



Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

A solid pseudopapillary neoplasm of the pancreas in a man presenting with acute pancreatitis: A case report



Emi Chikuie^a, Saburo Fukuda^{b,*}, Hirofumi Tazawa^b, Toshihiro Nishida^c,
Hideto Sakimoto^{b,d}

^a Department of Surgery, Yoshida General Hospital, 3666 Yoshida, Yoshida-Chou, Akitakata City, Hiroshima, 731-0595, Japan

^b Department of Surgery, Chugoku Rousai Hospital, 1-5-1 Hirotagaya, Kure, Hiroshima, 737-0193, Japan

^c Department of Diagnostic Pathology, Chugoku Rousai Hospital, 1-5-1 Hirotagaya, Kure, Hiroshima, 737-0193, Japan

^d Faculty of Medicine, Hiroshima University, 1-2-3 Kasumi, Hiroshima, 734-8551, Japan

ARTICLE INFO

Article history:

Received 17 December 2016

Accepted 14 January 2017

Available online 17 January 2017

Keywords:

Solid pseudopapillary neoplasm

Acute pancreatitis

Pancreatic cancer

ABSTRACT

INTRODUCTION: A solid pseudopapillary neoplasm (SPN) of the pancreas is rare neoplasm that occurs predominantly in young women. The clinical presentation of SPNs is nonspecific, but acute pancreatitis is rare in the reported literature.

PRESENTATION OF CASE: A 36-year-old man was referred to our hospital because of upper abdominal pain and elevation of serum amylase. A computed tomography (CT) scan showed swelling of the pancreas body and a poorly enhanced and indistinct mass in the pancreas body. He was diagnosed with acute pancreatitis. The symptom was improved with conservative treatment, but acute pancreatitis recurred twice during a period of 2 months. Magnetic resonance cholangiopancreatography (MRCP) and endoscopic retrograde cholangiopancreatography (ERCP) showed stenosis of the MPD adjacent to the mass. Distal pancreatectomy was performed because the mass in the pancreas body seemed to cause repeated acute pancreatitis and malignant pancreatic cancer could not be excluded. Immunohistochemically, a diagnosis of SPN of the pancreas was made from the resected specimen.

DISCUSSION: To the best of our knowledge, only 6 cases have been reported in the literature concerning the SPN presenting with acute pancreatitis.

CONCLUSION: We report a man with a small SPN of the pancreas presenting with acute pancreatitis and mimicking pancreatic cancer. We should be aware that this rare pancreatic tumor can become a cause of acute pancreatitis.

© 2017 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

A solid pseudopapillary neoplasm (SPN) of the pancreas is rare pancreatic tumor and occurs predominantly in young women [1,2]. The clinical presentation of SPNs is nonspecific, but acute pancreatitis is rare in the reported literature. Herein, we report a man with a small SPN of the pancreas who presented with acute pancreatitis and mimicking pancreatic cancer.

Abbreviations: CT, computed tomography; MPD, main pancreatic duct; MRCP, magnetic resonance cholangiopancreatography; ERCP, endoscopic retrograde cholangiopancreatography; Syn, synaptophysin; CgA, chromogranin A; SPN, solid pseudopapillary neoplasm; MRI, magnetic resonance imaging; EUS-FNA, endoscopic ultrasonography guided fine-needle aspiration.

* Corresponding author.

E-mail address: safukuda@aol.com (S. Fukuda).

<http://dx.doi.org/10.1016/j.ijscr.2017.01.026>

2210-2612/© 2017 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

2. Presentation of case

A 36-year-old man was referred to our hospital because of upper abdominal pain and elevation of serum amylase. His medical history was unremarkable except for a duodenal ulcer. He was a moderate drinker but had no history of acute pancreatitis. Laboratory data on admission showed elevated serum levels of pancreatic enzymes: amylase, 1600 IU/L (normal range, 40–129 IU/L); p-amylase, 1541 IU/L (18–55 IU/L); and lipase, 2243 IU/L (17–57 IU/L). Serum levels of the tumor markers carcinoembryonic antigen and carbohydrate antigen 19-9 were within normal ranges. A contrast-enhanced computed tomography (CT) scan showed diffuse enlargement of the pancreas with inflammatory change in peri-pancreatic fatty tissue and a poorly enhanced and indistinct nodule in the pancreas body. The distal side of the main pancreatic duct (MPD) was dilated (Fig. 1a and b). He was admitted with a diagnosis of acute pancreatitis. The pancreatitis was improved with conservative treatment and he was discharged three weeks later. However, acute pancreatitis recurred twice during a period

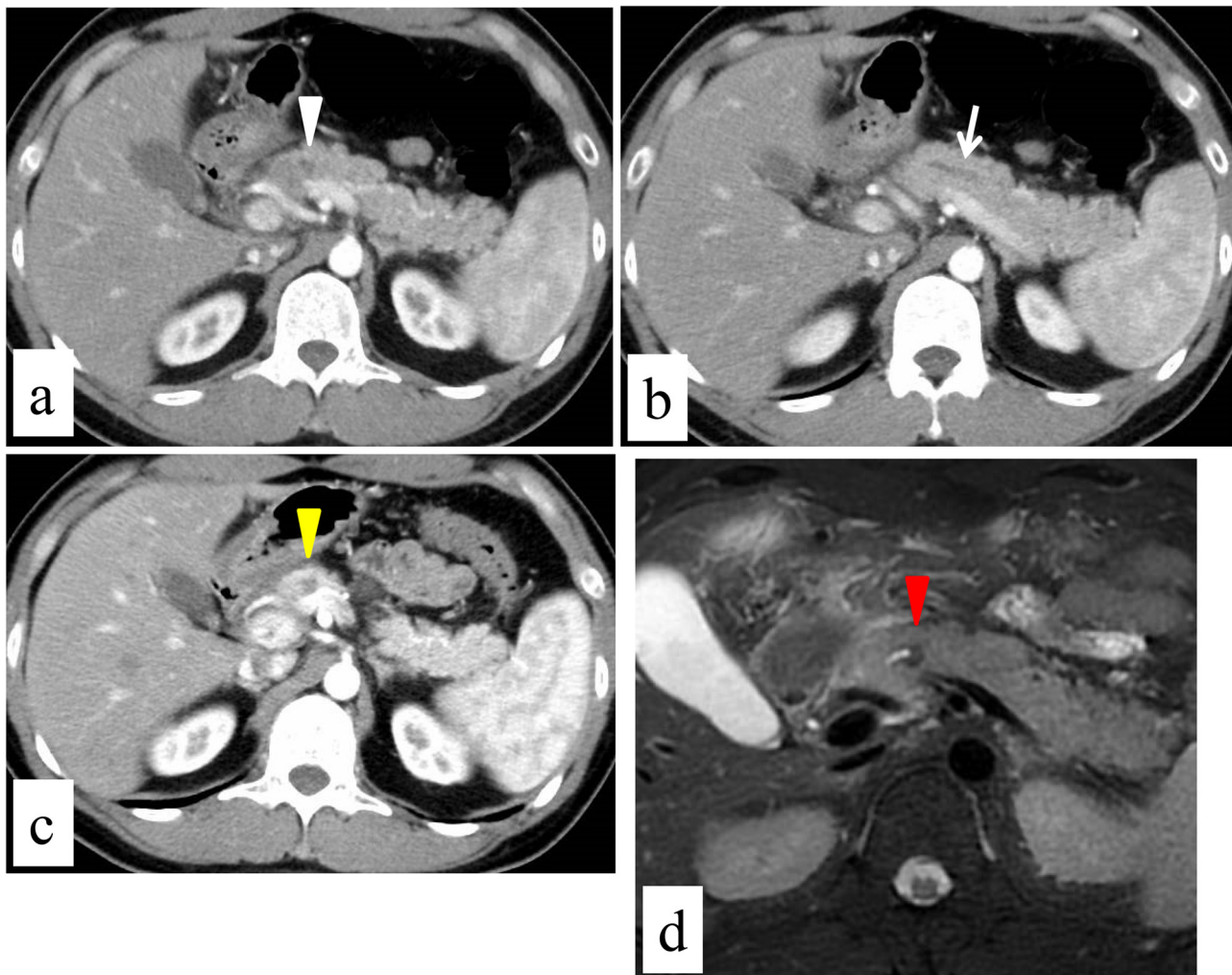


Fig. 1. (a) Contrast-enhanced computed tomography (CT) scan showed a poorly enhanced and indistinct nodule in the pancreas body (white arrowhead). (b) The distal side of the main pancreatic duct (MPD) was dilated (white arrow). (c) The mass in the pancreas body was 10 mm in diameter 53 days later (yellow arrowhead). (d) The mass showed low signal intensity on T2-weighted images obtained by magnetic resonance imaging (MRI) (red arrowhead).

of two months with repeated admission and discharge. The mass in the pancreas body gradually became clearer, and a mass of 10 mm in diameter was detected 53 days later by CT (Fig. 1c). The mass contained no calcification or cystic lesion. The mass showed low signal intensity on T2-weighted images and diffusion-weighted images obtained by magnetic resonance imaging (MRI) (Fig. 1d). Magnetic resonance cholangiopancreatography (MRCP) showed stenosis of the MPD adjacent to the mass (Fig. 2a). Endoscopic retrograde cholangiopancreatography (ERCP) also showed stenosis of the MPD (Fig. 2b). Cytology of pancreatic juice collected during the ERCP did not reveal malignant cells (Fig. 2c). Although a definitive diagnosis could not be made, the recurrent acute pancreatitis was thought to be related to the stenosis of the MPD caused by the mass in the pancreas body. Furthermore, the possibility of a malignant disease such as pancreatic cancer for the mass could not be excluded from the radiological findings. Distal pancreatectomy with regional lymph node dissection was performed. Macroscopically, a well-circumscribed mass, measuring 12 mm, in the pancreas body was observed (Fig. 3a and b). Microscopically, most of the mass consisted of a fibrous nodule, but an irregularly arranged pseudopapillary structure composed of fairly uniform tumor cells was seen around the fibrous nodule. The tumor cells had proliferated invasively in the pancreatic parenchyma (Fig. 3c and d). The tumor located adjacent to the MPD and the fibrous

and degenerative change had caused a deformity of the wall of the MPD (Fig. 3e). The border region between the fibrous nodule and the tumor contained calcification, old hemorrhage and cholesterol crystals (Fig. 3f). Immunohistochemically, the tumor was positive for β -catenin, vimentin, CD10, synaptophysin (Syn) (Fig. 4a–d), CD56, and neuron-specific enolase (NSE), but negative for chromogranin (CgA) (Fig. 4e) and insulin. The Ki67 labeling index was approximately 3% (Fig. 4f). Based on these findings, a diagnosis of SPN of the pancreas was made. Lymph node metastasis was not found. The patient's postoperative course was uneventful and he was discharged on the 22nd day after the operation. At a 32-month follow-up after resection, the patient did not have any recurrence of SPN or pancreatitis.

3. Discussion

SPN of the pancreas is a rare neoplasm accounting for only about 0.13–2.7% of all pancreatic tumors [1]. SPN is regarded as a low-grade malignant potential tumor with an excellent prognosis after complete resection. In a large review of the English literature including 718 patients with SPN [2], the male-to-female ratio was 1:9.78 with a median age of 21.97 years and the mean diameter of the tumors was 6.08 cm. The clinical presentation is nonspecific. The majority of patients presented with abdominal

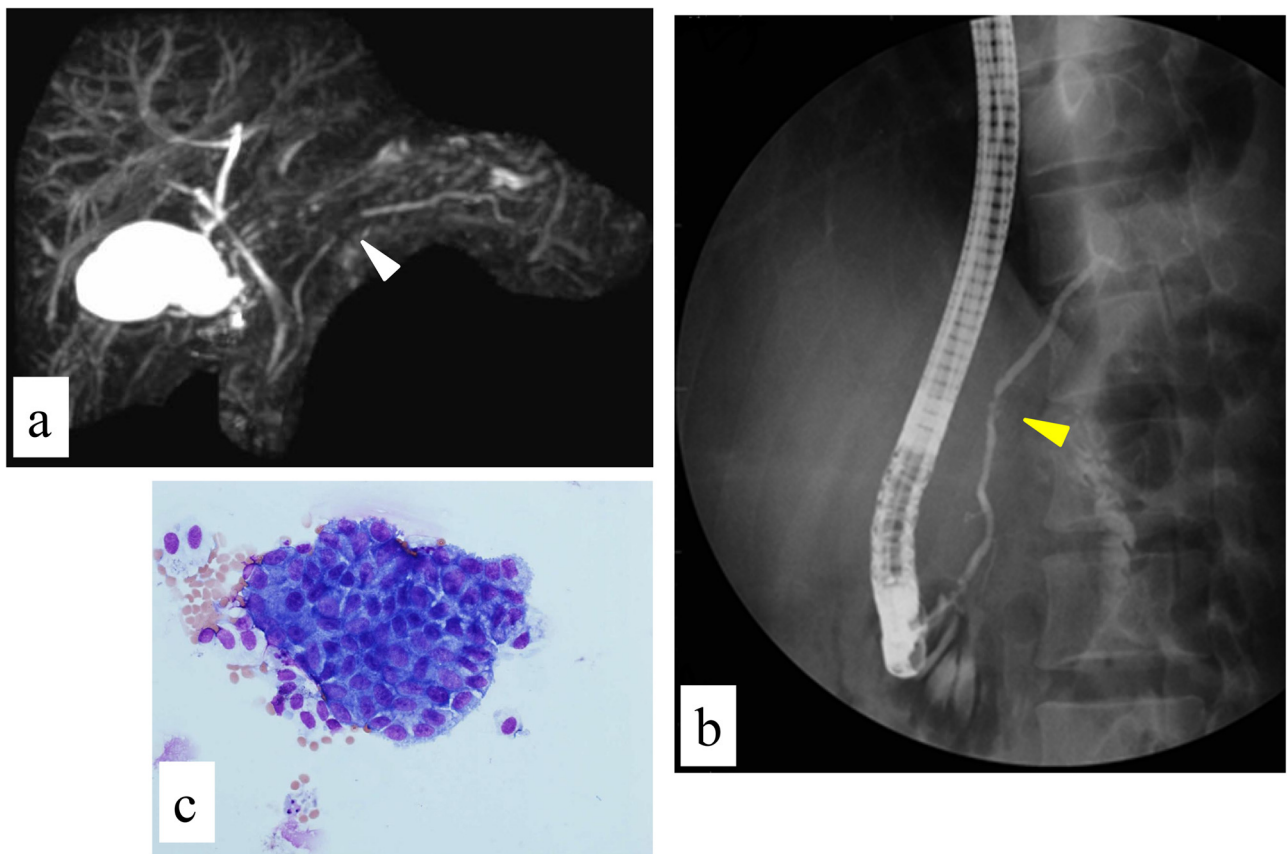


Fig. 2. (a) Magnetic resonance cholangiopancreatography (MRCP) showed stenosis of the MPD adjacent to the mass (white arrowhead). (b) Endoscopic retrograde cholangiopancreatography (ERCP) also showed stenosis of the MPD (yellow arrowhead). (c) Cytology of pancreatic juice did not reveal malignant cells.

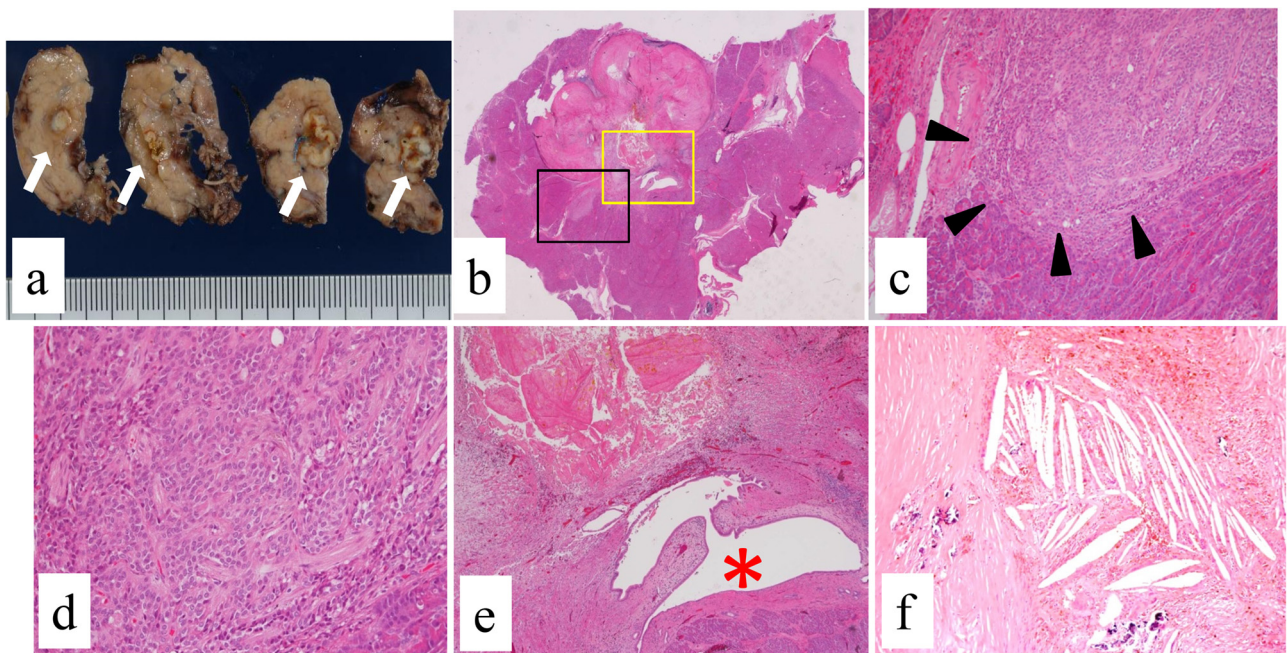


Fig. 3. (a) Macroscopically, a well-circumscribed mass in the pancreas body was observed (white arrows). (c, d) Magnified view of the black frame area in (b). The tumor cells had proliferated invasively in the pancreatic parenchyma (black arrowhead). An irregularly arranged pseudopapillary structure composed of fairly uniform tumor cells was seen around the fibrous nodule. (e) Magnified view of the yellow frame area in (b). Fibrous and degenerative change induced a deformity of the wall of the MPD (asterisk indicating MPD). (f) The border region between the fibrous nodule and the tumor contained calcification, old hemorrhage and cholesterol crystals.

pain, abdominal mass or fullness, but gastrointestinal obstruction, anemia, pancreatitis [3], bile duct obstruction, or traumatic rup-

ture with hemoperitoneum [4] was less frequently noted. On the basis of the review, typical SPN occurs predominantly in young

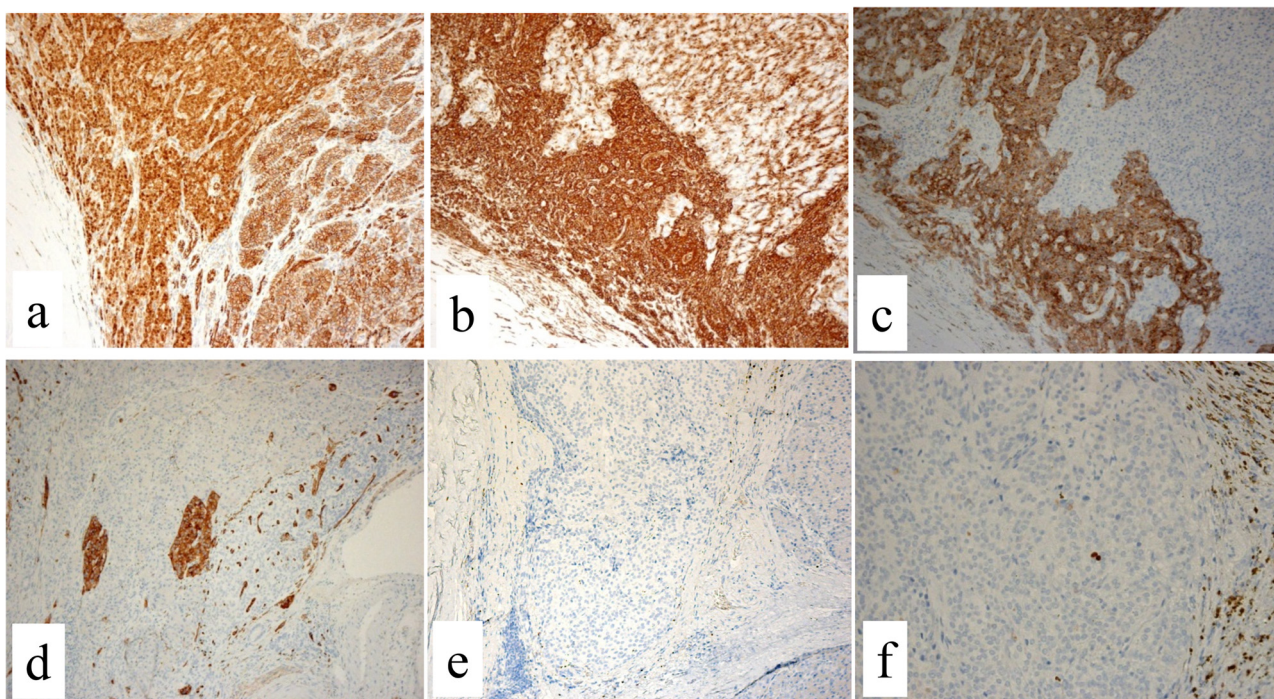


Fig. 4. Immunohistochemically, the tumor was positive for β -catenin (a), vimentin (b), CD10 (c), and synaptophysin (Syn) (d) but negative for chromogranin (CgA) (e). The Ki67 labeling index was approximately 3% (f).

Table 1
Reported cases of SPN presenting with acute pancreatitis.

	Case 1 [11]	Case 2 [12]	Case 3 [13]	Case 4 [14]	Case 5 [15]	Our case
Age	27	21	31	12	55	36
Gender	Female	Female	Female	Female	Male	Male
Location	Tail	Tail	Body	Tail	Body	Body
Size (cm)	3.2	8.0	5.0	8.0	5.5	1.2
MPD imaging	Obstruction	n.d	Narrowing and dilatation	n.d	Compression	Stenosis
Preoperative diagnosis	s/o Cystic pancreatic tumor	n.d	s/o SPN	n.d	SPN (EUS-FNA)	s/o Pancreatic cancer
Operation	DP	DP	DP	Enucleation	DP	DP
Prognosis	2 yrs no recurrence	3.5 yrs no recurrence	2 yrs no recurrence	n.d	6 mo no recurrence	2.5 yrs no recurrence

MPD, main pancreatic duct; DP, distal pancreatectomy; SPN, solid pseudopapillary neoplasm; s/o, suspected of; EUS-FNA, endoscopic ultrasonography guided fine-needle aspiration; n.d, not described.

females and the tumor is usually large and is accompanied by non-specific abdominal complaints. The current case is not typical in some aspects compared with typical SPNs. First, the tumor developed in a middle-aged man. Second, the tumor was quite small and mimicked a malignant pancreatic tumor. Third, the first clinical presentation was acute pancreatitis.

Male patients with SPN have accounted for less than 10% of the patients. SPN shows a clear female predilection with very few reported cases in male patients [2]. Several reports suggested that sex hormones play a role in the pathogenesis or growth of SPN [5]. Machado et al. examined clinical differences between male and female patients. The mean age of male patients was higher than that of female patients. Although tumor size, location and symptoms were the same, tumor aggressiveness such as portal vein invasion and/or development of metastases was more frequent in male patients. They concluded that SPN in male patients has delayed onset and aggressiveness compared with female patients [6].

The typical CT imaging features of SPNs are a large well-encapsulated mass with varying solid and cystic components caused by hemorrhagic degeneration. However, diagnosis can be difficult, especially for in small tumors. Baek et al. reported that SPNs of less than 3 cm in diameter were purely solid lesions

and lacked a cystic component [7]. Furthermore, 75% of small SPNs lacked any type of calcification. Consequently, small atypical SPNs have been frequently misdiagnosed as pancreatic cancer [7]. Because of its distinctive biological characteristics and prognosis, SPN should be distinguished from other pancreatic cancers.

Endoscopic ultrasound-guided fine needle aspiration (EUS-FNA) has been reported to be a useful diagnostic tool for pancreatic tumors [8]. Preoperative diagnosis of SPN is made in 75% of cases on the basis of EUS-FNA cytology [9]. However, seeding of the needle tract by cancer cells and complications such as bleeding, pancreatic fistula and biliary fistula during the procedure have been reported [10]. FNA should be performed only for cases in which radiological diagnosis is not clear.

In our case, the tumor was small and lacked a cystic component and calcification on CT imaging. Additionally, the MPD showed stenosis adjacent to the tumor. Due to these atypical images, it was difficult to make a speculation about SPN. This case might have been a good indication for EUS-FNA, but the tumor was difficult to detect by EUS and EUS-FNA was not performed. Because the tumor seemed to cause the recurrent acute pancreatitis and the possibility of a malignant pancreatic tumor could not be excluded,

distal pancreatectomy with regional lymph node dissection was performed.

To examine the rarity of SPNs with acute pancreatitis, we performed searches of the PubMed database and Japan Medical Abstracts Society using the following keywords: (acute pancreatitis) and (SPN). To the best of our knowledge; only 6 cases including the current case have been reported in the literature (Table 1) [11–15]. Four patients were female and two were male. The mean age was 30.3 years and the mean size of tumors was 5.15 cm. All of the tumors were located in the pancreas body or tail. MPD imaging was described for four cases: obstruction in one case; narrowing (or stenosis) and dilatation in two cases and compression in one case. Despite the large size of the mass; MPD stenosis or dilatation has been reported to be rare in SPN. Generally; stenosis or dilatation of the MPD is useful for differentiating SPNs from malignant pancreatic tumors [7]; but we should be aware that MPD stenosis or dilatation is seen also in SPNs. The operative procedures were distal pancreatectomy in five cases and tumor enucleation in one case. After complete resection of the tumor; postoperative courses of all but one of the cases were uneventful with no recurrence of SPN or acute pancreatitis. There is no doubt that the recurrent acute pancreatitis in our case was related to the stenosis of the MPD caused by the SPN. The question arises as to why the SPN induced acute pancreatitis because SPN of the pancreas is considered to be a slow-growing neoplasm [16]. The exact mechanism of acute pancreatitis is still unclear; but Sakagami et al. hypothesized that non-traumatic internal bleeding may lead to rapid tumor expansion; which results in acute pancreatitis triggered by ischemia; distension or duct obstruction [12]. In our case; we speculate that the fibrous and degenerative change around the tumor located adjacent to the MPD induced a deformity in the wall of the MPD. This change might have induced stenosis of the MPD and acute pancreatitis.

4. Conclusion

We report a man with a small SPN of the pancreas presenting with acute pancreatitis and mimicking pancreatic cancer. We should be aware that this rare pancreatic tumor can become a cause of acute pancreatitis.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Author contribution

EC and SF made substantial contributions to conception and design and acquisition and interpretation of data. HT, TN and HS were involved in drafting the manuscript or revising it critically for important intellectual consent. SF gave final approval of version to be published. All authors read and approved the final manuscript.

Conflict of interest

The authors declare that they have no competing interests.

Funding

None.

Open Access

This article is published Open Access at scimedirect.com. It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.

Ethical approval

This study does not include a research study.

Registration of research studies

This is not research study.

Guarantor

Saburo Fukuda.

Acknowledgements

None.

References

- [1] R.C. Martin, D.S. Klimstra, M.F. Brennan, K.C. Conlon, Solid-pseudopapillary tumor of the pancreas: a surgical enigma? *Ann. Surg. Oncol.* (2002) 35–40.
- [2] T. Papavramidis, T. Papavramidis, S. Papavramidis, Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in english literature. *J. Am. Coll. Surg.* 96 (2005) 5–72.
- [3] A.K. Madan, C.B. Weldon, W.P. Long, D. Johnson, A. Raafat, Solid and papillary epithelial neoplasm of the pancreas. *J. Surg. Oncol.* 850 (2004) 193–198.
- [4] C. Mao, M. Guvendi, D.R. Domenico, K. Kim, N.R. Thomford, J.M. Howard, Papillary cystic and solid tumors of the pancreas: a pancreatic embryonic tumor? Studies of three cases and cumulative review of the world's literature. *Surgery* 118 (1995) 821–828.
- [5] V. Canzonieri, M. Berretta, A. Buonadonna, M. Libra, E. Vasquez, E. Barbagallo, A. Bearz, S. Berretta, Solid pseudopapillary tumour of the pancreas. *Lancet Oncol.* 4 (2003) 255–256.
- [6] M.C. Machado, M.A. Machado, T. Bacchella, J. Jukemura, J.L. Almeida, J.E. Cunha, Solid pseudopapillary neoplasm of the pancreas: distinct patterns of onset, diagnosis, and prognosis for male versus female patients. *Surgery* 143 (2008) 29–34.
- [7] J.H. Baek, J.M. Lee, S.H. Kim, S.J. Kim, S.H. Kim, J.Y. Lee, J.K. Han, B.I. Choi, Small (<3 cm) solid pseudopapillary tumors of the pancreas at multiphase multidetector CT. *Radiology* 257 (2010) 97–106. <http://dx.doi.org/10.1148/radiol.10092089>.
- [8] M.J. Hewitt, M.J. McPhail, L. Possamai, A. Dhar, P. Vlavianos, K.J. Monahan, EUS-guided FNA for diagnosis of solid pancreatic neoplasms: a meta-analysis. *Gastrointest. Endosc.* 75 (2012) 319–331. <http://dx.doi.org/10.1016/j.gie.2011.08.049>.
- [9] N. Jani, J. Dewitt, M. Eloubeidi, S. Varadarajulu, V. Appalaneni, B. Hoffman, W. Brugge, K. Lee, A. Khalid, K. McGrath, Endoscopic ultrasound-guided fine-needle aspiration for diagnosis of solid pseudopapillary tumors of the pancreas: a multicenter experience. *Endoscopy* 40 (2008) 200–203.
- [10] G. Pettinato, D. Di Vizio, J.C. Manivel, S.E. Pambuccian, P. Somma, L. Insabato, Solid-pseudopapillary tumor of the pancreas: a neoplasm with distinct and highly characteristic cytological features. *Diagn. Cytopathol.* 27 (2002) 325–334.
- [11] Y. Mitsui, S. Yamamoto, T. Kunieda, N. Mimura, Y. Iguchi, M. Shimabara, M. Kuboki, K. Ohmoto, S. Iwamoto, T. Tsunoda, A case of small solid-pseudopapillary tumor of the pancreas complicated by lymph node metastasis. *Kawasaki Igakkaishi* 28 (2002) 109–113. In Japanese with English abstract.
- [12] J. Sakagami, K. Kataoka, Y. Sogame, A. Taii, T. Ojima, D. Kanemitsu, R. Takada, R. Ito, T. Motoyoshi, H. Yasuda, S. Mitsufuji, T. Okanoue, Solid pseudopapillary tumor as a possible cause of acute pancreatitis. *JOP* 5 (2004) 348–352.
- [13] Y. Morimoto, K. Kawamoto, T. Paku, K. Ogasawara, A case of solid-pseudopapillary tumor of the pancreas with acute pancreatitis. *J. Jpn. Coll. Surg.* 31 (2006) 992–995. In Japanese.
- [14] Y. Ozturk, O.B. Soyulu, B. Gurcu, R. Ortac, H. Cakmakci, A. Coker, Solid pseudopapillary tumor of the pancreas as a cause of recurrent pancreatitis. *Acta Gastroenterol. Belg.* 71 (2008) 390–392.
- [15] S. Kato, T. Honbu, M. Zakimi, K. Chinen, T. Kubota, M. Arashiro, K. Kikuchi, T. Murakami, F. Kunishima, A case of solid-pseudopapillary neoplasm in a middle-aged male preoperatively diagnosed by endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA). *Suizo* 29 (2014) 263–270. In Japanese.
- [16] T. Kato, N. Egawa, T. Kamisawa, Y. Tu, M. Sanaka, N. Sakaki, A. Okamoto, N. Bando, N. Funata, T. Isoyama, A case of solid pseudopapillary neoplasm of the pancreas and tumor doubling time. *Pancreatol.* 2 (2002) 495–498.