CASE REPORT

Solid pseudopapillary tumour (Frantz's tumour) of the pancreas in childhood

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SUMMARY

An 11-year-old girl presented with acute pancreatitis and mass in the head of the pancreas. MRI revealed a heterogeneous right-upper quadrant retroperitoneal mass measuring 6.8×6.1×5.5 cm arising from the pancreatic head. Endoscopic ultrasound with fine-needle aspirate revealed a solid pseudopapillary tumour (SPT) of the pancreas. The patient underwent a pylorus-preserving Whipple procedure. Pathology confirmed SPT. First described by Frantz, SPT represents less than 3% of all exocrine tumours. It is especially rare in children and shows different clinical features compared with adults. In our patient, tumour cells were arranged at the periphery of fibrovascular cores, but they did not show definite gland formation, keratinisation or cytoplasmic pigment accumulation. A periodic acid-Schiff stain without diastase did not show appreciable glycogen within the tumour cells, classic for Frantz's tumour. The literature, diagnosis, management and pathogenesis on this rare entity in children are reviewed and discussed.

BACKGROUND

Solid pseudopapillary tumour (SPT) of the pancreas is an uncommon neoplasm of low malignant potential.¹ First described by Virginia Kneeland Frantz,² many names have been used to describe this tumour, including papillary cystic tumour, solid-and-papillary tumour and Frantz's tumour.¹⁻³ In 1996, the WHO simplified the classification and nomenclature, resulting in SPT as the accepted terminology.³ *It is especially rare in children and shows different clinical features compared with adults.* The literature, diagnosis, management and pathogenesis on this rare entity in children are reviewed and discussed.

CASE PRESENTATION

An 11-year-old girl presented with vomiting and right-upper quadrant abdominal pain. The history revealed 8 months of chronic abdominal pain and progressive weight loss. Examination revealed a weight-for-age below the fifth centile and a flat, soft abdomen; mild epigastric tenderness; but no palpable mass. Lipase was elevated at 1829 IU/L, liver enzymes and hepatic function were essentially normal and tumour markers α -fetoprotein, cancer antigen 19-9, carcinoembryonic antigen and human chorionic gonadotropin were within normal range.

INVESTIGATIONS

A CT scan revealed a 6.4×5.9×5.2 cm right-upper quadrant, well-circumscribed heterogeneous mass of uncertain origin, without biliary or pancreatic duct dilation (figure 1). MRI demonstrated a

6.8×6.1×5.5 cm retroperitoneal, well-circumscribed heterogeneous mass centred in the region of the pancreatic head, with mild pancreatic ductal dilation and normal common bile duct, associated with medial displacement of the portal vein and posterolateral displacement of the second portion of the duodenum. Endoscopic ultrasound with fine-needle aspiration (EUS-FNA) described a well-circumscribed hyperechoic mass, at least 5.9 cm in diameter, in the head of the pancreas and mild pancreatic duct dilation at 2.4 mm, with no significant internal vascular flow, calcifications, or invasion of vascular structures. The pathology was consistent with SPT of the pancreas. Plans for operative intervention with pancreaticoduodenectomy (Whipple) were made, although the patient suffered from persistent pancreatitis.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses include a variety of pancreatic tumours including non-functioning islet tumour, pancreatoblastoma, acinar cell cancer, mucinous cystic neoplasm, serous cystadenomas, lymphoma and pseudocyst. The only way to confirm diagnosis is biopsy or excision, and each has radically different outcomes.

TREATMENT

A pylorus-preserving pancreaticoduodenectomy was performed. Of note, the pancreatic and biliary anastomoses were created to a roux-en-Y jejunal limb to try to prevent a long-term bile reflux. A mucosal-mucosal anastomosis was created for the pancreaticojejunostomy. Closed suction drains were left adjacent to each of these anastomoses. A pylorus-preserving method was chosen to prevent postgastrectomy syndromes. The procedure was technically challenging because of persistent pancreatic inflammation. The splenic vein was avulsed off the junction with the superior mesenteric vein (SMV) necessitating ligation of the splenic vein and direct repair of the SMV/portal vein. The patient was transfused 20 mL/kg of packed red blood cells and 10 mL/kg of fresh frozen plasma. Pathology confirmed the diagnosis. The tumour cells were arranged at the periphery of fibrovascular cores but did not show definite gland formation, keratinisation or cytoplasmic pigment accumulation. A periodic acid-Schiff (PAS) stain without diastase did not show appreciable glycogen within the tumour cells, classic for Frantz's tumour.

OUTCOME AND FOLLOW-UP

Postoperatively the patient did well. She was discharged home on postoperative day (POD) 17 on a

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Figure 1 Cross-section CT of the abdomen and pelvis with contrast showing pancreatic head mass.

low-fat diet. No pancreatic fistula developed, and all drains were removed prior to discharge. On POD 45 she was advanced to a regular diet. She has had no evidence of recurrent pancreatitis or tumour on CT 1 year later and MRI 2 years later. She was doing well at her 2-year follow-up.

Table 1 Clinical data, ranging from 53 to 183 cases, of solid pseudopapillary tumour of the pancreas from the paediatric literature^{6 7 9–36}

Category	Findings
Age (years) [SD]	Mean: 13.1 [2.4], range: 8–18
Sex	Male: 25 (19.2%) Female: 105 (80.8%)
Presentation	Abdominal pain: 88 (48.1%) Palpable mass: 46 (25.1%) Vomiting: 12 (6.6%) Trauma: 13 (7.1%) Dyspepsia: 7 (3.8%) Incidental: 5 (2.7%) Other: 12 (6.6%)
Location	Head, neck and/or body: 58 (45%) Body and/or tail: 68 (52.7%) Unknown: 3 (2.3%)
Size, largest diameter (cm) [SD]	Mean: 9.3 [4.1], range: 1–20
Operation	PPPD: 17 (13%) Whipple: 16 (12.2%) Duodenum-sparing head resection: 6 (4.6%) Distal pancreatectomy: 46 (35.1%) Local resection: 6 (4.6%) Enucleation: 3 (2.3%) Other: 6 (4.6%) Unspecified: 31 (23.7%)
Metastasis	4 (3.1%)
Follow-up (months) [SD]	Mean: 62.7 [58.4], range: 6–240
Recurrence	10 (8.7%)
Time to recurrence (months) [SD]	Mean: 43.5 [33.1], range: 0–96
Mortality	1
PPPD, pylorus-preserving pancreaticoduodenectomy.	

DISCUSSION

Two and a half per cent of all primary pancreatic tumours are SPT.⁴ SPT is considered rare in children, but the true incidence is difficult to discern. This may be in part due to the plethora of nomenclature prior to 1996. In a review of 58 cases of pancreatic tumours identified in the National Cancer Institute's Surveillance Epidemiology and End Results database, which provides information on cancer statistics, Perez *et al*⁵ reported the age-adjusted incidence of SPT in 2000 as 0.005/100 000 (ages 0–19), representing 17.2% of their cohort. Zhou *et al*⁶ reported 0.01 case/100 000 population/year from a referral base of two million patients. Lam and Zhou each reported a 20% incidence of SPT in children in their respective case series.^{4 6} Interestingly Zhou (Hong Kong) and Jaksic (Toronto) reported that 50% of all paediatric pancreatic cases were SPT.^{6 7}

SPT is usually seen in young women in their early 20s.⁸ In children and adolescents, the average age is 13.⁶ Our review of the paediatric population with SPT reached a similar conclusion. An extensive summary of the literature of paediatric tumours is presented in table 1.^{6 7 9–36}

The female:male ratio is 10:1¹⁴ with the incidence reported in females of 86% in adults and children.⁶ Historically, abdominal pain was the most common presentation of SPT.⁸ Recent reviews of the paediatric literature, however, have reported a palpable abdominal mass as the most common presentation in children. Abdominal pain, vomiting, post-traumatic and jaundice followed.^{6 20} Diagnosis from incidental imaging obtained for reasons other than suspected SPT was exceedingly rare in children^{6 20}; however, this was the most common reason for diagnosis among the adults.²⁰ Although pancreatitis was not specifically mentioned, elevated levels of amylase and lipase have been reported.^{6 25 27 35} Our patient presented with acute pancreatitis and non-palpable abdominal mass.

In children, SPT most commonly presents in the head of the pancreas (60–70%) in contrast to adults where the majority occur in the body or tail of the pancreas (80%).^{6 20} Unfortunately, preoperative laboratories and imaging are not specific for SPT. Ultrasound, CT and MRI reveal a well-demarcated mass ranging from mostly cystic to mostly solid.^{6 7 37} Imaging findings are detailed in box 1.

EUS-FNA to solidify the preoperative diagnosis and plan for appropriate treatment are well reported in adults.^{41–43} Nadler *et al*²⁴ report the successful preoperative diagnosis of SPT by use of EUS-FNA, but note that only a few paediatric gastroenterologists have experience with this procedure. An adult gastroenterologist was consulted to perform this procedure in our patient. Prior to this report, only 3 of 11 paediatric patients were successfully diagnosed preoperatively with SPT.^{42 44–52} Differential diagnoses include a non-functioning islet tumour,

Box 1 Imaging findings in solid pseudopapillary tumour.

- ▶ CT scan may show calcification in the tumour.⁷
- ▶ MRI may have areas of the following in the tumour:
 - High signal intensity corresponding to foci of haemorrhagic necrosis or debris in T1-weighted images⁶
 - Signal intensity ranges from very low to high in T2-weighted images.³⁸
- ▶ Angiography may show little or no blood supply in the tumour.^{39 40}

pancreatoblastoma, acinar cell cancer, mucinous cystic neoplasm, serous cystadenomas, lymphoma and pseudocyst.^{7 14 50 53 54} In this setting, an EUS-FNA was performed to rule out lymphoma—a Whipple is contraindicated for this diagnosis.

SPT has been shown to be of low malignant potential with few recurrences if completely excised. Although many surgical approaches have been described, the approach of choice is most dependent on location of the tumour and involvement of surrounding structures. Since pancreatic surgery is not without morbidity, Campanile *et al* questioned whether aggressive surgery was justified in young children. By reviewing data on 11 patients who had incomplete resections, they found a median survival rate of 5.7 years (69.5 months), which they deemed unacceptable in children and suggested that complete resection of the tumour was justified even if it required a 'mutilating' surgery.⁵⁵ Unfortunately, pancreaticoduodenectomy (the Whipple procedure) is often felt to be 'too big' of a surgery for small children. Grosfeld *et al*¹⁷ addressed this question directly by stating that a paediatric surgeon should be equipped to perform this surgery if so indicated. The strategy devised for our paediatric patient differed from that typical in the adult population. Our patient underwent a pylorus-preserving Whipple procedure to avoid dumping and diarrhoea. A roux-en-Y pancreaticojejunostomy and hepaticojejunostomy were performed to prevent a long-term primary duodenogastric bile reflux gastritis, similar to the management of choledochal cysts in children.⁵⁶ No complications were noted 2 years postoperatively.

SPT is an exocrine pancreatic tumour that has an unclear origin. Theories of pathophysiology for SPT include

1. Arising from centroacinar cells since it has matching immunohistochemical patterns^{57–59};
2. Originating from pancreatic duct cells^{44 60};
3. Arising from pluripotential embryonic pancreatic stem cells.^{61–63}

Since the majority of tumour cells stain positive for β -catenin in the cytoplasm and nucleus, it has been suggested that the Wnt signalling pathway is responsible for its growth.⁶⁴ Box 2 details the pathological findings of SPT. As noted by Grosfeld *et al*,¹⁷ SPT is differentiated from acinar cell tumours by the absence of PAS-positive cytoplasmic granules, ultrastructural

zymogen and prezymogen granules. Our patient's pathology mirrored these classic findings.

Paediatric patients with SPT fare better than adults. According to one review of 83 patients with SPT (children and adults), no patient under 20 years of age developed metastases or died, but in those >20 years of age, 16% (40/255) had identifiable metastases and 1.2% (3/255) died from the tumour.⁶⁵ In a review of *paediatric* patients with SPT, 1 of 78 had metastases and 11/78 had contiguous organ invasion by the tumour.⁶ Survival at 5 years was 21/78 patients and survival at 10 years was 10 patients. Overall mortality was 1% (1/78) from distant metastases, and recurrence rate was 8%. Risk factors for recurrence included older age and contiguous organ invasion.⁶ In the malignant or locally aggressive cases the prognosis was good, as local recurrence and metastasis are rare.

Unfortunately, in a large case series comparing children with adults by Lee *et al*,²⁰ no factor was identified that was predictive of the malignant potential of an SPT. Regardless, when evaluating *all* patients with SPT, surgical resection is the mainstay of therapy with overall 5-year survival greater than 95% (even in the presence of metastases).¹⁴ Fried *et al*⁶⁶ described radiotherapy for unresectable SPT. Of 78 children with SPTs, only two received radiotherapy and chemotherapy postoperatively.⁶⁶ The role of neoadjuvant or adjuvant therapy is speculative at best. Recently, Sugito *et al*³² reported the successful utilisation of radiofrequency ablation to shrink an unresectable SPT, which in turn led to a successful complete resection 24 months later. Successful treatment of early recurrence, with resection and/or adjuvant chemotherapy, has also been reported.^{10 13 18–20 25 26 36 49 65} Given the success in treating recurrences, and the role surveillance imaging has played in identifying the recurrence,^{10 13 18 65} paediatric patients with SPT should have close follow-up. We recommend serial CT or MRI every 3 months for the first year spacing out to 6 months during the second year and yearly thereafter for a total of 5 years.

Box 2 Pathological findings in solid pseudopapillary tumour

- Gross
 - Tumour is oval and well encapsulated.
 - Tumour grows exophytically.
 - Cut surface reveals
 - ▷ A solid area around the periphery
 - ▷ Cystic degeneration in the centre
 - May be haemorrhagic and necrotic
 - ▷ Central calcifications occasionally.⁴⁹
- Histology—a mixture of papillary and solid/cystic patterns.⁴
 - Papillary
 - ▷ The tumour cells have an eosinophilic cytoplasm.
 - ▷ They are uniform in size.
 - ▷ They are arranged around a central fibrovascular stalk.
 - The cystic area is composed of sheets of cells with variable degeneration.

Learning points

- In children, solid pseudopapillary tumour (SPT) most commonly presents in the head of the pancreas (60–70%) in contrast to adults where the majority occur in the body or tail of the pancreas (80%).
- In comparison with the adult population, children with SPT generally have a better prognosis.
- SPT has been shown to be of low malignant potential with few recurrences if completely excised. Although many surgical approaches have been described, the approach of choice is most dependent on location of the tumour and involvement of surrounding structures.
- A Whipple procedure is safe in children. The strategy devised for our paediatric patient differed from that typical in the adult population. Our patient underwent a pylorus-preserving Whipple procedure to avoid dumping and diarrhoea. A roux-en-Y pancreaticojejunostomy and hepaticojejunostomy were performed to prevent a long-term primary duodenogastric bile reflux gastritis and oesophagitis, similar to the management of choledochal cysts in children.

Contributors All the authors contributed to conception and design, or analysis and interpretation of the data; drafting the article or revising it critically for important intellectual content and final approval of the version to be published.

Competing interests None.

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REFERENCES

- Yu PF, Hu ZH, Wang XB, *et al.* Solid pseudopapillary tumor of the pancreas: a review of 553 cases in Chinese literature. *World J Gastroenterol* 2010;16:1209–14.
- Frantz VK. ed. Tumors of the pancreas. In: *Atlas of tumor pathology, 1st series, fascicle 27–28*. Washington, DC: US Armed Forces Institute of Pathology, 1959:32–3.
- Kloppel G, Solcia E, Longnecker DS, *et al.* Histological typing of tumors of the exocrine pancreas. In: Kloppel G, Solcia E, Longnecker DS, Capella C, Sobin LH, eds. *World Health Organization international histological classification of tumors*. 2nd edn. Berlin: Springer, 1996:120–8.
- Lam KY, Lo CY, Fan ST. Pancreatic solid-cystic-papillary tumor: clinicopathologic features in eight patients from Hong Kong and review of the literature. *World J Surg* 1999;23:1045–50.
- Perez EA, Gutierrez JC, Koniaris LG, *et al.* Malignant pancreatic tumors: incidence and outcome in 58 pediatric patients. *J Pediatr Surg* 2009;44:197–203.
- Zhou H, Cheng W, Lam KY, *et al.* Solid-cystic papillary tumor of the pancreas in children. *Pediatr Surg Int* 2001;17:614–20.
- Jaksic T, Yaman M, Thorner P, *et al.* A 20-year review of pediatric pancreatic tumors. *J Pediatr Surg* 1992;27:1315–17.
- Papavramidis T, Papavramidis P. Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. *J Am Coll Surg* 2005;200:965–72.
- Al-Qahtani S, Gudinchet F, Laswed T, *et al.* Solid pseudopapillary tumor of the pancreas in children: typical radiological findings and pathological correlation. *Clin Imaging* 2010;34:152–6.
- Andronikou S, Moon A, Ussher R. Peritoneal metastatic disease in a child after excision of a solid pseudopapillary tumour of the pancreas: a unique case. *Pediatr Radiol* 2003;33:269–71.
- Casanova M, Collini P, Ferrari A, *et al.* Solid-pseudopapillary tumor of the pancreas (Frantz tumor) in children. *Med Pediatr Oncol* 2003;41:74–6.
- Chao HC, Kong MS, Lin SH, *et al.* Papillary cystic neoplasm of the pancreas in children: report of three cases. *Acta Pediatr Taiwan* 2000;41:101–5.
- Choi SH, Kim SM, Oh JT, *et al.* Solid pseudopapillary tumor of the pancreas: a multicenter study of 23 pancreatic cases. *J Pediatr Surg* 2006;41:1992–5.
- Crucitti A, Grossi U, Giustacchini P, *et al.* Solid pseudopapillary tumor of the pancreas in children: report of a case and review of the literature. *Updates Surg* 2010;62:69–72.
- Dall'igna P, Cecchetto G, Bisogno G, *et al.* Pancreatic tumors in children and adolescents: the Italian TREP project experience. *Pediatr Blood Cancer* 2010;54:675–80.
- Faraj W, Jamali F, Khalifeh M, *et al.* Solid pseudopapillary neoplasm of the pancreas in a 12-year-old female: case report and review of the literature. *Eur J Pediatr Surg* 2006;16:358–61.
- Grosfeld JL, Vane DW, Rescorla FJ, *et al.* Pancreatic tumors in childhood: analysis of 13 cases. Presented at the 38th Annual Meeting of the Surgical Section of the American Academy of Pediatrics. Chicago, Illinois, 21–23 October 1989.
- Hassan I, Celik I, Nies C, *et al.* Successful treatment of solid-pseudopapillary tumor of the pancreas with multiple liver metastases. *Pancreatol* 2005;5:289–94.
- Lack EE, Cassady JR, Levey R, *et al.* Tumors of the exocrine pancreas in children and adolescents: a clinical and pathologic study of eight cases. *Am J Surg Pathol* 1983;7:319–27.
- Lee SE, Jang JY, Hwang DW, *et al.* Clinical features and outcome of solid pseudopapillary neoplasm: differences between adults and children. *Arch Surg* 2008;143:1218–21.
- Melotti G, Cavallini A, Butturini G, *et al.* Laparoscopic distal pancreatectomy in children: case report and review of the literature. *Ann Surg Oncol* 2007;14:1065–9.
- Meshikhes AW, Atassi R. Pancreatic pseudopapillary tumor in a male child. *JOP* 2004;5:505–11.
- Moholkar S, Sebire NJ, Roebuck DJ. Solid-pseudopapillary neoplasm of the pancreas: radiological-pathological correlation. *Pediatr Radiol* 2005;35:819–22.
- Nadler EP, Novikov A, Landzberg BR, *et al.* The use of endoscopic ultrasound in the diagnosis of solid pseudopapillary tumors of the pancreas in children. *J Pediatr Surg* 2002;37:1370–3.
- Park M, Koh KN, Kim BE, *et al.* Pancreatic neoplasms in childhood and adolescence. *J Pediatr Hematol Oncol* 2011;33:295–300.
- Rebhandl W, Felberbauer FX, Puig S, *et al.* Solid-pseudopapillary tumor of the pancreas (Frantz tumor) in children: report of four cases and review of the literature. *J Surg Oncol* 2001;76:289–96.
- Saw HP, Ho ML, Chen JY. Solid cystic pseudopapillary tumor of the pancreas: report of one case. *Acta Pediatr Taiwan* 2003;44:368–71.
- Schroth M, Razeghi S, Jänig U, *et al.* Solid-pseudopapillary tumor of the pancreas. *Med Pediatr Oncol* 1999;33:508–9.
- Shorter NA, Glick RD, Klimstra DS, *et al.* Malignant pancreatic tumors in childhood and adolescents: the Memorial Sloan-Kettering experience, 1967 to present. *J Pediatr Surg* 2002;37:887–92.
- Snajdauf J, Rygl M, Petru O, *et al.* Duodenum-sparing technique of head resection in solid pseudopapillary tumor of the pancreas in children. *Eur J Pediatr Surg* 2009;19:354–7.
- Sokolov YY, Stonogin SV, Donskoy DV, *et al.* Laparoscopic pancreatic resections for solid pseudopapillary tumor in children. *Eur J Pediatr Surg* 2009;19:399–401.
- Sugito K, Kusafuka T, Hoshino M, *et al.* Application of radiofrequency ablation for giant solid pseudopapillary tumor of the pancreas. *Pediatr Int* 2010;52:e29–31.
- Tapia B, Ahrens W, Kenney B, *et al.* Acinar cell carcinoma versus solid pseudopapillary tumor of the pancreas in children: a comparison of two rare and overlapping entities with review of the literature. *Pediatr Dev Pathol* 2008;11:384–90.
- Uchida H, Goto C, Kishimoto H, *et al.* Laparoscopic spleen-preserving distal pancreatectomy for solid pseudopapillary tumor with conservation of splenic vessels in a child. *J Pediatr Surg* 2010;45:1525–9.
- Vergauwen W, Op de Beeck B, Hagendorens M, *et al.* A solid pseudopapillary tumour of the pancreas presenting after an abdominal trauma. *Acta Chir Belg* 2010;110:390–3.
- Yu DC, Kozakewich HP, Perez-Atayde AR, *et al.* Childhood pancreatic tumors: a single institution experience. *J Pediatr Surg* 2009;44:2267–72.
- Poustchi-Amin M, Leonidas JC, Valderrama E, *et al.* Papillary-cystic neoplasm of the pancreas. *Pediatr Radiol* 1995;25:509–11.
- Ohtomo K, Furui S, Onoue M, *et al.* Solid and papillary epithelial neoplasm of the pancreas: MR imaging and pathologic correlation. *Radiology* 1992;184:567–70.
- Kuo TT, Su JJ, Chien CH. Solid and papillary neoplasm of the pancreas. Report of three cases from Taiwan. *Cancer* 1984;54:1469–74.
- Choi BI, Kim KW, Han MC, *et al.* Solid and papillary epithelial neoplasms of the pancreas: CT findings. *Radiology* 1998;166:413–16.
- Bondeson L, Bondeson AG, Genell S, *et al.* Aspiration cytology of a rare solid and papillary epithelial neoplasm of the pancreas. Light and electron microscopic study of a case. *Acta Cytol* 1984;28:605–9.
- Pelosi G, Iannucci A, Zamboni G, *et al.* Solid and cystic papillary neoplasm of the pancreas: a clinic-cytopathologic and immunocytochemical study of five new cases diagnosed by fine needle aspiration cytology and a review of the literature. *Diagn Cytopathol* 1995;13:233–46.
- Pettinato G, Manivel JC, Ravetto C, *et al.* Papillary cystic tumor of the pancreas. A clinicopathologic study of 20 cases with cytologic, immunohistochemical, ultrastructural, and flow cytometric observations, and a review of the literature. *Am J Clin Pathol* 1992;98:478–88.
- Alm P, Jonsson PE, Karp W, *et al.* A case of papillary-cystic epithelial neoplasm of the pancreas. *Acta Pathol Microbiol Scand* 1981;89:125–32.
- Itho H. A case of pancreas tumor in which pancreatic pseudocyst was suspected initially. *Pediatr Oncol (Shoni-gan)* 1984;20:211–12.
- Jeng LB, Chen MF, Tang RP. Solid and papillary neoplasm of pancreas: emphasis on surgical treatment. *Arch Surg* 1993;128:433–6.
- Ky A, Shilyansky J, Gerstle J, *et al.* Experience with papillary and solid epithelial neoplasms of the pancreas in children. *J Pediatr Surg* 1998;33:42–4.
- Ludvik B, Scheithauer W, Wrba F, *et al.* Gestagen receptor-positive, solid-cystic acinar-cell tumor of the pancreas. *Dtsch Med Wochenschr* 1989;114:903–6.
- Todani T, Shimada K, Watanabe Y, *et al.* Frantz's tumor: a papillary and cystic tumor of the pancreas in girls. *J Pediatr Surg* 1988;23:116–21.
- Ward HC, Leake J, Spitz L. Papillary cystic cancer of the pancreas: diagnostic difficulties. *J Pediatr Surg* 1993;28:89–91.
- Yamaguchi K, Hirakata R, Kitamura K. Papillary cystic neoplasm of the pancreas: radiological and pathological characteristics in 11 cases. *Br J Surg* 1990;77:1000–3.
- Yang YJ, Chen JS, Chen CJ, *et al.* Papillary cystic tumor of the pancreas in children. *Scand J Gastroenterol* 1996;31:1223–7.
- Jung SE, Kim DY, Park KW, *et al.* Solid and papillary epithelial neoplasm of the pancreas in children. *World J Surg* 1999;23:233–6.
- Wunsch LP, Flemming P, Werner U, *et al.* Diagnosis and treatment of papillary cystic tumor of the pancreas in children. *Eur J Pediatr Surg* 1997;7:45–7.
- Campanile M, Nicolas A, LeBel S, *et al.* Frantz's tumor: is mutilating surgery always justified in young patients? *Surg Oncol* 2011;20:121–5.
- Shimotakahara A, Yamataka A, Yanai T, *et al.* Roux-en-Y hepaticojejunostomy or hepaticoduodenostomy for biliary reconstruction during the surgical treatment of choledochal cyst: which is better? *Pediatr Surg Int* 2005;21:5–7.
- Bombi JA, Milla A, Badal JM, *et al.* Papillary-cystic neoplasm of the pancreas: report of two cases and review of the literature. *Cancer* 1984;54:780–4.
- Kallichanda N, Tsai S, Stabile BE, *et al.* Histogenesis of solid pseudopapillary tumor of the pancreas: the case for the centroacinar cell of origin. *Exp Mol Pathol* 2006;81:101–7.
- Kloppel G, Morohoshi T, John HD, *et al.* Solid and cystic acinar cell tumor of the pancreas. A tumor in young women with favorable prognosis. *Virchows Arch A Pathol Anat Histopathol* 1981;392:171–83.

- 60 Haumoudi AB, Misugi K, Grosfeld JL, *et al.* Papillary epithelial neoplasm of pancreas in a child. Report of a case with electron microscopy. *Cancer* 1970;26:1126–34.
- 61 Mao C, Guvendi M, Domenico DR, *et al.* 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* 1995;118:821–8.
- 62 Matsunou H, Konishi F. Papillary-cystic neoplasm of the pancreas. A clinicopathologic study concerning the tumor aging and malignancy of nine cases. *Cancer* 1990;65:283–91.
- 63 Zinner MJ, Shurbaji MS, Cameron JL, *et al.* Solid and papillary epithelial neoplasms of the pancreas. *Surgery* 1990;108:475–80.
- 64 Tanaka Y, Kato K, Notohara K, *et al.* Frequent beta-catenin mutation and cytoplasmic/nuclear accumulation in pancreatic solid-pseudopapillary neoplasm. *Cancer Res* 2001;61:8401–504.
- 65 Wang KS, Albanese C, Dada F, *et al.* Papillary cystic neoplasm of the pancreas: a report of three pediatric cases and literature review. *J Pediatr Surg* 1998;33:842–5.
- 66 Fried P, Cooper J, Balthazar E, *et al.* A role for radiotherapy in the treatment of solid and papillary neoplasms of the pancreas. *Cancer* 1985;56:2783–5.

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