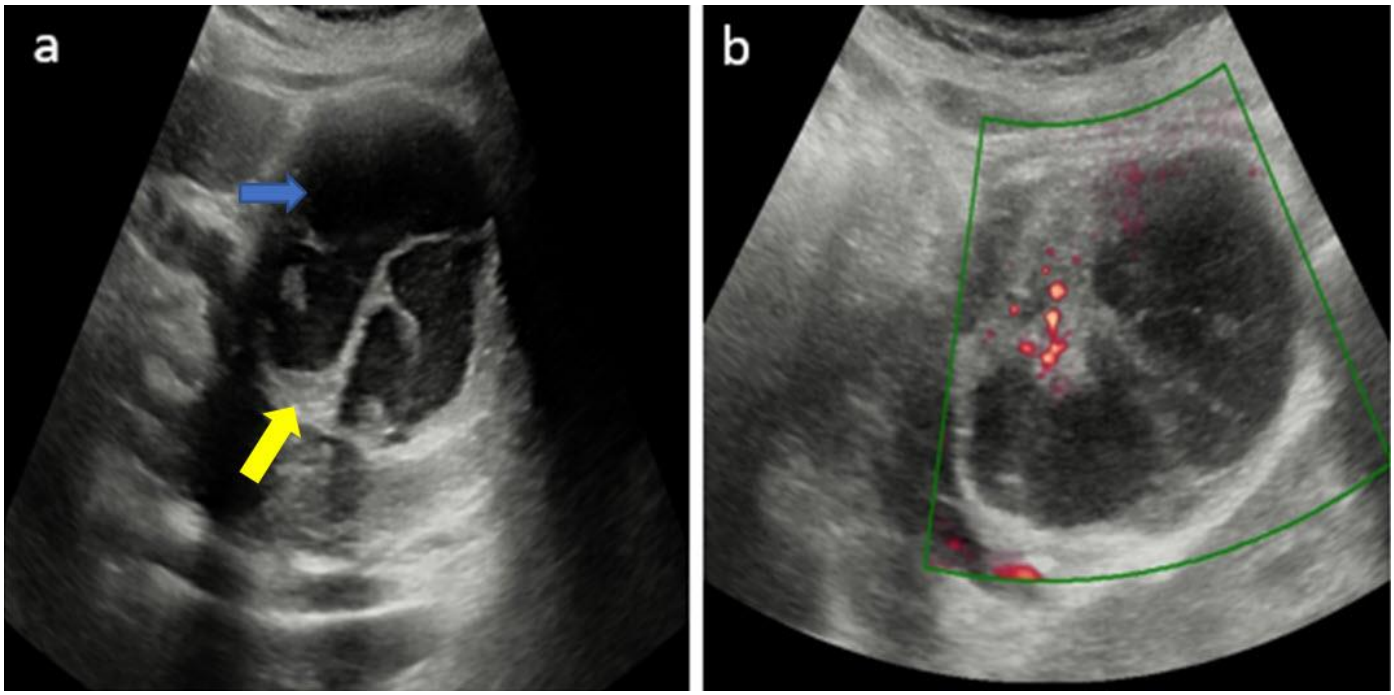

Figure 4: 19-year-old female with Solid Pseudopapillary Epithelial Neoplasm of the pancreas.

Findings - Gray scale ultrasound image showing solid cystic lesion with peripheral solid component (yellow arrow) and central cystic component (blue arrow) in image (4a). Color Doppler ultrasound image showing vascularity within the solid component in image (4b).

Technique: Gray scale ultrasound of the epigastric region of abdomen, using a 3-12 MHz convex array transducer (Samsung RS80A), in axial plane in image (4a). Color Doppler ultrasound of the epigastric region of abdomen, using a 3-12 MHz convex array transducer (Samsung RS80A), in axial plane in image (4b).

Etiology	Unknown
Incidence	0.13–2.7% percent of all pancreatic neoplasms
Gender ratio	M: F 5:1
Age predilection	Second and third decades of life. Mean age of occurrence is 23.9 years
Risk factors	No known
Treatment	**Localized tumor:** Complete surgical resection which involves a radical procedure (e.g., Whipple or distal/partial pancreatectomy) **Metastatic tumor:** Complete surgical resection of the tumor with chemotherapy
Prognosis	Good prognosis with a reported 5-year survival of 97%.
Findings on imaging	**Ultrasound:** Heterogenous mass with areas of internal hemorrhage and cystic degeneration with fluid-debris levels within and posterior acoustic enhancement **Computed tomography:** Heterogenous density mass with fluid density in the central region and soft tissue density in the peripheral region. Enhancement in the soft tissue component on contrast studies. **Magnetic Resonance Imaging:** Heterogenous signal intensity on T1 and T2 weighted images. Thick fibrous capsule on T2 weighted images seen as a rim of low signal intensity. On gadolinium-enhanced dynamic MR imaging, early heterogenous enhancement of the fibrous capsule with progressive fill-in

Table 1: Summary table of Solid Pseudopapillary Epithelial Neoplasm of the pancreas.

	Occurrence	Age	Gender	Imaging features
SPEN	3 % of the pancreatic neoplasms	2nd–4th decade ("daughter" lesion)	Females >males	**CT**: Heterogeneous solid and/or cystic with hyperattenuating areas from hemorrhage **MRI**: T1W and T2W: variable owing to internal necrosis and hemorrhage
Pseudocyst of pancreas	Secondary to history of pancreatitis.	May occur at any age	Males> females	**CT**: Cystic lesion with variable wall thickness; may have septae within **MRI**: T1W: low SI T2W: high SI; may have thick enhancing walls, septa, and/or nodules
Serous cystic neoplasm	16% of the pancreatic neoplasms	5th– 7th decade ("grandmother" lesion)	Females >males	**CT**: Multicystic, lobulated with enhancing septa **MRI**: T1W: low SI T2W: high SI Central scar
Mucinous cystic neoplasm	23% of pancreatic cystic lesions	4th–5th decade ("mother" lesion)	Almost exclusively in females	**CT**: Few cysts with occasional mural nodule and/or septa **MRI**: T1W: usually low SI T2W: high SI; may have thick enhancing walls, septa, and/or nodules
Intraductal papillary mucinous neoplasm	38% of pancreatic cystic lesions	6th–7th decade	Equally common in males and females	**CT**: BD: Cysts communicate with pancreatic duct MD: segmental or diffuse dilatation of pancreatic duct **MRI**: T1W: low SI T2W: high SI Enhancing nodules increase likelihood of malignancy

Table 2: Differential diagnosis table for Solid Pseudopapillary Epithelial Neoplasm of the pancreas.

ABBREVIATIONS

BD = branch duct, MD = main duct
CT = Computed Tomography
ERCP = Endoscopic Retrograde Cholangiopancreatography
EUS = Endoscopic ultrasound
FNAC = Fine Needle Aspiration Cytology
IPMN = Intraductal Papillary Mucinous Neoplasm
MCN = Mucinous Cystic Neoplasm
MRI = Magnetic Resonance Imaging
SI = signal intensity
SPEN = Solid Pseudopapillary Epithelial Neoplasm
T1W = T1-weighted MRI
T2W = T2-weighted MRI
U/L = units per liter

KEYWORDS

Solid pseudopapillary epithelial neoplasm; Frantz tumor; pancreatic tumors; cystic pancreatic neoplasm; computed tomography; pancreas

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