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Case Report

Solid pseudopapillary tumor of pancreas: A report of two cases and literature review ☆,☆☆

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ABSTRACT

Solid pseudopapillary neoplasm of the pancreas is rarely encountered in clinical practice. It is a tumor with a good prognosis and overall curative rates. It primarily affects young females in their twenties. It has characteristic imaging appearances, but a definite diagnosis requires histopathological examination. The treatment goal of solid pseudopapillary neoplasm is almost always curative and aims for complete resection of the mass. Here, we present 2 cases of this rare neoplasm. The first case was managed by laparoscopic distal pancreatectomy, while the second underwent a Whipple procedure for pancreatic head involvement.

Introduction

Solid pseudopapillary neoplasm (SPN) is a very rare type of pancreatic neoplasm with low malignant potential and an overall good prognosis [4]. However, some cases may exhibit local aggressive growth, while others may metastasize early and involve the liver, lung, and skin [13]. The pathogenesis of this neoplasm remains unclear [8]. SPN tends to predominantly affect young females, with a mean age of 28 [13].

The clinical presentation of the disease varies widely, with abdominal pain or discomfort being the most common presenting symptom [16]. Additionally, in some patients, this neo-

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plasm can be incidentally detected during imaging studies or routine physical examination [9]. The treatment goal of SPN is curative through complete surgical excision in most cases [14]. In this paper, we present 2 cases of this rare clinical entity. The first case involves a 22-year-old female who underwent laparoscopic distal pancreatectomy for an incidentally found SPN during investigations for back pain in the neurosurgery clinic. The second case describes a 14-year-old female who underwent a Whipple procedure for a pancreatic head SPN detected during abdominal ultrasound in the gynecology clinic.

Case report 1

A 22-year-old female, smoker, with no significant medical history, initially presented to the neurosurgical clinic with complaints of back pain. However, she was later referred after the incidental detection of an abdominal mass. The patient reported experiencing severe sharp pain that intensified with movement over the past 4 months. Subsequently, she developed dull epigastric pain accompanied by nausea, unrelated to food intake. The patient denied experiencing vomiting, changes in bowel habits, or weight loss. Upon further inquiry, it was revealed that her sister had passed away from uterine cancer at the age of 23.

Upon examination, no abdominal masses were detected, and her examination was otherwise unremarkable. Her laboratory results, including tumor markers, were reported as within normal limits.

The patient underwent an MRI scan, revealing an oval-shaped lesion measuring 4.8 by 3.7 cm on the distal part of the pancreatic "tail". Subsequently, a contrasted abdominal computerized tomography (CT) scan was ordered, which demonstrated a 5 by 4 cm mass on the anterior aspect of the body and tail of the pancreas (see Fig. 1). The mass appeared iso-dense on the delayed phase, with a central area showing breakdown suggestive of a "cystic component rather than necrosis". The liver, spleen, and lymph nodes appeared uninvolved on the CT scan. The differential diagnosis included pancreatic adenocarcinoma and solid pseudopapillary neoplasm (SPN) of the pancreas.

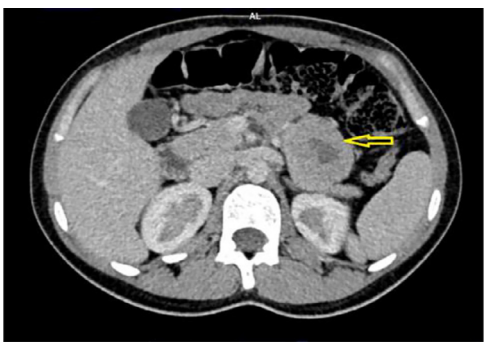

Fig. 1 – A 5 x 4 cm solid pseudopapillary tumor (SPT), (yellow arrow) protruding from anterior aspect of the pancreatic body and tail, appearing hypodense to the pancreatic parenchyma on venous phase of contrast, also it shows central area of cystic component.

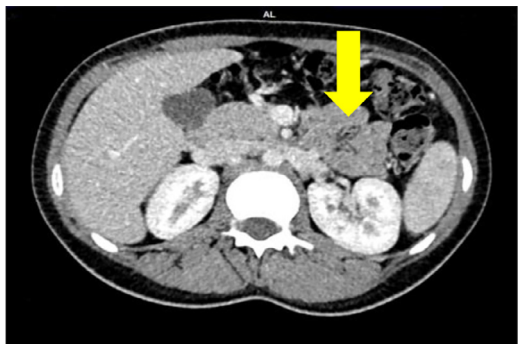

Fig. 2 – Three months post partial pancreatectomy, there is a thin walled localized fluid collection seen at the site of removed pancreatic tail, yellow arrow (mostly residual pseudocyst or walled post operative collection), with no evidence of tumoral residue or recurrence.

Laparoscopic distal pancreatectomy was performed without complications after unsuccessful attempts to separate the mass from the capsule for enucleation. The spleen was preserved during the surgery. Microscopic examination revealed a well-demarcated proliferation of neoplastic cells arranged in a solid and pseudopapillary pattern. Immunostaining showed positivity for Beta-catenin and CD10 in the tumor cells.

The tumor was staged as pT3N0 according to the pTNM AJCC 8th edition. However, the M staging could not be determined from the submitted specimen. The final report confirmed a solid-pseudopapillary neoplasm of the pancreas abutting the anterior, posterior, and inferior surfaces. Three months post optratively, Figure 2 showed that there was a thin walled localized fluid collection seen at the site of removed pancreatic tail, with no evidence of tumoral residue or recurrence.

Case report 2

A 14-year-old female patient, with no significant past medical or surgical history, presented with complaints of abdominal pain and distention lasting for 1 year. She was referred to the surgery clinic by her gynecologist after an incidental abdominal mass was detected on abdominal ultrasound. The pain was localized in the epigastric area and was described as mild and dull in nature. It persisted for months without relief or exacerbation by specific maneuvers, medications, or food intake. The pain did not significantly affect the patient's daily activities. She denied experiencing nausea, vomiting, weight loss, jaundice, upper gastrointestinal bleeding, or back pain. On physical examination, her vital signs were within normal limits, and her abdomen was soft and lax, with no tenderness to palpation. No abdominal masses were palpable, and normal bowel sounds were auscultated.

The patient underwent an MRI scan, which revealed a large, rounded, well-defined mass with heterogeneous signal intensity on both T1 and T2 sequences. The mass exhibited a few small cystic areas and one hemorrhagic focus, originating from the pancreatic head and exerting significant mass effect on the duodenum, common bile duct, and gallbladder. It measured 9 cm in maximum diameter. The remainder of the pancreas and pancreatic duct were not involved by the mass. No upper abdominal lymphadenopathy or ascites were observed. A presumptive diagnosis of a pseudo-papillary tumor of the pancreas was made following an endoscopic guided biopsy of the mass.

The patient underwent pancreaticoduodenectomy (Whipple procedure), which was performed successfully without complications. The recovery period was uneventful. Postoperatively, the patient was monitored for 24 hours in the surgical ICU before being transferred back to the general floor. She was discharged on postoperative day 7.

The histopathological examination of the surgical specimen revealed a cellular tumor comprised of sheets of small, uniform polygonal cells with occasional grooves, interspersed with delicate vasculature. These cells were arranged in a pseudopapillary pattern and exhibited positive staining for B-catenin, CD10, PR, and CD56 immunostains. Additionally, focal areas of necrosis were observed, although rare mitotic figures were present. Lymph node examination yielded negative results for malignancy. The tumor was staged as pT3N0 according to the pTNM AJCC 8th edition. However, the M staging could not be determined from the submitted specimen.

The patient was followed up in the outpatient clinic over a year after the surgery. She underwent a follow-up abdominal MRI, which revealed suspicious masses at the site of anastomosis. However, no liver or bone metastases were detected.

Figs. 3 and 4 showed A well-defined rounded pancreatic head mass (yellow arrow) measuring about 9 cm in maximum diameter with heterogenous appearance due to its cystic and solid components (1 hemorrhagic component is also demonstrated, yellow arrow head). This lesion exerts significant mass effect on the liver and gallbladder.

Figs. 5A–C showed the pattern of enhancement on the arterial, portal venous and delayed phases. The mass showed minimal heterogenous enhancement across the 3 phases. Figure 6 showes no mass at site of asastomosis, and no local or distant metastases one year post Whipple procedure.

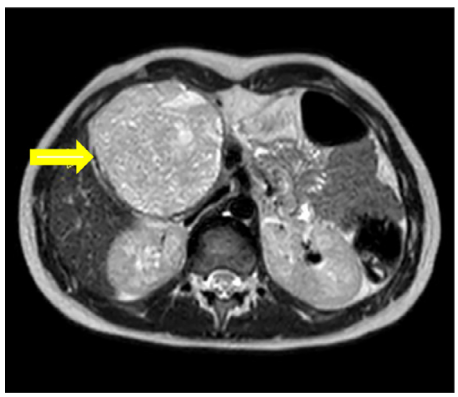

Fig. 3 – T2 MRI, axial section.

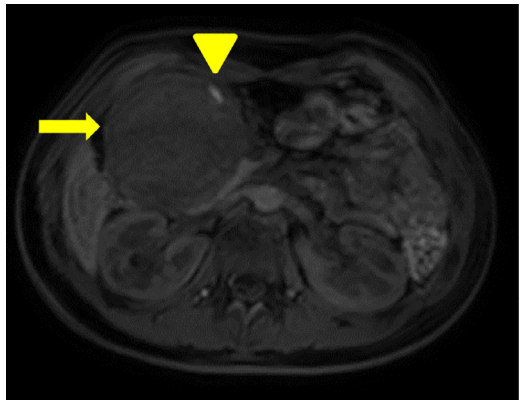

Fig. 4 – T1 MRI, axial section.

Discussion

Solid pseudopapillary tumors of the pancreas are rare exocrine pancreatic tumors, accounting for approximately 1%-2% of pancreatic neoplasms [6]. Various names were used to describe the tumor until 1996 when the World Health Organization (WHO) adopted the term “solid pseudopapillary tumor” for pancreatic lesions [15].

Solid pseudopapillary tumor (SPT) of the pancreas is a low-grade malignant neoplasm that predominantly affects young Asian and African American females aged between 20 and 30 years old. However, males can also be affected, and the disease tends to be more aggressive in this population [6]. The tumor commonly presents with symptoms such as abdominal pain, a slowly growing mass in the upper abdomen, or incidentally on imaging performed for other reasons. Less common symptoms may include nausea, vomiting, jaundice, constitutional symptoms (such as fever, loss of appetite, etc.), and metastatic symptoms, among others [13].

Imaging plays a crucial role in diagnosing pancreatic neoplasms, particularly this subtype. Radiologically, most pancreatic SPTs present as well-encapsulated masses with heterogeneous central cystic and peripheral solid components, exhibiting a pattern of enhancement that varies based on lesion’s size [2].

On CT, large lesions (>3 cm) appear hypodense without IV contrast, lacking enhancement on pancreatic and portal venous phases in contrasted images. CT effectively highlights common peripheral calcifications in these cases. Conversely, small lesions (<3 cm) exhibit mostly solid, homogeneous components without calcifications. They demonstrate progressive enhancement on hepatic venous phases with IV contrast, appearing isodense [6].

MR findings mirror those on CT, with the added advantage of better visualization of hemorrhagic components. Large tumors are typically hyperintense with a rim of low-signal cap-

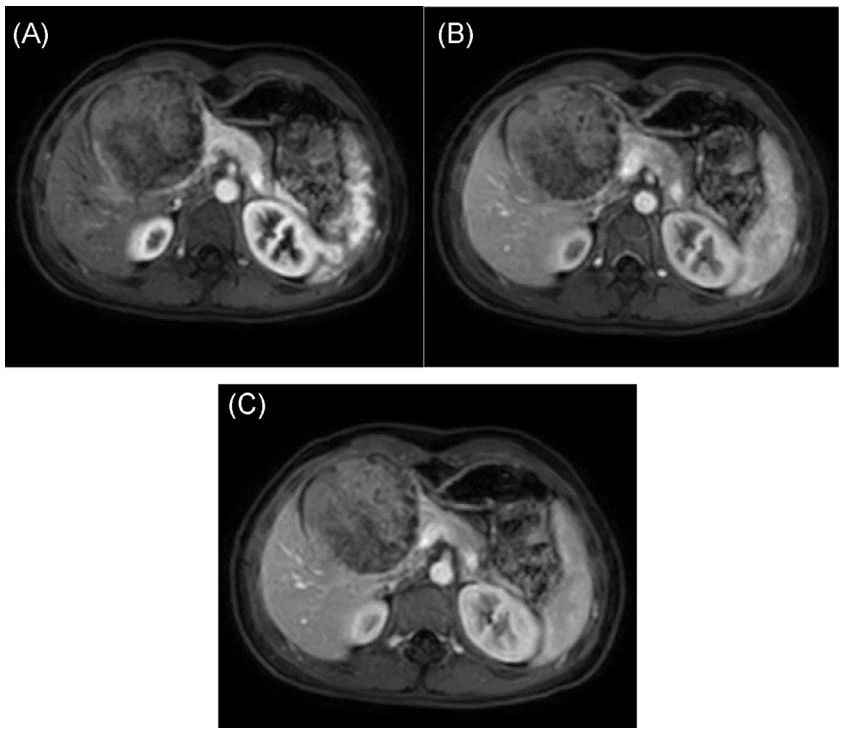

Fig. 5 – (A) T1 MRI with contrast, arterial phase. (B) T1 MRI with contrast, portal venous phase. (C) T1 MRI with contrast, delayed phase.

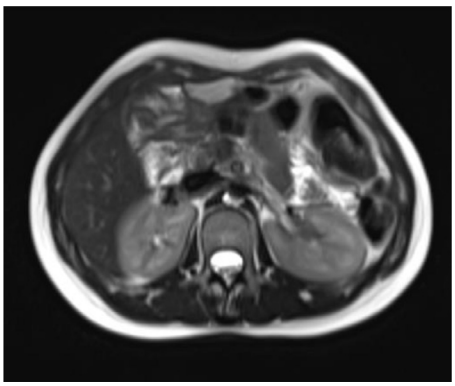

Fig. 6 – One year post Whipple procedure, showing no mass at site of anastomosis, and no local metastases found.

sule on T1, while small lesions are hypointense on T1 and hyperintense on T2, lacking the low-signal capsule [6].

In our case, the radiological appearance deviates slightly, presenting a large lesion arising from the pancreatic head with heterogeneous solid and cystic components and a low-signal capsule. Notably, there's a low T1 signal, potentially attributed to a few small cystic areas observed. The pathogenesis of SPT is still unclear. However, recent studies state that SPT arise from multipotent stem cells which have abnormal WNT-Beta Catenin pathway due to a mutation in exon 3 of Beta Catenin gene leading to abnormal nuclear and cytoplasmic localization of Beta Catenin [1].

Histopathologically, solid pseudopapillary tumors (SPTs) are characterized by eosinophilic round cells with decreased mitotic activity, forming the solid component along with areas of hemorrhage, calcifications, and necrosis, resulting in a pseudopapillary appearance [9]. There is consensus in the literature that positive staining for Beta-catenin and the absence of membranous expression of E-cadherin are typical features of this tumor [3,12,14]. Positive staining for CD10 may also be observed in the majority of cases, although with lower specificity and sensitivity [11,12]. Negative results for neuroendocrine markers such as synaptophysin and chromogranin are indicative, as these markers are highly sensitive in detecting pancreatic neuroendocrine tumors [3]. Furthermore, a negative result for CK7 can be useful in ruling out pancreatic adenocarcinoma, as its sensitivity can be as high as 96% [10].

The management of SPT is primarily surgical, with various approaches depending on the size and location of the tumor, boasting a cure rate of approximately 95%. For very small tumors, enucleation may suffice. In cases where the tumor is located in the tail of the pancreas, distal pancreatectomy with spleen preservation is typically performed, unless there is splenomegaly or vascular or hilar invasion of the spleen [5]. Tumors located in the pancreatic head are managed by pylorus-preserving pancreaticoduodenectomy, commonly known as the Whipple procedure [9]. Routine lymph node dissection and surgical debulking of metastases are rarely performed [13].

The prognosis of solid pseudopapillary tumors (SPT) of the pancreas is generally favorable, even in cases where metastasis is present, with a 5-year survival rate reported as high

as 97% [16]. However, follow-up after surgery is necessary as there is a rare but existing risk of recurrence, estimated to be around 3% based on some studies [7].

Conclusion

We have presented 2 cases of pseudopapillary tumors of the pancreas. This indolent, low-grade neoplasm is often diagnosed incidentally. While characteristic radiological findings can aid in diagnosis in many cases, histopathological examination remains the cornerstone diagnostic method. Treatment of this neoplasm primarily aims for complete curative surgical resection.

Data availability

The authors confirm that all relevant data to this case were included in the article. Further data will be available directly from the author upon request.

Author contribution

Abdalhameed aldmour, Moayad Shaf'ei, Omar Ihmoud, Zaid Ahmad, Omar Ifdielat, Mohammad Sharayah, Abdallah Sharayah, Mohammad Salahaldeen contributed to data curation, investigation, and contributed to writing original draft. Salam Daradkeh contributed to editing and review the final manuscript. All authors gave final approval and agreed to be accountable for all aspects of work ensuring integrity and accuracy. Abdalhameed aldmour, Moayad Shaf'ei, Omar Ihmoud, Zaid Ahmad, Omar Ifdielat, Mohammad Sharayah, Abdallah Sharayah, Mohammad Salahaldeen contributed to data curation, investigation, and contributed to writing original draft. Salam Daradkeh contributed to editing and review the final manuscript. All authors gave final approval and agreed to be accountable for all aspects of work ensuring integrity and accuracy.

Ethics statement

Verbal and written consents were obtained from the patients before writing the case or using investigations to participate.

Patient consent

Written consents were obtained from the patients stating that the patients agree to be included in the case report named: Solid Pseudopapillary Tumor of the Pancreas. The patients were informed that we will be using data about their hospital stay, the related treatments and that the case report may be published and presented to others to read. The patients were assured that confidentiality of the patients' information will be maintained, and no personally identified details will be disclosed in the case report.

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