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POLARISING FRANTZ TUMORS – BENIGN AND MALIGNANT SOLID PSEUDOPAPILLARY EPITHELIAL NEOPLASM OF PANCREAS - TWIN CASE REPORTS

Radio-Diagnosis

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ABSTRACT

Solid pseudo-papillary epithelial neoplasm (SPEN), also known Hamoudi tumors or Franz tumors, are rare pancreatic neoplasm which are almost always seen in young women predominately of non-Caucasian descent with only a small minority of cases diagnosed in men [1]. The first published description of an SPN was by Frantz in 1959[2].It is a rare tumor comprising of less than 3 percent of all pancreatic tumor. It is seen most often in the region of tail of pancreas. This tumor is mostly asymptomatic and usually detected when it reaches large size. These are tumors with low malignant potential and rarely vascular invasion and metastatic disease can be seen in aggressive cases. Here we report two cases, one each of benign SPEN and malignant SPEN with contrasting imaging findings and polar outcomes.

KEYWORDS

SPEN, Pancreatic tumor, Pseudo-papillary,β-catenin, Ki-67

INTRODUCTION:

This tumor is known by many aliases like solid cystic tumor, papillary cystic tumor and solid pseudo-papillary tumor of the pancreas. It is a rare tumor comprising of less than 3 percent of all pancreatic tumor. It is seen to occur mostly in young women. It is seen most often in the region of tail of pancreas.

This tumor is mostly asymptomatic and usually detected when it reaches large size. These are tumors with low malignant potential and rarely vascular invasion and metastatic disease can be seen in aggressive cases. On imaging, these are seen as well-defined encapsulated masses which have a combination of solid and cystic components.

MATERIALS AND METHODS:

This work has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) and with approval of Institutional Ethical Committee.

Verbal Consents were obtained from both the patients whose case reports are discussed here. Furthermore, all radiological images and data are anonymized by removing patient identifiers to protect patient identity.

Since the patient's age and gender are vital demographic parameters for the diagnosis of SPEN, they are retained in the study with the consent of patient.

Two Cases of SPEN with varied presentation, imaging findings and clinical course are discussed below:

CASE 1:

A 20 year old female presented with insidious onset of progressive abdominal discomfort and feeling of lump in the abdomen with gradually progressing, episodic, epigastric pain. No history of vomiting, jaundice. Patient complained of loss of appetite with loss of weight, however there were no other gastrointestinal symptoms.

On clinical examination, a mass was palpable in the epigastric region. Initially abdominal ultrasonography was done which showed a large solid-cyst complex mass lesion in epigastrium with few septations and internal debris in the region of tail of pancreas and subsequently contrast enhanced CT was done for the patient.

RADIOLOGICAL IMAGING:

FINDINGS:

On cross sectional imaging of abdomen by contrast enhanced CT – A large well defined heterogenous solid - cystic lesion with thin walls was seen in the region of tail of the pancreas measuring ~8.8 x 9.2 x 10.5 cm. On contrast administration the lesion was showing heterogenous enhancement. Enhancing solid components were predominantly seen in the periphery of the lesion with multiple internal non-enhancing areas. No internal calcifications, hyperdense content, hemorrhagic components were noted. No definite pancreatic ductal communication could be demonstrated.

Posteriorly, the lesion was seen to compress the splenic vein, artery and the anterior cortex of left kidney. Anteriorly, it was seen to abut the greater curvature of the body of the stomach and antero-inferiorly it was seen abutting the anterior abdominal wall. It was seen to displace the jejunal loops medially. Fat planes with the adjacent organs maintained with no evidence of invasion of adjacent viscera No noted. Main pancreatic duct was not dilated. No intraductal or parenchymal calcifications were seen. No other similar lesions were seen elsewhere in pancreas. No significant adenopathy and fluid collection were noted.

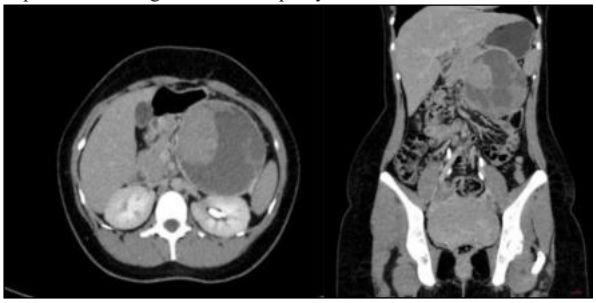


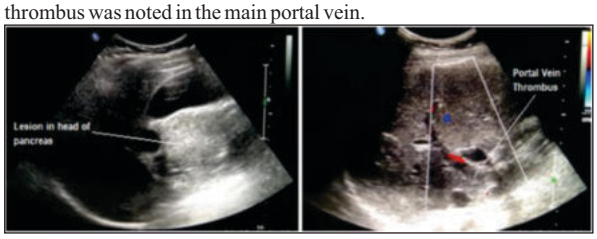
Figure 1 - CECT abdomen axial (1A) and coronal section shows (1B) a well-defined solid-cystic lesion in the tail region of pancreas with enhancing solid component (White arrow) noted in the periphery.

CASE 2:

A 12 year old female presented with complain of abdominal pain and nausea for 2 months. Patient had repeated episodes of vomiting during the period. Patient was afebrile. No history of abdominal distension,

jaundice, gastrointestinal bleed, fever, shortness of breath, decreased urine output, altered sensorium were present. Patient complained of loss of appetite with loss of weight. On clinical examination, a mass was palpable in the epigastric region. There was no icterus pallor, or tenderness on palpation.

Ultrasound was done for the patient, which showed a well defined heterogenous hyperechoic lesion in the head of pancreas. Few eccentric acoustic shadows likely due to calcifications were noted. No significant internal vascularity was noted. A mixed echogenic thrombus was noted in the main portal vein.



Ultrasonography of abdomen- Figure 2A- Well defined hyperechoic lesion in the head of pancreas. Figure 2B - Echogenic thrombus in the main portal vein.

Subsequently contrast enhanced CT scan revealed–

There was evidence of a spherical mildly enhancing mass lesion in the head of pancreas. Peripheral curvilinear calcifications were seen along the surface. Supero-medially the mass was seen extending upto the porta hepatis and mildly compressing the adjacent 2nd part of duodenum. The mass was seen partially encasing the common hepatic artery near porta hepatis and invading the confluence of portal vein and main portal vein. A partial bland thrombus was also seen within the superior mesenteric vein. The lesion was seen to compress the common bile duct displacing it posteriorly. Few non enhancing necrotic areas also were seen within the mass. Few portal collaterals were seen at peripancreatic region. Main pancreatic duct was not dilated.The superior mesenteric artery, IVC was uninvolved. No regional adenopathy was noted.

Few well defined round hypodense lesions were seen in liver largest of those was seen in segment III. The lesion showed significant peripheral enhancement in arterial phase. Other smaller lesions also revealed mild peripheral enhancement.

CECT Abdomen -

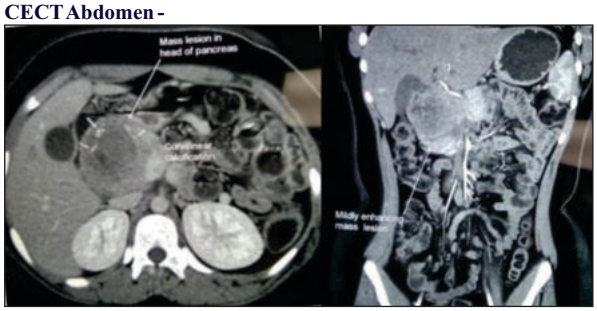


Fig. 3A. Enhancing mass lesion in Axial Section.

Fig. 3B. Coronal Section

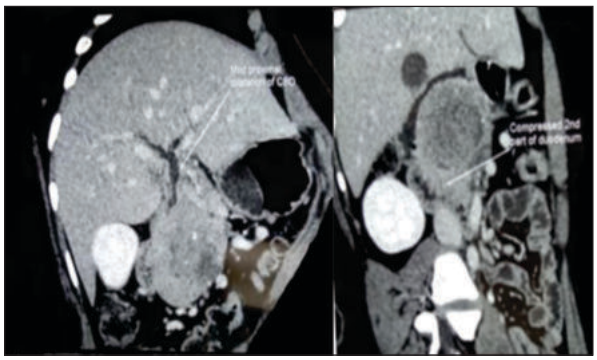


Fig. 3C. Coronal Section Showing CBD.

Fig. 3D. Sagittal Section Showing Compression Of Duodenum.

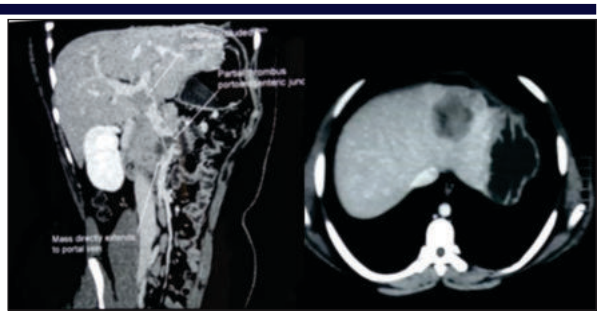


Figure 3E- Coronal Section showing portal vein invasion

Fig. 3F- Metastatic lesion in liver

DIAGNOSIS AND MANAGEMENT:

CASE 1:

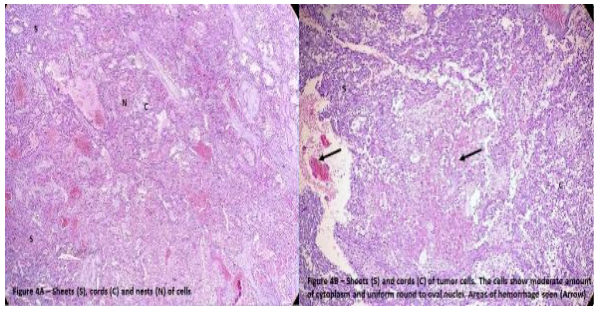
In our first case, considering the age and sex of the patient, radiological diagnosis of benign SPEN was given and the patient underwent complete excision of the mass along with splenectomy and omentectomy and the specimens were sent for HPE. HPE showed well circumscribed solid and cystic neoplasm composed of sheets cords and nests of eosinophilic cells with moderate eosinophilic to vacuolated cytoplasm and uniform round to oval nuclei with finely dispersed chromatin with inconspicuous nucleoli and sparse nuclear grooving. Areas with fragile capillaries, vascular congestion were seen. No mitotic figures were noted. Areas of hemorrhage were seen. Resected pancreatic margins, spleen and omentum were free from infiltration.

Post operative recovery was uneventful and patient was advised to follow up in medical gastroenterology and endocrinology departments. Thus, surgical resection proved to be successful in this case with no evidence of recurrence in 2 yr interval imaging follow-up.

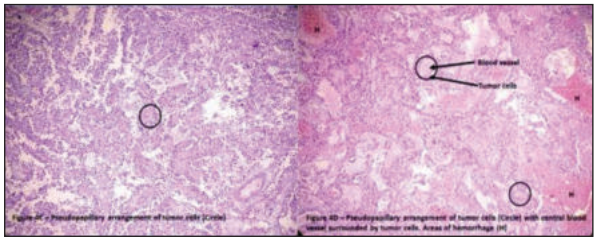
CASE 2:

In our second case, Based on the USG diagnosis of heterogenous cystic lesion, possibility of hydatid cyst or a pancreatic inflammatory/ infective pseudocyst were ruled out by a negative echinococcal serology. UGI endoscopy revealed eccentric bulge in the region of second part of duodenum. Then taking into consideration of the CT picture of local aggressive nature of the tumor, vascular invasion and hepatic metastasis, primary radiological diagnosis of pancreatic adenocarcinoma was given. However considering the atypical enhancement characteristics, heterogenous composition of tumor and age of the imaging differential diagnosis of malignant SPEN and neuroendocrine tumor of pancreas were also given. Patient was advised serum CA 19-9,Serum LDH, Serum AFP, neuroendocrine hormone assays and Image guided biopsy and HPE. CA 19-19 levels were normal. USG guided percutaneous core biopsy of pancreatic mass and hepatic metastasis was done and sent for histopathological exam was performed. IHC markers like Ki-67, β-catenin , synaptophysin, chromogrannin A and pancytokeratin were done. Serum CA 19-19, AFP was normal. Serum LDH was raised. HPE showed features consistent with malignant SPEN with positive IHC markers like Ki-67 and β-catenin which confirmed our radiological diagnosis of malignant SPEN with portal vein and hepatic metastasis. Considering the inoperable nature of the disease, alternative chemoradiation regimens were suggested. Patient was started on trial chemotherapy regimen of pancreatic cystadenocarcinoma, but patient ultimately succumbed to the sinister nature of the disease.

Histopathogical Findings:



Figures 4A & 4B - Sheets, cords and nests of uniform small fairly round cells with moderate eosinophilic to vacuolated cytoplasm and uniform round to oval nuclei.



Figures 4C, 4D - Areas of necrosis with pseudopapillary arrangement of the tumor cells around the blood vessels and areas of hemorrhage.

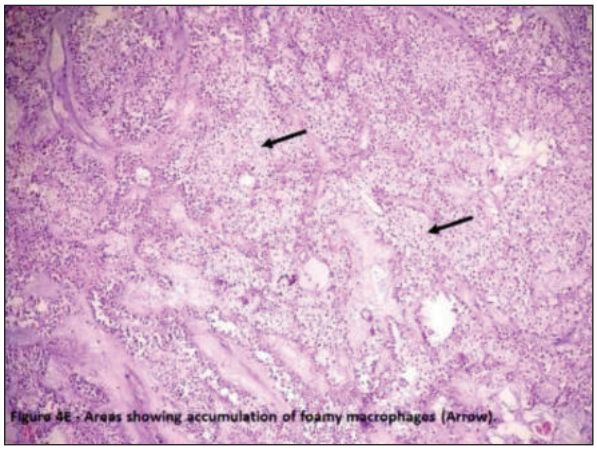


Figure 4E- Areas Of Foamy Macrophages Accumulation.

DISCUSSION:

Solid pseudo-papillary epithelial neoplasm is an uncommon pancreatic tumor seen chiefly in young females. Lesions can be large at time of diagnosis (median size ~8 cm)[3]. Patients are generally asymptomatic with abdominal pain being the most common symptom. Other symptoms include nausea, vomiting, weight loss and jaundice. Gastrointestinal bleeding is seen rarely. Abdominal mass is palpable especially in children. They may occasionally present with a gradually enlarging abdominal mass or vague abdominal pain. These tumors frequently contain varying amounts of necrosis, hemorrhage, and cystic changes. WHO classification defines SPEN as low grade malignancies; however lesions may demonstrate signs of local aggressive spread and malignant potential in the form of perineural invasion or microscopic vascular invasion [8]. Gross pathological features include areas of central hemorrhage and necrosis surrounded by solid and pseudo-papillary structure. Thick, hypervascular capsule surrounding mixture of solid and cystic areas with hemorrhage and necrosis is seen. Microscopically lesion shows appears as homogenous, small epitheloid cells as single cells or aggregates, small sheets or papillary structures. Complications include hemorrhage, biliary obstruction. There is a predilection to involve the pancreatic tail.

On ultrasound it appears as solid mass with cystic components with fluid-debris levels, posterior acoustic enhancement and very little internal color flow /vascularity on doppler. On CT, it appears as well-defined, heterogeneous, encapsulated mass with thick, enhancing capsule having frequent peripheral or central calcification [4].Frequent peripheral or central calcification is seen. Most often the tumor is solid but can have variable cystic components and intra-tumoral hemorrhage. Usually no biliary or pancreatic ductal obstruction is seen. Gross vascular invasion or occlusion on imaging is rare. Metastatic disease is very uncommon, but if present it often it metastasizes to liver and locoregional lymph nodes. On MRI, it appears as a large, well-demarcated mass with central areas of low and high T1 signal intensity (hemorrhage). Presence of internal hemorrhages a characteristic feature, and may result in fluid-fluid or hematocrit levels. On T1WI post contrast, it appears as solid-cystic mass with minimal enhancement. Capsule appears as rim of low T2 signal intensity and enhances on post-gadolinium imaging.

Surgical intervention is the mainstay of treatment which includes enucleation, Whipple's resection or subtotal pancreatectomy depending upon the size and extent of the tumor [5].Recurrences are uncommon but can occur in up to 15% of cases which show aggressive behavior[6].The overall median five-year survival rate is >95%[7].

CONCLUSION:

SPEN finds a place in the imaging differential diagnoses of pancreatic neoplasms especially in young adolescent females. These are benign lesions in majority of cases with a classical clinicoradiological scenario - A large well-encapsulated mass that demonstrates calcification and regions of hemorrhagic degeneration seen in a young woman. Classic imaging characteristics include large size, mixed solid and cystic nature, encapsulated appearance and presence of hemorrhage. Surgery is curative in the vast majority of cases.

However in around 15% of the cases there may be overt malignant changes and metastasis at the time of diagnosis. Owing to the rarity and overlapping imaging features, malignant SPENs are almost always misdiagnosed as cystadenocarcinomas and other neuroendocrine malignancies on imaging. More awareness is required about the possible malignant nature of the lesion and malignant SPENs though rare should be considered as imaging differential diagnosis and confirmed with prompt HPE with appropriate IHC markers, to allow for early curative resection if possible.

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ABBREVIATIONS –

USG - Ultrasonography, SPEN- Solid Peudopapillary Epithelial Neoplasm. CT – Computed tomography, CECT – Contrast enhanced CT, MRI – Magnetic resonance imaging, HPE- Histopathological Examination, IHC- Immunohistochemistry.

DECLARATIONS:

Competing interests - The authors declare that they have no competing interests.
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All authors have read and approved the manuscript.

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