Clinical Journal of Gastroenterology (2023) 16:937–941
https://doi.org/10.1007/s12328-023-01850-6

CASE REPORT

Solid pseudopapillary neoplasm in a woman presenting with acute pancreatitis: a case report and review of literature

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Received: 10 February 2023 / Accepted: 17 August 2023 / Published online: 1 September 2023

Abstract
Solid pseudopapillary neoplasm (SPN) is a rare pancreatic tumor that typically affects young women in the body and tail of the pancreas. SPN is often asymptomatic in the early stages, so it is initially discovered as a large tumor. In this report, we experienced a case of a relatively small SPN discovered in the setting of acute pancreatitis. Because there have been few reports of SPN being discovered in the situation like our case, we report this case based on a review of the literature.

Keywords Solid pseudopapillary neoplasm · SPN · Acute pancreatitis

Introduction

Solid pseudopapillary neoplasm (SPN) is a rare disease that accounts for 2–3% of all pancreatic tumors, first described by Frantz in 1959 and classified as a potentially malignant neoplasm by the World Health Organization in 2010 [1–3]. It is solid and epithelial in nature, with an unknown direction of differentiation. It is thought to degenerate over time, resulting in internal bleeding, necrosis, calcification, and cyst formation [3, 4]. It is more common in young women, mostly in the body and tail of the pancreas, and has no symptoms in the early stages. Therefore, it is frequently discovered as a large tumor. Surgery is the standard treatment, with a favorable prognosis. However, metastasis and invasion of surrounding tissues have been reported in 5–15% of patients, resulting in a poor prognosis [2, 5]. Therefore, physicians must accurately diagnose this disease in its early stages, which is difficult due to a lack of symptoms.

In this report, we present a case of SPN discovered in the setting of acute pancreatitis, which is rare and has been discussed in the literature. SPN should be considered as a differential diagnosis in young women with acute pancreatitis of unknown cause. This information, we believe, will be useful in the diagnosis and treatment of similar cases.

Case report

A 20-year-old woman was brought to our hospital with upper abdominal pain and vomiting. There were no special notes on her medical history, medications, or family history. She had been drinking alcohol about once a week, consuming approximately 40 g of pure alcohol per drink.

An abdominal examination revealed tenderness in the left upper abdomen but no fever or significant vital signs. Her laboratory findings revealed a leukocyte count of 15,500/μL, a neutrophil percentage of 91.3%, and a C-reactive protein level of 0.26 mg/dL, indicating a neutrophil predominance and mild inflammatory response. Her pancreatic-derived amylase and lipase levels were found to be elevated (797 U/L and 1106 IU/L, respectively). A contrast-enhanced computed tomography (CT) scan revealed pancreatic swelling mainly in the body and tail of the pancreas, as well as a

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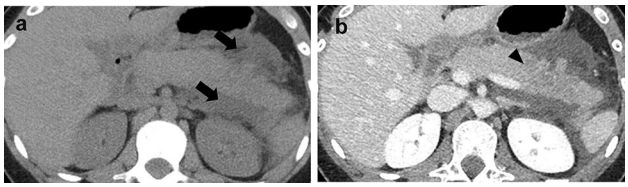

Fig. 1 Contrast computed tomography (CT) scan findings on admission. **a** A plain CT scan showing swelling in the body and tail area of the pancreas, with high density of adipose tissues caused by inflammation (arrows). **b** A contrast CT showing a poorly contrasted area at the tail of the pancreas caused by acute pancreatitis (arrowheads)

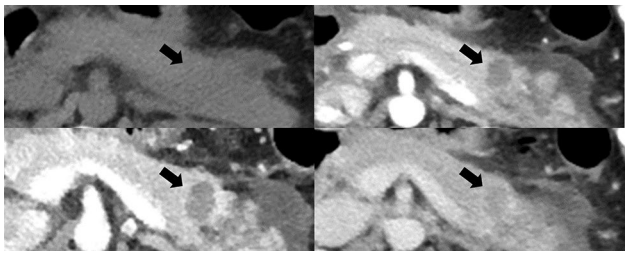

Fig. 2 Dynamic computed tomography (CT) scan findings two days after admission. Four phases of dynamic CT focusing on the pancreatic tumor and showing the tumor in the pancreatic tail with a progressive contrast effect (arrows)

poor contrast area in the tail. She was diagnosed with acute pancreatitis (contrast-enhanced CT Grade 1) and hospitalized (Fig. 1). Following admission, treatment was started with a large volume of fluid replacement. The next day, after admission, the abdominal pain and vomiting subsided. On the second day of hospitalization, a dynamic CT of the pancreas revealed that the extent of peripancreatic inflammation had been mildly reduced, but the area of poor contrast at the pancreatic tail had not changed significantly. However, a low-absorption area approximately 15 mm in size was observed caudally with a contrast-impaired area, with a progressive contrast effect at the boundary between the normal and poor contrast areas (Fig. 2). Hence, pancreatitis associated with the tumor was suspected. The dilation of the main pancreatic duct caudal to the tumor site was obscured by inflammatory effects of pancreatitis. After starting treatment, abdominal findings and blood tests gradually improved, and the patient was placed on a diet on the sixth day of the disease. On the twelfth day, magnetic resonance imaging (MRI) was performed to look for the nodule on the pancreatic tail that had been identified on CT. The nodule was located on the main pancreatic duct and had a high signal on T2-weighted and diffusion-weighted images as well as a low apparent diffusion coefficient value (Fig. 3a, b). T1-weighted image showed low signal at the tumor. There is no evidence of hemorrhage inside the tumor (Fig. 3c).

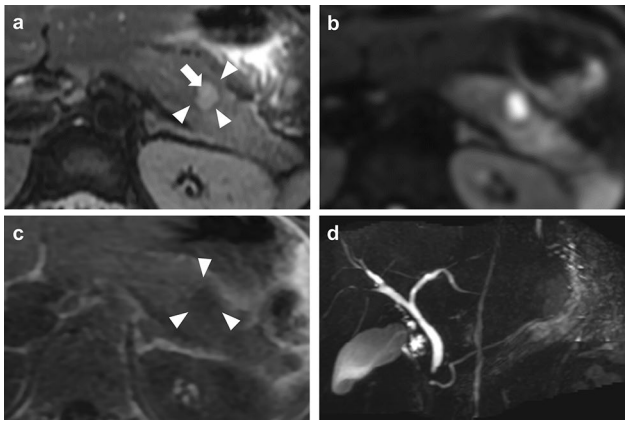

Fig. 3 Magnetic resonance image (MRI) findings. **a** The MRI T2 phase showing high signal in the tumor (arrow), with a film-like structure around the tumor (arrowhead). **b** Diffusion-weighted MRI imaging. **c** MRI T1 phase showing low signal in the tumor (arrowhead). **d** The main pancreatic duct in the head and body were normal and caudal to the tumor site was indistinct

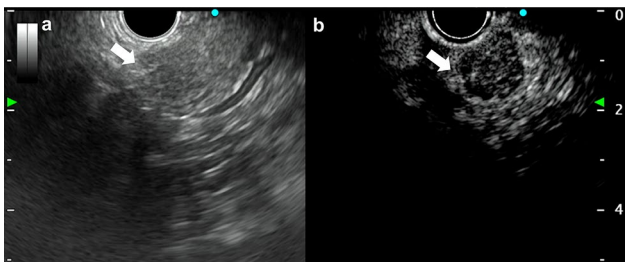

Fig. 4 Endoscopic ultrasound (EUS) findings. **a** EUS B-mode showing hypoechoic findings in the tumor (arrow). **b** Contrast-enhanced EUS showing that the tumor is persistently stained by contrast medium (arrow)

Magnetic resonance cholangiopancreatography showed that the dilation of the main pancreatic duct was obscured by inflammation (Fig. 3d). Based on these imaging findings, SPN or atypical neuroendocrine tumor was raised as differentials and was most suspected due to age and gender. The patient was discharged on the 17th day with an objective to carry out tissue diagnosis of the tumor after the pancreatitis was in remission.

Two months after the onset of pancreatitis, endoscopic ultrasonography identified the nodule in the pancreatic tail as a hypoechoic mass, and contrast-enhanced ultrasonography revealed a weak contrast effect and borderline clarity for about 15 s (Fig. 4). There was no evidence of chronic pancreatitis in the background. An endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) was performed on the nodule using a 22-gauge FNA needle. Cytology by Papanicolaou staining shows round small cells with nucleomegaly clinging to thin branching vessels, which are the findings of SPN. The specimen was

pathologically examined, revealing small, poorly atypical round cells with relatively homogeneous nuclei. Immunohistochemically, β-catenin and vimentin were positive, while CD10 was partially positive and chromogranin A, trypsin, and cytokeratin CK19 were negative (Fig. 5a–g). Based on these findings, a diagnosis of SPN was made. Two months after the onset of pancreatitis, CT findings showed no change in tumor size and laparoscopic pancreatic resection with splenic sparing was performed. Gross finding of the surgical specimen showed that the tumor was 20 × 17 mm in diameter and was located at the pancreatic tail (Fig. 6a). The main pancreatic duct was found to be incorporated into the tumor and running through it at the tumor site (Fig. 6b). The pathology findings were SPN with findings similar to those of EUS-FNA. The pathology also confirmed that the main pancreatic duct recognized in Hematoxylin and eosin staining and immunohistochemistry staining by CK19, which is in pancreatic duct into the tumor (Fig. 6b–f). The diameters of the main pancreatic duct were evaluated at three different positions: on the head side; 1000 × 200 µm, inside the tumor; 380 × 85 µm, and on the caudal side; 950 × 85 µm. This showed that the deformation of the main pancreatic duct was present inside the tumor. The caudal pancreatic parenchyma was atrophic compared with the pancreatic head side, and fibrosis was observed between the lobes regarding to the findings of the change after pancreatitis. There was no recurrence of pancreatitis or SPN metastasis eight months after surgery.

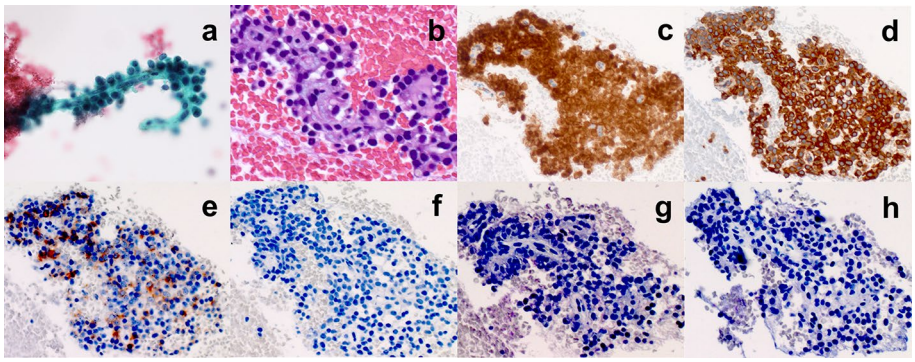

Fig. 5 Cytological and histological findings of the pancreas. Pancreatic cytology and tissue were collected from the tumorous area using an endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA). **a** Papanicolaou staining (400×). **b** Hematoxylin and eosin staining (100×). **c** β-catenin (100×). **d** Vimentin (100×). **e** CD10 (100×). **f** Chromogranin A (100×). **g** Trypsin (100×). **h** CK19 (100×)

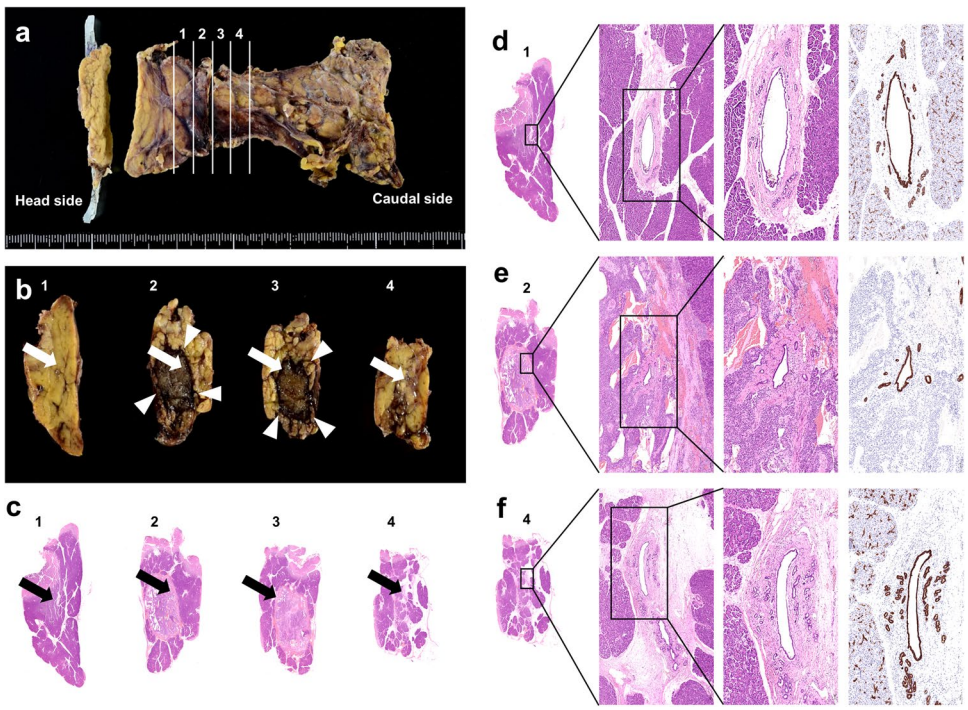

Fig. 6 Surgical specimen findings. **a**, **b** Pathological specimen. The arrows indicate the main pancreatic duct. The arrowheads indicate the tumor. **c** Hematoxylin and eosin staining (Overview). The arrows indicate the main pancreatic duct. **d–f** Hematoxylin and eosin staining (Overview, 50×, 100×) and immunohistochemistry staining CK19 (100×)

Table 1 Summary of reported cases

No	Age	Sex	Size (mm)	Location	CT/MRI imaging findings	Preoperation diagnosis	Diagnostic method	Etiology of pancreatitis	References
1	27	F	32	Tail	Calcification and poorly contrast effect	Suspect of cystic tumor	Imaging findings	Unknown	[8]
2	21	F	80	Tail	Cystic changes with hemorrhage	N/A	N/A	Internal bleeding	[9]
3	31	F	50	Body	Light contrast effect with cystic changes	Suspect of SPN	Imaging findings	Unknown	[10]
4	12	F	80	Tail	Cystic changes	N/A	N/A	Internal bleeding	[11]
5	11	F	68	Head	Heterogeneous density at T1/T2 phage	SPN	EUS-FNA	Unknown	[12]
6	55	M	55	Body	Light contrast effect with cystic changes	SPN	EUS-FNA	Deformation of MPD	[13]
7	36	M	12	Body	Poorly contrast effect	Suspect of pancreatic cancer	Imaging findings, Cytology of pancreatic juice	Deformation of MPD	[14]
8	14	M	30	Tail	Light contrast effect	Suspect of SPN	Imaging findings	Deformation of MPD	[15]
Our case	20	F	20	Tail	Light contrast effect	SPN	EUS-FNA	Deformation of MPD	

Discussion

SPN is a spherical tumor that grows widely on the outer pancreas. Because SPN is a hypervascularized enhancing tumor, dynamic CT and MRI show a spherical tumor with a contrast effect, which tends to occur in the body and tail of the pancreas. It may also present with calcification at the tumor margins and inside the tumor and undergo repeated intra-tumor hemorrhage and necrosis, leading to cystic changes [6]. As a result, there may be a mixture of substantial and cystic components, with corresponding imaging findings [3].

When the above imaging diagnosis suggests SPN, EUS-FNA is performed to confirm the diagnosis, or direct surgical treatment is used for diagnostic purposes. Pathologically, SPN shows a fold and pseudopapillary pattern. Immunological staining is positive for vimentin, CD10, CD56, and β-catenin, whereas chromogranin A, which is positive in neuroendocrine tumor, is often negative [3]. These findings are consistent with the present case.

In our case, contrast-enhanced CT made it difficult to identify the tumor at first. This may be because the inflammation spread to the pancreas and surrounding area, making it more difficult to distinguish between the tumor and pancreatic parenchyma, and because it was a small tumor with a maximum diameter of 20 mm and no cystic component, calcification, or other SPN-associated findings. Therefore, dynamic CT or MRI is useful in diagnosing the presence of a tumor and providing more definitive information based on the finding characteristics of SPN obtained.

SPN may be clinically manifested as abdominal pain or discomfort, but it is often asymptomatic. Therefore, many cases are discovered in an expanded state [7]. In a study of 302 cases, the average tumor diameter at onset was 75 mm [4]. Our SPN case was discovered in the setting of acute pancreatitis. There were eight cases diagnosed with SPN at the onset of acute pancreatitis using the search terms "SPN" or "acute pancreatitis" in PubMed and these cases were included in the literature review. A literature review for SPN with acute pancreatitis revealed tumor diameters ranging from 12 to 80 mm, with an average of 50.9 ± 24.8 mm in eight cases (Table 1) [8–15]. Among these, the findings consistent with SPN by imaging findings include cysts and calcification; however, the smaller tumors in Case No. 7 and 8, being 12 mm and 30 mm in size respectively, do not show these characteristic findings.

In these cases, two cases were diagnosed by EUS-FNA, both of which were relatively large in size (55 mm and 68 mm). Alternatively, in the present case, although the size of the tumor was as small as 20 mm, it was possible to make a definitive diagnosis by EUS-FNA at an early stage.

Two mechanisms of acute pancreatitis in SPN have been proposed: (1) rapid enlargement of the tumor due to intra-tumor hemorrhage causing ischemia and obstruction of the pancreatic duct, and (2) fibrosis and degeneration of the tumor adjacent to the pancreatic duct, causing deformation

of the main pancreatic duct wall [9, 14]. In our case, the main pancreatic duct ran through the tumor, and it was contemplated that the deformation of the main pancreatic duct by the tumor caused the pancreatitis. As mentioned in the literature review, in Case No. 7 and 8 with small tumors of 12 mm and 30 mm in size, respectively, smaller sized SPN is positioned along the main pancreatic duct and deformation may be the cause of pancreatitis. On the other hand, in Case No. 2 and 4, where the tumor size was as large as 80 mm, intratumoral hemorrhage was the cause. In the case of pancreatitis caused by a small tumor, it may be difficult to recognize the tumor, and this possibility should be considered when the cause is unknown.

In conclusion, we experienced a case of SPN diagnosed after acute pancreatitis. SPN should be considered in the differential diagnosis of young women with acute pancreatitis. Even if the tumor is small, SPN can cause acute pancreatitis. However, when the tumor diameter is small, characteristic imaging findings such as cysts and calcification may be missed, and dynamic CT and MRI are considered useful for diagnosis. Furthermore, EUS-FNA with histological examination may be useful for definitive diagnosis.

Acknowledgements The author would like to thank Enago for the English language review.

Compliance with ethical standards

Conflict of interest The authors declare that they have no current financial arrangement or affiliation with any organization that may have a direct influence on their work.

Human/animal Rights All procedures were approved by the University of Niigata Institutional Review Board and conducted in accordance with the principles outlined in the Declaration of Helsinki.

Informed consent A written informed consent was obtained from the patient for the clinical data collection and to publish the results based on them.

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