

## Case Report

# Solid and Papillary Epithelial Neoplasm (SPEN) of the Pancreas in a Pregnant Lady

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## ABSTRACT

State-of-the-art cross-sectional imaging has led to the realization that the prevalence of cystic neoplasms in the pancreas is much higher than initially estimated. This has also permitted the differentiation of most cystic neoplasms based on cyst size, shape, wall thickness, internal architecture, location and presence of calcifications. The

solid and papillary epithelial neoplasm (SPEN) of the pancreas is a rare low-grade cystic malignancy, seen mainly in adolescent and young adult females. We present the imaging findings of SPEN of the pancreas in a young pregnant lady that was accurately diagnosed preoperatively.

KEYWORDS: imaging, pancreas, SPEN

## INTRODUCTION

Solid and papillary epithelial neoplasm of the pancreas (SPEN) is a rare low-grade malignancy mainly found in adolescent and young women of Asian and far Eastern origin. Clinically, patients are asymptomatic or may present with non-specific signs and symptoms including nausea, vomiting and upper abdominal discomfort caused by the usually great size of the tumor. Surgical resection is curative in most instances<sup>[1,2,3]</sup>.

## CASE REPORT

A 34-yr-old Arab pregnant lady, a mother of six, presented with complaints of a dragging sensation in the upper abdomen. The gestational age of her fetus was 16 weeks at the time of examination. There was no history of previous biliary tract disease, pancreatitis, or trauma. She denied rearing animals at home. There were no complaints of arthralgia. Her laboratory tests including her blood and eosinophil count were normal.

Ultrasound of 8/2/2003 revealed a 14 x 11 cm well encapsulated complex, heterogeneous mass with cystic and solid components in the tail of the pancreas, displacing the spleen laterally and compressing the left kidney posteriorly without evidence of invasion. The cystic mass showed posterior enhancement (Fig. 1). The other organs, including the liver, spleen, kidneys, gallbladder and biliary tract were normal. Pelvic ultrasound revealed a single live fetus (of gestational age 16

weeks) in the uterine cavity. The ovaries and adnexae were normal. There was no free fluid.

Because of the high radiation dose delivered by computed tomography (CT) abdomen to the patient and in particular to the fetus, it was decided to proceed to magnetic resonance imaging (MRI) of the abdomen rather than CT scan. MRI revealed a well encapsulated predominantly cystic mass (as revealed by the low signal on T1- Fig. 2 and high signal on T2- Fig. 3) with peripheral mural or frond-like areas of intermediate signal intensity due to the solid component. The fat suppressed images emphasized the cystic nature of the mass (Fig. 4). Gross hemorrhage was not present, although a minimal hemorrhagic component could not be excluded. The fibrous capsule was seen as a hypointense rim on T1 and T2. Administration of gadolinium would have enhanced the evaluation of this tumor but was refused by the patient, even though its use is safe for the mother and the fetus. Because of the clinical presentation and the radiographic findings, the strong possibility of a SPEN of the pancreas was raised.

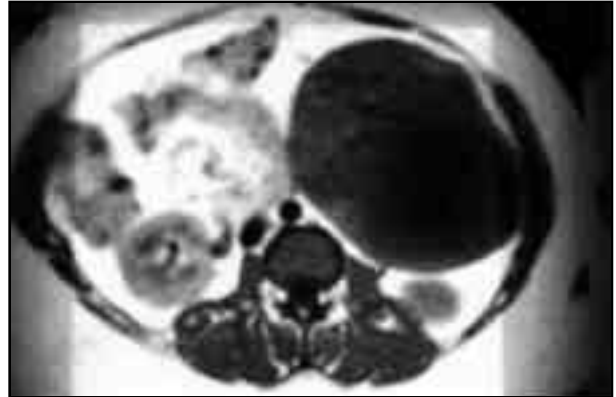
On ultrasound guided aspiration of the cyst, 40 ml watery blood stained fluid was withdrawn. Cytology from the solid area of the cyst and fluid showed few sheets and dispersed small cuboidal epithelial cells without nuclear atypia and with scanty cytoplasm, macrophages, eosinophils and neutrophils. Findings were suggestive of a benign epithelial neoplasm.

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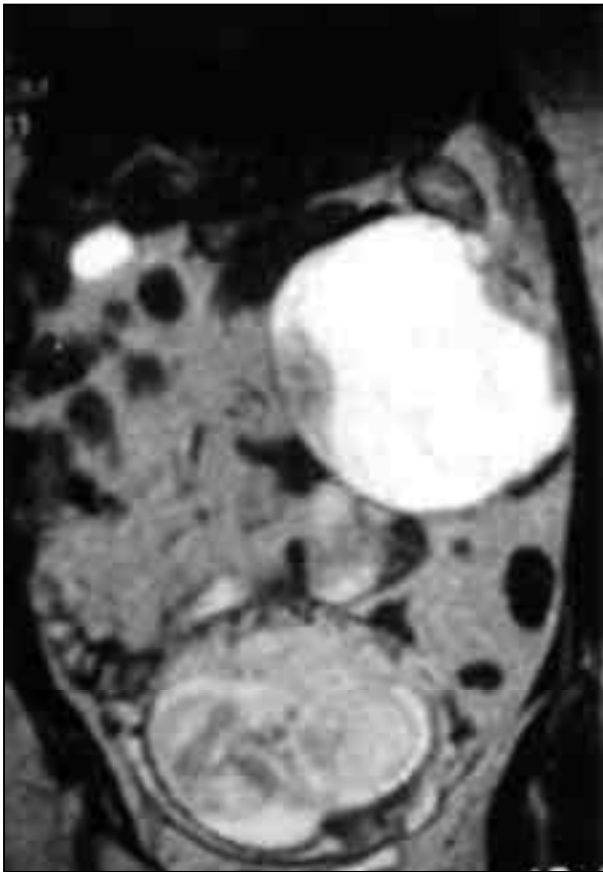
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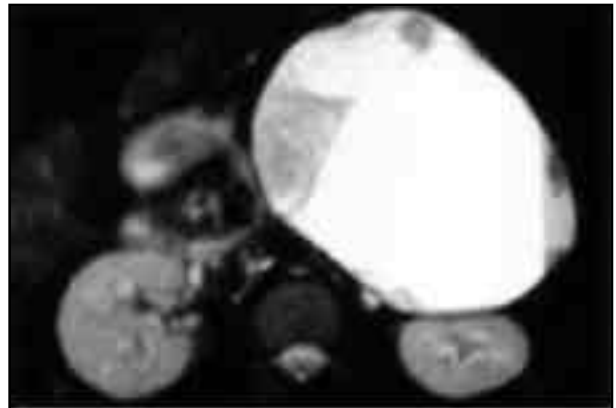
**Fig. 1:** Ultrasound of SPEN, tail of pancreas, showing the large, well-encapsulated, predominantly cystic tumor with a frond of solid tumor in the right lower corner



**Fig. 2:** Axial T1W MR (500/10) shows the homogenous low signal intensity of the fluid and low-intermediate signal of the wedge-shaped solid component



**Fig. 3:** Coronal T2W MR (4000/110) demonstrates the well-encapsulated cystic mass in the tail of the pancreas. Don't miss the in-house guest in the pelvis.



**Fig. 4:** Axial Fat Suppressed T2W MR (5181/90) showing the cystic component of the tumor as high signal intensity with the peripheral wedge shaped solid component as intermediate signal intensity.



**Fig. 5:** CT abdomen with oral and I.V. contrast showing a small post-operative collection in the tail of pancreas

On 12/7/2003, the patient underwent a cesarian section. The incision was extended upwards and the cyst was inspected. It was nearly 20 cm in diameter and arising from the tail of the pancreas. 1500 ml of hemorrhagic fluid was aspirated. The cyst was then excised in toto. The distal end of the pancreas was closed with TIAstapler and oversewn with vicryl 00. It was also covered with omentum. Hemovac drain was put in the lesser sac and the abdomen was closed. Miniheparin started preoperatively was continued postoperatively till the patient

became fully mobile. Fluid per Hemovac drain showed raised amylase for three days, and the patient was discharged on the 11th postoperative day.

Two weeks later, the patient complained of abdominal pain in the left upper quadrant. Ultrasound followed by CT abdomen showed a small collection in the region of the tail of the pancreas (Fig. 5) as well as a small infarct in the tip of the spleen (Fig. 6). Both were managed conservatively,

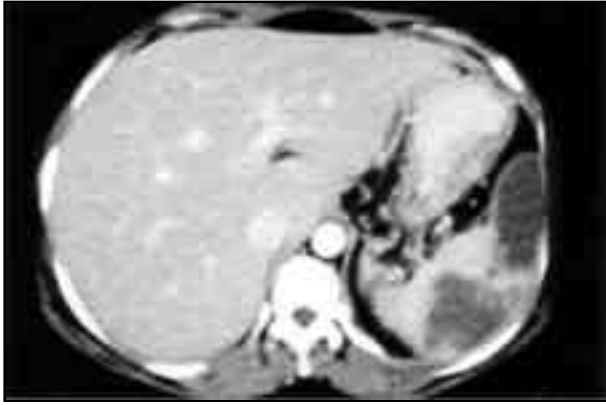


Fig. 6: CT abdomen with oral and I.V. contrast showing post-operative infarcts in the spleen

and the patient has since been well.

Pathologically, the gross specimen measured 20 x 16 x 11 cm. It weighed 1.450 kg. It had a thin fibrous capsule and a hemorrhagic, partly necrotic center. Microscopically, the tumor had solid, cystic, pseudo-papillary and trabecular pattern of growth. The solid component showed poorly supported vessels. Areas of myxoid change with thin blood filled channels were noted. The tumor cells had ovoid nuclei and granular eosinophilic cytoplasm. At one focus, invasion of the capsule was noted. Findings confirmed the diagnosis of SPEN.

## DISCUSSION

Cystic neoplasms of the pancreas are relatively uncommon, representing only 10-15% of pancreatic cysts and 1% of pancreatic cancers<sup>[4]</sup>. SPENs are rare pancreatic tumors with a reported incidence of 0.17% to 2.5% among exocrine pancreatic tumors<sup>[5]</sup>. Pre-operative diagnosis is important, since surgical resection is curative. They occur in young females with an average age of 24 years<sup>[3]</sup>. When found in older females, these tumors are associated with a higher grade of malignancy<sup>[6]</sup>.

SPENs are large tumors with an average diameter of 9 to 18 cm<sup>[6]</sup> and about 58% arise in the tail of the pancreas<sup>[1]</sup>. On imaging, these tumors can be solid<sup>[3]</sup>, but are usually cystic with a well formed capsule.

It is well characterized on ultrasound by areas of internal hemorrhage seen as increased internal echoes, along with cystic degeneration, fluid-debris levels and posterior enhancement due to the cystic component. Peripheral calcification may be present, but is better detected by CT<sup>[4,5]</sup>. Mural nodules are also occasionally seen by ultrasound.

On computed tomography, the SPEN has a solid periphery and central cystic degeneration with hemorrhage. The fluid contents hence show density greater than water. Peripheral calcification, when present, is best detected by this modality. A well-

defined hypodense capsule is seen, which shows enhancement with contrast. Tumor vascularity is moderate<sup>[2,4,6]</sup>.

MR has proved to be a valuable modality in the evaluation of SPEN. Because MR imaging is sensitive to fluid, it has great potential in the assessment of cystic neoplasms<sup>[6,7]</sup>. T1 and T2W images demonstrate a heterogeneous well-demarcated mass with areas of low and high signal intensity respectively. This appearance reflects the complex nature of the solid, cystic, hemorrhagic and necrotic components<sup>[1]</sup>. The cyst fluid improves the contrast resolution within the mass, delineating subtle irregularities of the cyst wall which aid in the differential diagnosis between benign and malignant neoplasms, and pseudocysts<sup>[7]</sup>. It also increases the specificity by demonstrating the hemorrhagic component on T1 weighting due to the paramagnetic component of methemoglobin<sup>[6,8]</sup>. The fibrous capsular rim is hypointense on T1 and T2. The capsule and the solid portion of the tumor may enhance with gadolinium. This mass can be seen to displace adjacent structures, as in this study, without invasion<sup>[1,6]</sup>.

Pathologically, SPEN is a large well-encapsulated mass. It has a thick fibrous capsule with mixed cystic, solid and pseudopapillary pattern. The solid areas consist of sheets of epithelial cells whereas the papillary areas consist of a fibrovascular core lined with cuboidal or columnar epithelial cells. Hemorrhage occurs due to the rupture of the poorly supported vascular network traversing the tumor<sup>[1,2,5]</sup>.

The differential diagnosis would include the gamut of cystic pancreatic neoplasms including the serous or microcystic adenomas, mucinous or macrocystadenomas, cystic non functioning islet cell tumors and intraductal papillary mucinous tumors<sup>[1,2,5]</sup>. The age of presentation, the site of the tumor within the pancreas (*i.e.*, head, body or tail) and the cross-sectional imaging characteristics of size of the tumor, presence or absence of calcification and septae and the presence of a capsule would help in the differentiation of these lesions. The main distinguishing features are as follows:

Serous or microcystic cystadenoma or glycogen rich cystadenoma presents in 60 year old females, usually in the region of the head of the pancreas. It has thin septae separating multiple cysts 1 mm to 2 cm in size which are distributed in a peripheral location within the tumor giving it a honeycomb or a lace like appearance. Central stellate scars with a sunburst pattern of calcification can be seen. MR imaging shows delayed enhancement of the scar on contrast enhanced images. These are usually benign tumors<sup>[2,8,9]</sup>.

Mucinous or macrocystic cystadenoma present in 40-50 year old females and are malignant or potentially malignant. These lesions have larger cysts (more than 2 cm in diameter) and are unilocular or multilocular. Thick septae, solid mural nodules and peripheral coarse calcifications are present<sup>[2,8,9]</sup>.

Intraductal papillary mucinous tumors are rare. Two-thirds of the patients are men, with a peak age of incidence at sixty. Most patients present with pancreatitis. The lesion is characterized by marked distension of the pancreatic duct with a large amount of mucus, leading to cyst formation. CT scan may reveal a cystic mass of the pancreas or a dilated pancreatic duct which may be similar to chronic pancreatitis<sup>[3]</sup>.

Cystic non-functioning islet cell tumors are small in size ranging from 4 mm to 2 cm and are very vascular. They also have a different enhancing pattern with intravenous contrast<sup>[2]</sup>.

Pseudocysts, though inflammatory in nature, deserve a mention in the list of differential diagnosis. They are the sequel of acute or chronic pancreatitis, trauma or pancreatic cancer. They are usually located within the pancreas but can present in the retroperitoneum, mediastinum or even the parenchyma of the liver, kidney or spleen. They are single, unilocular with internal echoes due to fluid or debris. They lack a solid component and are encapsulated by fibrous tissue<sup>[8, 9]</sup>.

The SPEN presents at a younger age than the above mentioned tumors. They generally lack internal septations and multiple loculations. The large size, the well-defined capsule, the heterogeneous mixed solid and cystic pattern and the hemorrhagic component seen as high signal

intensity on T1 weighted imaging serve to distinguish the SPEN from other cystic pancreatic neoplasms.

With the appropriate clinical setting, the imaging findings can be highly suggestive for the diagnosis of SPEN. This tumor should be primarily considered in a young female presenting with a large, well-defined cystic mass in the pancreas, with heterogeneous pattern. An accurate diagnosis is invaluable, since total surgical resection has an excellent prognosis.

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