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A Case of Solid Pseudopapillary Neoplasm Spontaneously Ruptured Into the Duodenum: Case Report

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There have been no reports of ruptured solid pseudopapillary neoplasm (SPN) into adjacent organs. A 22-year-old female was referred to our hospital for treatment of a pancreatic head tumor. Computed tomography (CT) examination at our hospital showed a 5-cm tumor containing air, although CT at a previous hospital revealed an 8-cm tumor without air. Thus, a spontaneous rupture of the tumor into the duodenum was suspected. Subtotal stomach preserving pancreaticoduodenectomy with combined resection of the portal vein was performed. Contrast radiography of resected specimen showed the medium injected into the tumor leaking out from the 2nd portion of the duodenum. Histologically, the patient was diagnosed as SPN. Microscopic invasion to the portal vein and duodenum were also confirmed. She did not experience any postoperative complications and has remained well without any signs of recurrence during 2 years of follow-up. Although there have been 14 studies reporting ruptured SPN, this is the first report of SPN that spontaneously ruptured into the duodenum. An extremely rare case of SPN of the pancreatic head that spontaneously ruptured into the duodenum was reported.

Key words: Solid pseudopapillary neoplasm – Rupture – Duodenum

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Solid pseudopapillary neoplasm (SPN) of the pancreas is an uncommon pancreatic tumor first described by Frantz[1] in 1959, and was included in the World Health Organization classification of tumors in 1996.[2,3] It represents 1% to 2% of pancreatic neoplasms and 10% to 15% of cystic tumors of the pancreas.[4] Although SPN is known to have a low malignant potential and most patients with SPNs have a favorable prognosis after radical resection, a considerable number of cases with microscopic invasion to adjacent organs including the portal vein (PV), duodenum, spleen, or kidney were reported.[5,6] However, there have been no reports of ruptured SPN into adjacent organs. Herein, we report a rare case of SPN that spontaneously ruptured into the duodenum and was successfully resected by pancreaticoduodenectomy combined with PV resection.

Case Presentation

The patient has given permission to publish her case. The patient was a 22-year-old female who visited another hospital in November 2012 with a complaint of upper abdominal and back pain. Abdominal computed tomography (CT) showed a well-defined heterogeneous 8-cm mass with calcification in the pancreatic head. Duodenum and PV were distorted by the tumor. Dilatation of the main pancreatic duct was also observed (Figs. 1A and 1B).

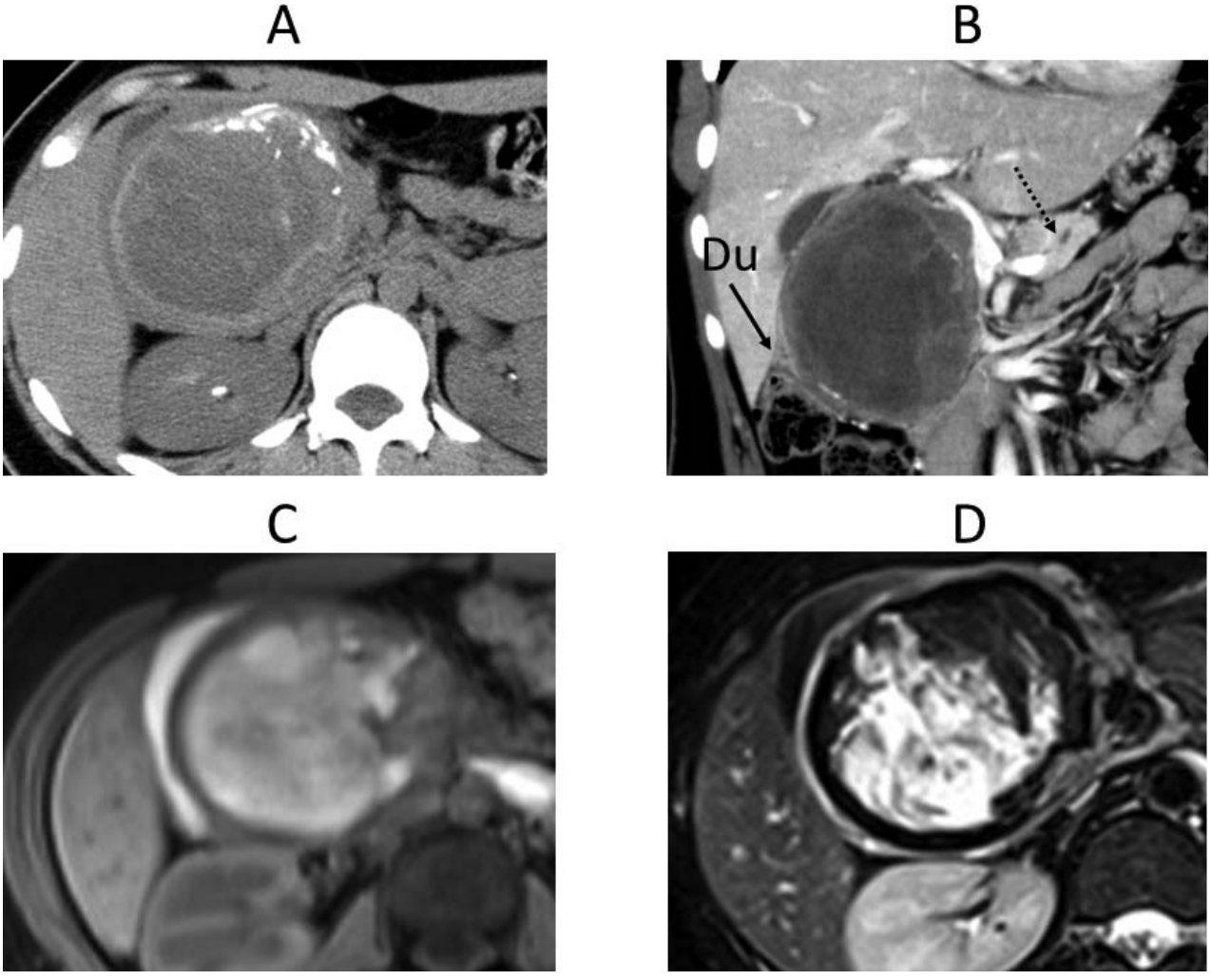

Fig. 1 (A) Nonenhanced CT examined at a previous hospital showed a well-defined heterogeneous 8-cm mass with calcification in the pancreatic head. (B) The duodenum (arrow) and portal vein were distorted by the tumor. Dilatation of the main pancreatic duct (dotted arrow) was also revealed by contrasted CT. (C, D) MRI revealed a mass encapsulated with fibrous membrane containing a solid part and various intensities of hemorrhage. (C) T1-weighted image. (D) T2-weighted image.

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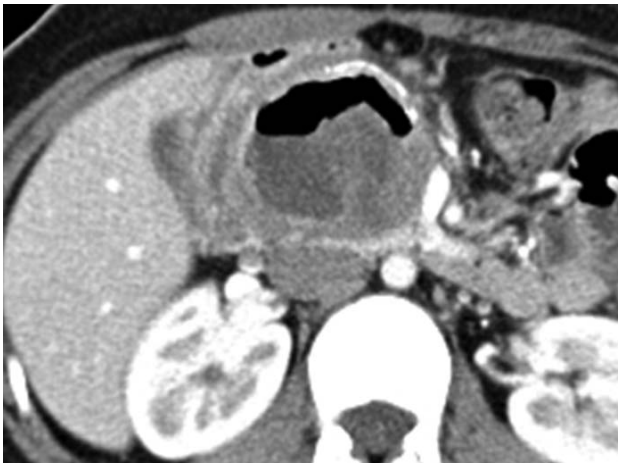

Fig. 2 Findings from CT examination at our hospital. The tumor evidently reduced in size to 5 cm and contained air.

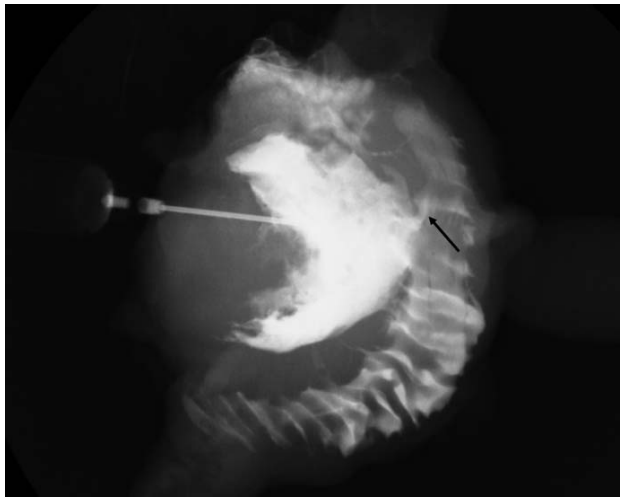

Fig. 3 Findings of contrast radiography of resected specimen. Contrast medium injected into the tumor leaked out from the 2nd portion of duodenum (arrow).

Magnetic resonance imaging (MRI) revealed a mass that was encapsulated by a fibrous membrane containing a solid part and various intensities of hemorrhage (Figs. 1C and 1D). The patient was referred to our hospital for surgical treatment in December. At that time, her body temperature was 37.6°. Laboratory tests showed an abnormal elevation of C-reactive protein to 16.5 mg/dL. To investigate the inflammation's origin, a re-examination of another abdominal CT was performed at our hospital. The results of CT indicated that her inflammation was due to tumor rupture into the duodenum, because the mass was evidently reduced in size to 5 cm and contained air (Fig. 2). Since she had no history of blunt abdominal trauma, the tumor was considered to have ruptured spontaneously. Endoscopic ultrasound-guided fine-needle aspiration was performed for histologic diagnosis. From the results, she was diagnosed with pathologic solid pseudopapillary neoplasm. She underwent operation in January 2013. On laparotomy, the tumor invaded the superior mesenteric vein and duodenum. Therefore, subtotal stomach preserving pancreaticoduodenectomy with combined resection of PV was performed. First, a wedge resection of PV was performed. However, since the intraoperative pathologic examination revealed the PV wall margin to be positive for SPN cells, an additional segmental resection of PV was done. The procedure took 458 minutes and involved 740 mL of blood loss. The resected specimen showed an 8 × 5.5-cm tumor in the pancreatic head. Before opening the duodenum, contrast radiography of the resected specimen was performed. The contrast medium injected into the tumor leaked out from the 2nd portion of the duodenum (Fig. 3). The gross appearance of the cut surface revealed an encapsulated round tumor composed of a solid area and hemorrhage. The cystic part of the tumor mostly existed close to the duodenum while the solid part existed at the peritoneal side (Fig. 4A). Histologically, the tumor had small uniform neoplastic epithelial cells growing with papillary structures and myxedematous stroma compatible with the diagnosis of SPN (Fig. 4B). Microscopic invasion to the portal vein (Fig. 4C) was also observed. An immunohistochemical staining study showed positivity for β-catenin and focally demonstrated positivity for synaptophysin. The labelling index of Ki-67 was <1%. The patient was discharged from the hospital 13 days after the operation without any complications. She has remained well without any signs of recurrence for over 2 years of follow up.

Discussion

Ruptured SPN was first reported by Bombí *et al*[7] in 1983. They treated a case of SPN presenting with acute abdominal pain and hemoperitoneum. From this literature onward, and to the best of our knowledge, there have been 16 case reports of ruptured SPN written in English, including ours. Among these, tumor rupture occurred at a primary lesion in 14,[7–19] and at a recurrent lesion (lymph node, liver metastasis) in 2 cases.[20,21] We summarized the clinical characteristics of 14 cases of

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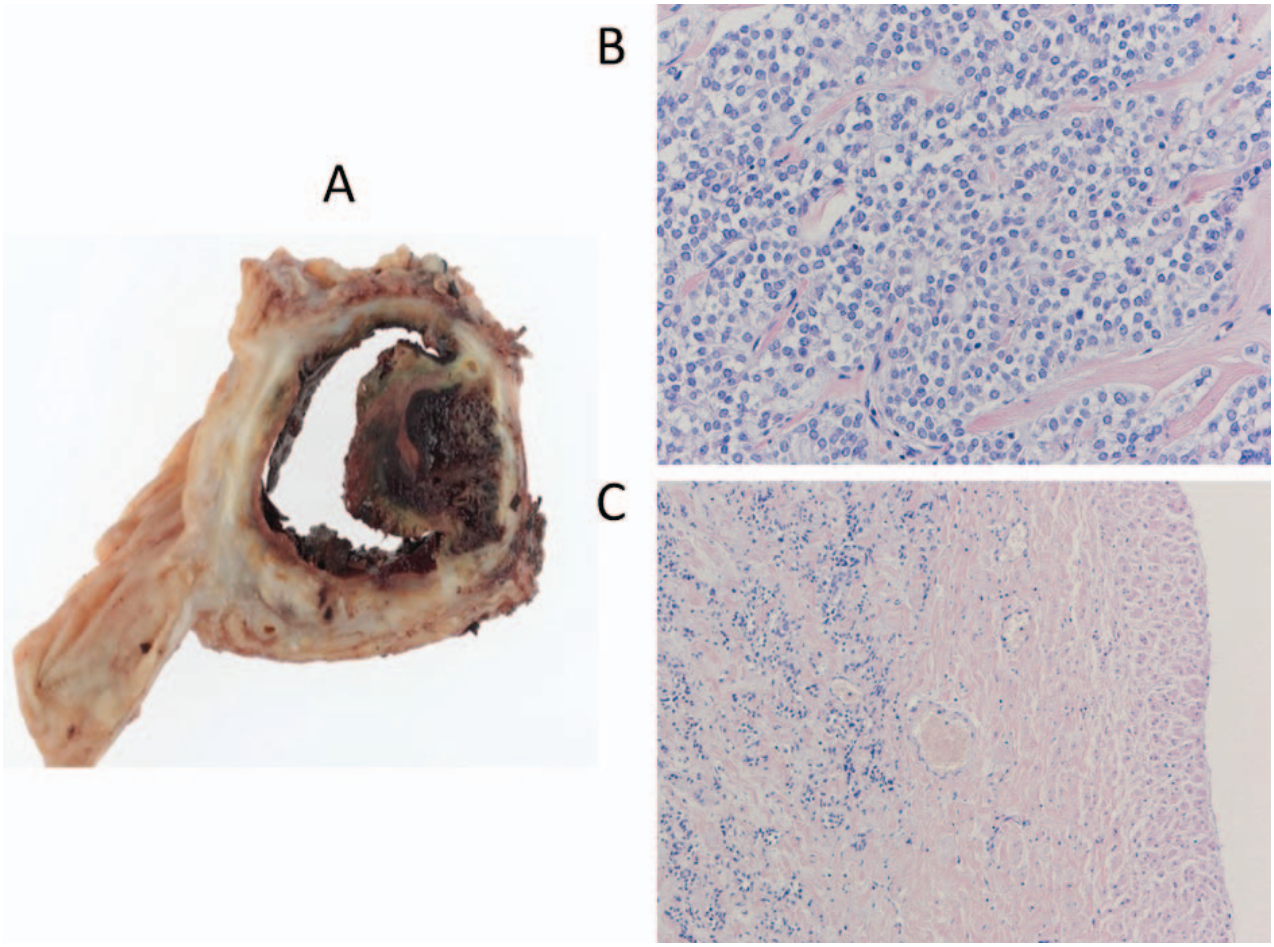

Fig. 4 (A) The gross appearance of the cut surface revealed an encapsulated round tumor composed of a solid area and hemorrhage. Most of the cystic part of the tumor was close to the duodenum while the solid part was near the peritoneal side. (B) Histologically, the tumor had small uniform neoplastic epithelial cells growing with papillary structures and myxedematous stroma compatible with the diagnosis of SPN [hematoxylin and eosin (H&E), high magnification]. (C) Microscopic invasion to the portal vein was observed (H&E, low magnification).

ruptured SPN at a primary lesion (Table 1). Among these, SPN rupturing into an adjacent organ was only seen in our case. Although the chief complaint was pain or shock due to hemoperitoneum in most reported cases, our patient had no symptom except for low-grade fever. Most likely this is because the contents of SPN tumor had not leaked into the peritoneal cavity. Spontaneous rupture was observed in 8 while traumatic rupture was seen in the remaining 6. The tumor was located at the pancreatic head in 3 cases and at the distal part, including pancreatic body or tail in 11 cases. In so far as the 8 cases that spontaneously ruptured, the tumor was located at a distal part in 7 cases while at the head only in our case. Thus, our case was extremely rare in 2 respects. First, SPN was ruptured into the duodenum, not into peritoneal cavity. Second, the spontaneously ruptured SPN was located at the pancreatic head. The median diameter of the tumors was 11.0 cm (range: 4–17 cm). Apparently, it was larger than the tumors reported in a thorough review, which revealed that the mean diameter of SPN was 6.08 cm (range: 0.5–34.5 cm).[22] This might be associated with a correlation between SPN size and tumor aggressiveness, which was demonstrated by Kang *et al*.[23]

Our case should be distinguished from other ruptured SPN cases regarding one more point: the involvement of adjacent organs including the duodenum and portal vein. Microscopically, SPN cells are known to delicately infiltrate into the surrounding pancreatic tissue, entrapping acinar

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Table 1 Reported 14 cases of ruptured SPN at primary lesion

Case no.	Author	Year	Age	Sex	Symptom	Cause of rupture	Tumor location	Size, cm	Involved other organs	Outcome[a]
1	Bombi[7]	1984	22	F	Sudden pain	Spontaneous	Body	12	None	2Y, A, NED
2	Lieber[8]	1987	13	F	Sudden pain	Trauma	Body	9	None	2Y, A, NED
3	Todani[9]	1988	16	F	Pain, anemia	Spontaneous	Tail	9	None	5Y, A, NED
4	Hernandez[10]	1989	22	F	Pain, fever, vomit	Spontaneous	Tail	16	Peritoneum	12M, A, unmeasurable rec
5	Sanchez[11]	1990	13	F	Pain, shock	Trauma	Distal part	10	None	4Y A, NED
6	Jeng[12]	1993	26	F	Pain, shock	Spontaneous	Body	13	None	66M, A, NED
7	Panieri[13]	1998	34	F	Severe pain	Spontaneous	Body	12	None	16 day, D
8	Potrc[14]	2004	14	M	Pain, shock	Trauma	Head	9	None	3Y, A, NED
9	Huang[15]	2005	19	F	Pain, shock	Trauma	Tail	8	None	6M, A
10	Tajima[16]	2012	12	F	Pain	Trauma	Head	14	None	7Y, A, peritoneal, LN rec
11	Takamatsu[17]	2013	13	F	Sudden pain	Spontaneous	Tail	4	None	2Y, A, NED
12	Huang[18]	2013	29	F	Sudden pain	Spontaneous	Body	17	None	8M, A, NED
13	Park[19]	2014	12	F	Unknown	Trauma	Body	15	None	LM→RFA, 97M, A, NED
14	Current	2015	22	F	Low-grade fever	Spontaneous	Head	8	Duodenum, portal vein	22M, A, NED

A, alive; D, dead; LM, liver metastasis; LN, lymph node; M, month; NED, no evidence of disease; Outcome, prognosis after surgery; rec, recurrence; RFA, radiofrequency ablation; Y, year.

cells and islets, although this tumor was grossly well demarcated from the normal pancreas.[3] However, it is also known that vascular invasion is rare. Papavramidis[22] reported that invasion to portal vein was observed in only 26 of 497 patients with data available regarding metastases or invasion. Cheng *et al*[5] demonstrated 10 cases of adjacent organ (SMV/PV or left kidney) resections for SPN not leading to postoperative mortality. They showed that 9 patients with microscopically negative margins survived without any recurrence and metastasis while only 1 patient with a microscopically positive margin developed liver metastasis. They concluded that en bloc synchronous adjacent organ resection should be applied. Additional segmental resection of PV following wedge resection for positive margins in our case was proved valid based on this standpoint.

There are 2 clinical questions about the etiology of our special case. First, why did SPN of our patient rupture spontaneously? Since there have been only 8 case reports of spontaneously ruptured SPN, little is known about its pathogenesis. Takamatsu *et al*[17] reported that SPN had a natural tendency to hemorrhage because the cystic part of it consisted of the degeneration following the intramural hemorrhage. Therefore, they concluded that the spontaneous rupture resulted from both abrupt massive hemorrhage and the increased pressure in the tumor. Second, why did it rupture into the duodenum and not into the peritoneal cavity as with all of other reported cases? It might have been because most of the cystic part of the tumor existed close to the duodenum while the solid part existed on the peritoneal side. Moreover, the wall of the duodenum might have been fragile due to tumor invasion. From the above, we considered that the pressure in the SPN had increased, perhaps due to sudden hemorrhage. Fortunately, the cystic part of the tumor located adjacent to the duodenum subsequently ruptured, breaking the weak wall of the duodenum.

Little is known about the prognosis of ruptured SPN. Kim *et al*[24] asserted that the tumor rupture might be a risk factor for recurrence after surgery for SPN. Indeed, recurrence was observed in 3 (21.4%) out of 14 ruptured SPNs. It was a higher incidence than that found in previously reported unruptured and completely resected SPNs.[24,25] On the other hand, peritoneal recurrence was evident in only 1 of 14 patients. Thus, the cause of the highly recurrent incidence of ruptured SPN might be associated with potential tumor aggressiveness rather than perito-

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neal dissemination. As for our patient, although the leaking of SPN contents into the peritoneal cavity was not developed, we still must pay close attention for recurrence. It is possible that the SPN cells had an aggressive nature because they invaded adjacent organs (duodenum and portal vein) and ruptured spontaneously.

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Conflicts of interest: The authors declare that they have no competing interests.

References

1. Frantz VK. Tumors of the pancreas. In: *Anonymous Atlas of Tumor Pathology*, Section 7, Fascicles 27 and 28. Washington, DC: Armed Forces Institute of Pathology, 1959:32–33
2. Kloppel G, Solcia E, Longnecker DS, Capella C, Sobin LH. Histological typing of tumors of the exocrine pancreas. In: World Health Organization. *International Histological Classification of Tumors*, 2nd ed. New York, NY: Springer, 1996:15–21
3. Kloppel G, Hruban RH, Klimstra DS, Maritra A, Morohoshi T, Notohara K *et al*. Solid-pseudopapillary neoplasm of the pancreas. In: Bosman FT, Carneiro F, Hruban RH, Theise ND, eds. *WHO Classification of Tumors of the Digestive System*. Lyon, France: International Agency for Research on Cancer, 2010: 327–30
4. Morikawa T, Onogawa T, Maeda S, Takadate T, Shirasaki K, Yoshida H *et al*. Solid pseudopapillary neoplasms of the pancreas: an 18-year experience at a single Japanese institution. *Surg Today* 2013;**43**(1):26–32
5. Cheng K, Shen B, Peng C, Yuan F, Yin Q. Synchronous portal-superior mesenteric vein or adjacent organ resection for solid pseudopapillary neoplasms of the pancreas: a single-institution experience. *Am Surg* 2013;**79**(5):534–539
6. Kim MJ, Choi DW, Choi SH, Heo JS, Sung Jy. Surgical treatment of solid pseudopapillary neoplasms of the pancreas and risk factors for malignancy. *Br J Surg* 2014;**101**(10):1266–1271
7. Bombí JA, Milla A, Badal JM, Piulachs J, Estape J, Cardesa A. Papillary-cystic neoplasm of the pancreas. Report of two cases and review of the literature. *Cancer* 1984;**54**(4):780–784
8. Lieber MR, Lack EE, Roberts JR Jr, Merino MJ, Patterson K, Restrepo C *et al*. Solid and papillary epithelial neoplasm of the pancreas. An ultrastructural and immunocytochemical study of six cases. *Am J Surg Pathol* 1987;**11**(2):85–93
9. Todani T, Shimada K, Watanabe Y, Toki A, Fujii T, Urushihara N *et al*. Frantz's tumor: a papillary and cystic tumor of the pancreas in girls. *J Pediatr Surg* 1988;**23**(2):116–121
10. Hernandez-Maldonado JJ, Rodriguez-Bigas MA, Gonzalez de Pesante A, Vazquez-Quintana E. Papillary cystic neoplasm of the pancreas. A report of a case presenting with carcinomatosis. *Am Surg* 1989;**55**(9):552–559
11. Sanchez JA, Newman KD, Eichelberger MR, Nauta RJ. The papillary-cystic neoplasm of the pancreas. An increasingly recognized clinicopathologic entity. *Arch Surg* 1990;**125**(11): 1502–1505
12. Jeng LB, Chen MF, Tang RP. Solid and papillary neoplasm of the pancreas. Emphasis on surgical treatment. *Arch Surg* 1993; **128**(4):433–436
13. Panieri E, Krige JE, Bornman PC, Graham SM, Terblanche J, Cruse JP. Operative management of papillary cystic neoplasms of the pancreas. *J Am Coll Surg* 1998;**186**(3):319–324
14. Potrc S, Kavalar R, Ivanecz A, Gadzijev EM. Acute presentation of a solid pseudopapillary tumor of the pancreas. *Wien Klin Wochenschr* 2004;**116**(2):100–103
15. Huang HL, Shih SC, Chang WH, Wang TE, Chen MJ, Chan YJ. Solid-pseudopapillary tumor of the pancreas: clinical experience and literature review. *World J Gastroenterol* 2005;**11**(9): 1403–1409
16. Tajima Y, Kohara N, Maeda J, Inoue K, Kitasato A, Nastsuda K *et al*. Peritoneal and nodal recurrence 7 years after the excision of a ruptured solid pseudopapillary neoplasm of the pancreas: report of a case. *Surg Today* 2012;42(8):776–780
17. Takamatsu S, Nagano H, Ohtsukasa S, Kawachi Y, Maruyama H. A case of spontaneous ruptured solid pseudopapillary tumor of pancreas resected by laparoscopic surgery. *Case Rep Med* 2013;2013:953240
18. Huang SC, Wu TH, Chen CC, Chen TC. Spontaneous rupture of solid pseudopapillary neoplasm of the pancreas during pregnancy. *Obstet Gynecol* 2013;**121**(2):486–488
19. Park JY, Kim SG, Park J. Solid pseudopapillary tumor of the pancreas in children: 15-year experience at a single institution with assays using an immunohistochemical panel. *Ann Surg Treat Res* 2014;**86**(3):130–135
20. Stömmer P, Kraus J, Stolte M, Giedl J. Solid and cystic pancreatic tumors. Clinical, histochemical, and electron microscopic features in ten cases. *Cancer* 1991;**67**(6):1635–1641
21. Sclafani LM, Reuter VE, Coit DG, Brennan MF. The malignant nature of papillary and cystic neoplasm of the pancreas. *Cancer* 1991;**68**(1):153–158
22. Papavramidis T, Papavramidis S. Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. *J Am Coll Surg* 2005;**200**(6):965–972

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23. Kang CM, Kim KS, Choi JS, Kim H, Lee WJ, Kim BR. Solid pseudopapillary tumor of the pancreas suggesting malignant potential. *Pancreas* 2006;**32**(3):276–280
24. Kim CW, Han DJ, Kim J, Kim YH, Park JB, Kim SC. Solid pseudopapillary tumor of the pancreas: Can malignancy be predicted? *Surgery* 2011;**149**(5):625–634
25. Reddy S, Cameron JL, Scudiere J, Hruban RH, Fishman EK, Ahuja N *et al*. Surgical management of solid-pseudopapillary neoplasms of the pancreas (Franz or Hamoudi tumors): a large single-institutional series. *J Am Coll Surg* 2009;**208**(5): 950–957

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