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Silent Presentation of a Solid Pseudopapillary Neoplasm of the Pancreas

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Female, 14
Final Diagnosis: Solid pseudopapillary neoplasm of the pancreas
Symptoms: None
Medication: —
Clinical Procedure: —
Specialty: Radiology

Objective: Rare disease

Background: Solid pseudopapillary neoplasm (SPN) is a rare tumor frequently found in the head or tail of the pancreas. It mainly presents in young women between the 2nd and 3rd decades of life. A predilection for African Americans and Asians has been observed and is rarely reported in children. Most patients are symptomatic, with abdominal pain as the most common presenting symptom. Clinical laboratory test results are usually normal and pancreatic markers are not typically elevated. Metastatic disease is very uncommon, but most often metastasizes to the liver and regional lymph nodes. Prognosis is usually excellent after surgical resection.

Case Report: We present the case of a 14-year-old Hispanic female who presented to the emergency department after a high-speed motor vehicle accident. She suffered multiple body traumas. Specifically, the patient referred severe epigastric pain. No significant past medical or surgical history was obtained. Laboratory workup was non-contributory. Further evaluation with abdomen and pelvis contrast-enhanced computed tomography and magnetic resonance imaging revealed a pancreatic tail mass. Distal pancreatectomy followed. Pathologic diagnosis of SPN was established.

Conclusions: SPN is a rare exocrine tumor with excellent prognosis following resection. Imaging findings are suggestive, but a pathology evaluation is necessary to make the final diagnosis. Differential diagnosis includes entities such as mucinous cystic pancreatic tumor, pancreatic ductal carcinoma, and pancreatic serous cystadenoma. Radiologists play a vital role in the diagnosis, since many times, as in our case, it presents as an incidental finding. A small percentage of SPN neoplasms are associated with metastasis or local recurrence. Therefore, the aim of our case presentation is to review key imaging findings to guide early management and surgical planning.

MeSH Keywords: Abdominal Pain • Multidetector Computed Tomography • Pancreatectomy • Pancreatic Neoplasms

Full-text PDF: http://www.amjcaserep.com/abstract/index/idArt/902867

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Background

SPN is a rare tumor (<3% of all pancreatic tumors) usually found in the head or tail of the pancreas [1]. It mainly presents in young women between the 2nd and 3rd decades of life. A predilection for African Americans and Asians has been observed and it is rarely reported in children [1,2]. Most patients are symptomatic, with abdominal pain as the most common presenting symptom. Other patients may be asymptomatic and present with a gradually enlarging palpable abdominal mass. Clinical laboratory test results are usually normal and pancreatic markers are not typically elevated. Metastatic disease is very uncommon (<10%), but it most often metastasizes to liver and regional lymph nodes. Treatment is with surgery. A laparoscopic distal pancreatectomy may be performed. Prognosis is usually excellent after surgical resection [3].

Case Report

We present the case of a 14-year-old Hispanic female who presented to the Emergency Department after a high-speed motor vehicle accident. The patient suffered multiple body traumas. Specifically, the patient referred severe epigastric pain at presentation. No significant past medical or surgical history was recorded. Laboratory workup was non-contributory. Evaluation with abdomen and pelvis contrast-enhanced computed tomography (CT) followed. CT images showed a hypodense, heterogeneously enhancing, partially exophytic lesion within the tail of the pancreas, measuring 3.3×2.9×3 cm (Figure 1). The lesion demonstrated progressive contrast enhancement of the capsule and few internal septations. Regions of hemorrhage were also identified within the lesion. There was no association with the splenic vein or artery. No peri-pancreatic fluid collection or evidence of inflammation was identified.

Magnetic resonance imaging (MRI) with emphasis to the pancreas was performed for further characterization. A well-defined pancreatic tail lesion measuring 2.3×3×3 cm was identified (Figure 2). A peripheral enhancing soft tissue component and few enhancing septations were identified within the lesion. In the 2-D and 3-D magnetic resonance images (MRCP), the pancreatic duct was identified with a normal course and no association with the lesion. No evidence of abdominal metastatic disease was identified.

Laparoscopic distal pancreatectomy was performed. The resected, encapsulated mass showed areas of hemorrhage and necrosis, surrounded by tissues with solid and papillary projections. The rest of the pancreas was unremarkable (Figure 3). Upon microscopic examination, the lesion showed areas of degeneration forming white clefts, conferring the tumor its pseudopapillary appearance (Figure 4). A pathologic diagnosis of SPN was established.

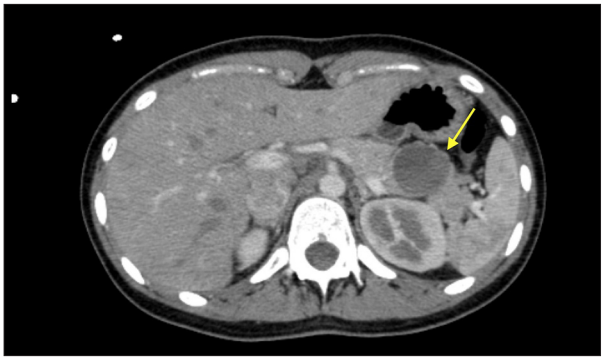

Figure 1. Axial contrast-enhanced CT image: Hypodense, heterogeneously enhancing, partially exophytic lesion within the tail of the pancreas, measuring 3.3×2.9×3 cm (arrow). The lesion is not associated with the splenic vein or artery. No peri-pancreatic fluid collection or evidence of inflammation.

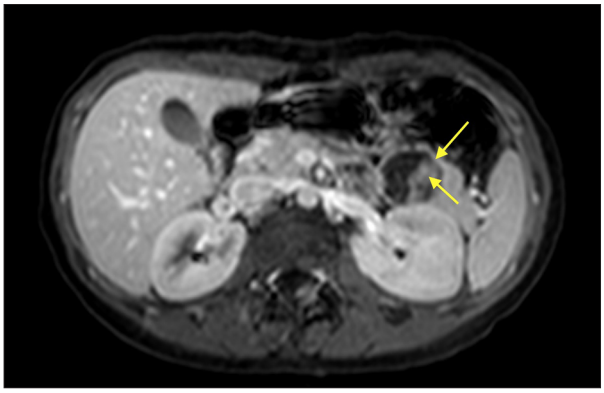

Figure 2. Axial contrast-enhanced arterial phase fat-saturated T1-weighted MR image: Well-defined pancreatic tail lesion with enhancing soft tissue component, as well as few enhancing septations (arrows). No connection with the pancreatic duct was seen in the MRCP images (not shown). No MRI evidence of malignant transformation was found, and there was no evidence of abdominal metastatic disease.

Discussion

SPN is a rare tumor (<3% of all pancreatic tumors) frequently found in the head or tail of the pancreas [1]. It mainly presents in young women between the 2nd and 3rd decades of life. A predilection for African Americans and Asians has been observed and it is rarely reported in children [1,2]. Most patients are symptomatic, with abdominal pain as the most common presenting symptom. Other patients may be asymptomatic and present with a gradually enlarging palpable abdominal mass. Clinical laboratory tests are usually normal and pancreatic markers are not typically elevated.

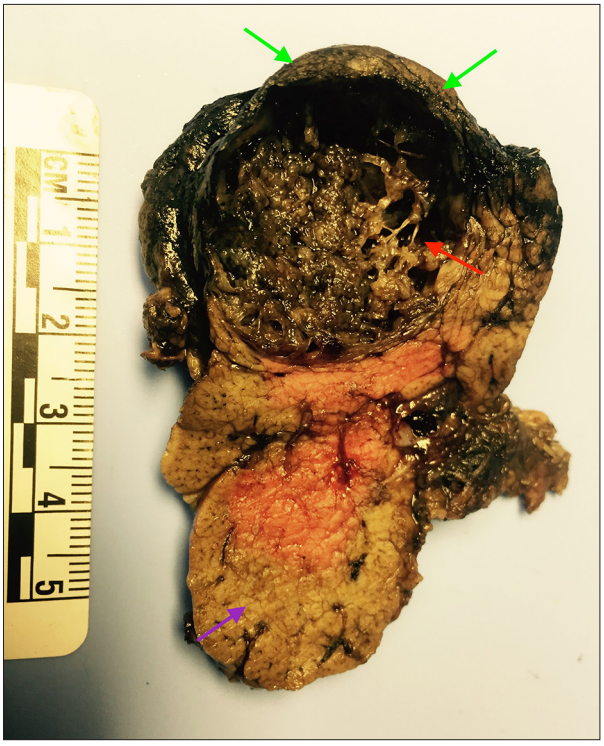

Figure 3. Distal pancreatectomy gross specimen: The resected mass shows areas of hemorrhage and necrosis, surrounded by tissues with solid and papillary projections (red arrow). The capsule varies in thickness (green arrows). The rest of the pancreas is unremarkable (violet arrow).

At gross examination, the mass is usually well circumscribed and encapsulated with varying areas of hemorrhage, necrosis, and cystic papillary projections.

Histologically, it shows fibrous septations and areas of degeneration forming white clefts; giving the tumor its pseudopapillary appearance. Peripheral or central calcifications may be seen in solid areas [1].

CT demonstrates a well-defined, encapsulated mass with a thick, enhancing capsule. Lesions are most often solid, with varying cystic components and intra-tumoral hemorrhage. They are usually quite large at presentation, with a mean size of more than 5 cm [1]. Following contrast administration, enhancing solid components are typically identified at the periphery, whereas cystic areas are more centrally located.

MR imaging demonstrates a well-demarcated mass with central areas of low and high T1 signal intensity, due to hemorrhagic products. Presence of internal hemorrhage is a characteristic feature and may result in fluid-fluid levels. The surrounding capsule typically appears as a rim of low T2 signal intensity. Solid components and capsule enhancement can be identified

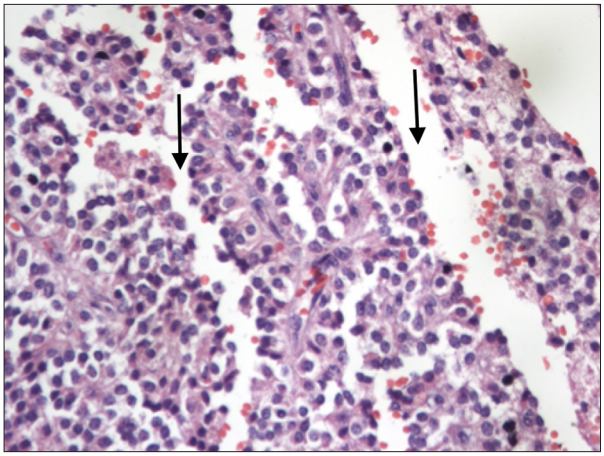

Figure 4. (400× H&E) High-power magnification shows areas of degeneration forming white clefts (arrows).

in post-gadolinium images. Additional 2-D and 3-D MRCP sequences can be performed to confirm the absence of biliary or pancreatic ductal obstruction.

Ultrasound findings may show fluid-debris levels within the mass with posterior acoustic enhancement. Scant internal vascularity is usually documented upon color flow interrogation.

Differential diagnosis for SPN of the pancreas includes entities such as mucinous cystic pancreatic tumor, pancreatic ductal carcinoma, and pancreatic serous cystadenoma [4].

Mucinous cystic pancreatic tumors are most commonly encountered in middle-aged to elderly women. Cystic lesions are found in the body or tail of the pancreas, consisting of a few cystic locules with thick walls.

Pancreatic ductal carcinoma most often occurs in older adults. It presents as a hypodense, poorly marginated, unencapsulated mass, resulting in pancreatic ductal dilatation and atrophy.

It has the tendency to obstruct the pancreatic and/or the bile ducts.

Pancreatic serous cystadenoma is usually seen in older women. It classically demonstrates a microcystic appearance with multiple small internal cystic components. Typically, no biliary or pancreatic ductal obstruction is observed.

MR imaging characteristics can also help differentiate SPN from other entities, such as neuroendocrine tumors. SPN cystic areas of high signal intensity on T1-weighted images and low signal intensity on T2-weighted images represent hemorrhagic products [5]. On the other hand, neuroendocrine tumors cystic components have corresponding increased T1 and T2 signal intensity.

Complete surgical resection is first-line treatment and is considered curative in most patients with SPN. Complications may include hemorrhage and biliary obstruction [3,6].

Prognosis is excellent after surgical resection. Metastatic disease is very uncommon, appearing only in <10% of cases. If metastasis does occur, it most often will arise in the liver and regional lymph nodes. Recurrence is extremely infrequent, with a mortality rate of less than 2% [5].

Conclusions

Solid pseudopapillary neoplasm of the pancreas is a rare exocrine tumor with excellent prognosis following resection. Imaging findings are suggestive but a pathology evaluation is necessary to make the final diagnosis. Differential diagnosis includes entities such as mucinous cystic pancreatic tumor, pancreatic ductal carcinoma, and pancreatic serous cystadenoma. Radiologists play a vital role in the diagnosis, since many times it presents as an incidental finding. The aim of our case presentation is to review imaging findings to guide appropriate treatment and surgical planning.

Conflict of interest

None declared.

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