

## CASE REPORT

# Solid Pseudo-Papillary Neoplasm of Pancreas: An Unusual Presentation and Management

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### ABSTRACT

Solid pseudo-papillary neoplasms are rare, indolent pancreatic tumours in young women. We report an acute, large ruptured solid pseudo-papillary neoplasms in a child. At index surgery only de-bulking with control of haemorrhage was done in view of extensive, multi-organ resection. Four cycles of chemotherapy allowed us to downstage the tumour and subsequently a distal pancreatectomy with splenectomy was able to achieve negative margins. Patient was asymptomatic and without recurrence at 12 months. This case is being reported for the rarity of the tumour in this age group as well as for the difference in treatment strategy adopted because of the unusual presentation.

### INTRODUCTION

Solid pseudo-papillary neoplasms (SPN) are uncommon pancreatic tumours in children. Most SPNs, although often large, are well circumscribed, and complete surgical resection is possible and usually associated with cure. We report an unusual case of a large, initially unresectable ruptured SPN in a child, which was managed with pre-operative chemotherapy followed by surgery in two stages. This case is being reported for the rarity of the tumour in this age group as well as for the difference in treatment strategy adopted because of the non-resectability.

### CASE REPORT

An eleven-year-old girl presented to our hospital with complaints of severe abdominal pain associated with vomiting and abdominal distention for four days. The pain was non-radiating and there was no fever, jaundice, haemoptysis or melena. There was reduced urine output and episodes of altered consciousness but no history of any trauma.

At presentation, she was drowsy but arousable. She had hypotension requiring inotropes. Abdominal examination revealed a grossly distended abdomen with tenderness but no rigidity or rebound tenderness. Her haemoglobin was 6.4 (11.5-15.5) g/dL and total leucocyte count was 20,400

(4,000-11,000) cells/ $\mu$ L with neutrophilic leukocytosis. The biochemical parameters were normal. CT imaging revealed a large poorly demarcated predominantly necrotic and haemorrhagic mass (**Figure 1**) in relation to the body and tail of pancreas with extension to the spleen and stomach, which was displacing the transverse and proximal descending colon. The mass was also abutting the duodeno-jejunal flexure and the left kidney. There was evidence of blood in the peritoneal cavity (**Figure 2**). There was no single feeding vessel supplying the mass and therefore angio-embolisation was not possible to control the bleeding.

In view of her haemodynamic instability the patient underwent emergency surgery. At operation, 1400 mL of blood was drained from the peritoneal cavity from a ruptured mass in the region of the tail of the pancreas which was adherent to the splenic flexure of the colon, spleen and the transverse mesocolon (**Figure 3**). The lesser sac was completely obliterated. In view of the extent of tumour and the patient's unstable condition, no attempt at resection was made. We performed a lavage of the peritoneal cavity and obtained biopsies from the mass after achieving haemostasis.

She required intensive care for two days following which she was shifted to the ward after her condition stabilized. Her fever improved with broad spectrum antibiotics. PET- CT done ten days after surgery to re-stage the disease revealed a large lobulated, peripherally FDG-avid, solid-cystic mass lesion in relation to the pancreatic tail with multiple FDG-avid enhancing lobulated solid mural nodules with a peripherally FDG-avid loculated collection along the margins of the mass. There was a mildly FDG-avid, moderate, left pleural effusion and mildly FDG-avid fluid in the pelvis (**Figure 4**). Histopathological evaluation of the

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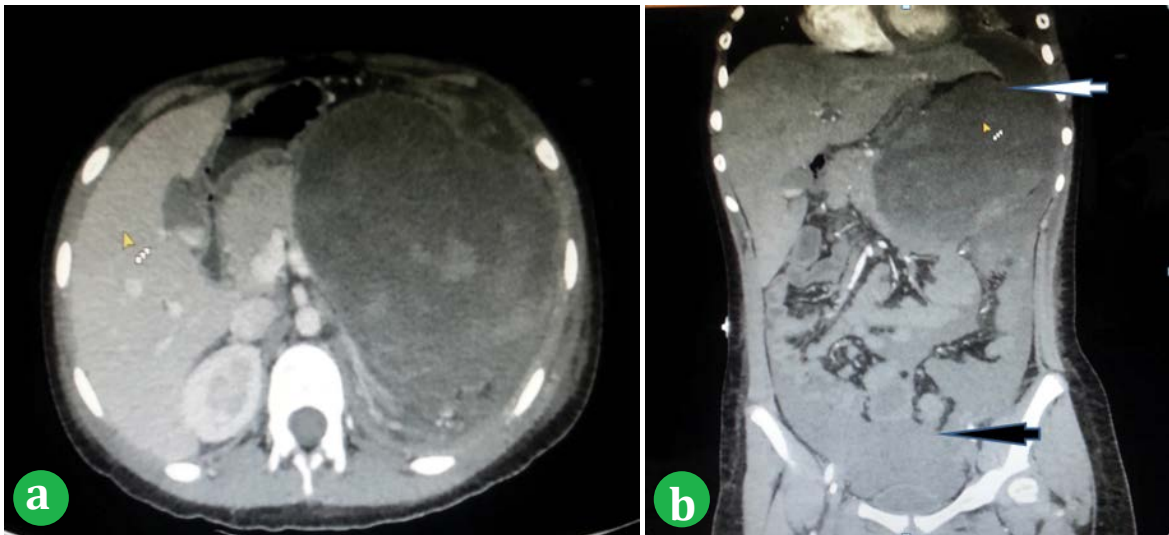
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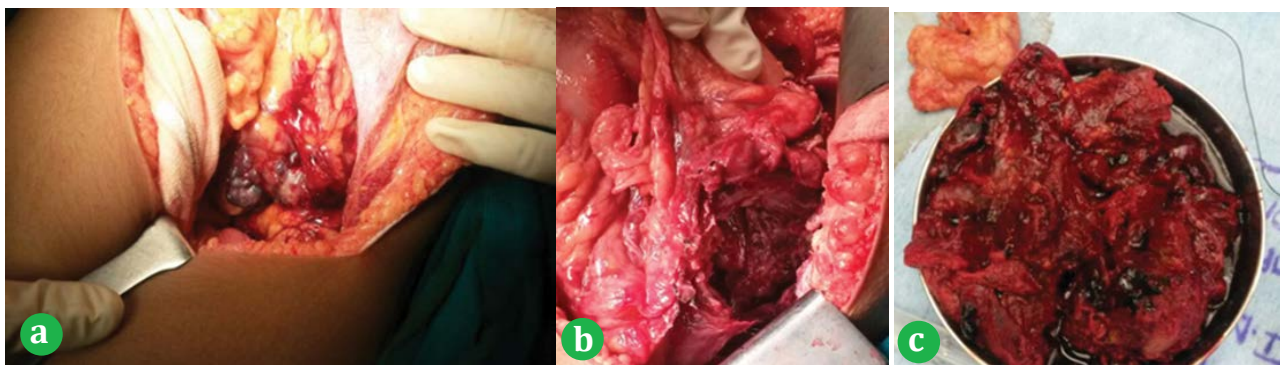
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**Figure 1.** (a) A large necrotic mass (13.6X10cm) with internal haemorrhage in relation to tail of pancreas, (b) Discontinuity in capsule (white arrow) with pelvic collection (black arrow) suggestive of rupture.



**Figure 2.** (a) Left-sided pleural effusion compressing the left lung. (b). Mass compressing the stomach with (c) Displacement of colon.



**Figure 3.** (a). Pancreatic mass in the region of the tail of the pancreas with nodular infiltration into the mesocolon, (b). ruptured pancreatic mass densely adherent to surrounding structures, (c). mass showing areas of necrosis and haemorrhage on cut section.

resected mass revealed solid pseudopapillary neoplasm of the pancreas. Her postoperative recovery was uneventful and she was discharged on post-operative day 16.

Definitive surgery for SPN at this juncture would require extensive bowel resection, splenectomy and complete pancreatectomy. In order to avoid a morbid surgery in a child, it was planned to treat her with neoadjuvant chemotherapy to improve the resectability of the mass. She was treated with four cycles of chemotherapy with vincristine (1.5 mg/m<sup>2</sup> IV x1), actinomycin-D (0.045 mg/kg IV x1) and cyclophosphamide (1200 mg/m<sup>2</sup> IV as 1 hour infusion with MESNA and fluids) at intervals of three weeks with ultrasound (USG) monitoring of the tumour response. She tolerated the chemotherapy well with no episodes of grade 3-4 toxicity. Ultrasonogram done after

4 cycles showed a reduction in size of the mass to 6x4 cm localized to the tail of the pancreas.

She subsequently underwent tumour resection with distal pancreateo-splenectomy (Figure 5). Her post-operative recovery was unremarkable and she was discharged six days after the procedure. Histopathology revealed features confirmed complete resection of solid pseudo-papillary neoplasm (solid cystic papillary cystic tumour) with focal infiltration into peri-pancreatic fat. The resected margins and lymph nodes were free of tumour. At 12 months patient is asymptomatic with no evidence of recurrence.

**DISCUSSION**

Primary malignant tumours of pancreas are extremely rare. SPN, pancreatoblastomas, adenocarcinomas,



neuroendocrine tumours have been reported in addition to sarcomas and lymphomas. Given the rare incidence, treatment experience is limited to institutional series and case reports.

SPN is usually reported in the 2<sup>nd</sup> and 3<sup>rd</sup> decade of life especially in women. It is often surrounded by a pseudocapsule and exhibits benign or low-grade malignancy. Conservative resection with preservation of as much pancreatic tissue as possible is the treatment of choice. According to the location of the tumour, surgical treatment entails a distal pancreatectomy with or without splenectomy, or pylorus-preserving pancreatoduodenectomy, or a standard Whipple operation. Despite a large tumour size or vascular involvement, SPNs are usually resectable [1]. Resection for metastases has been described at the time of primary resection or even for isolated single site recurrence [2]. Extended resections for advanced or metastatic disease has been reported to be associated with good long term survival (>10 years) justifying this aggressive approach [3, 4, 5, 6, 7]. Lymph node metastasis is uncommon [8, 9, 10]. Moreover, lymph nodal involvement does not affect the long term survival in these patients [3, 11, 12]. Hence, there is no role for radical lymphadenectomy in these tumours. Positive margins post resection again has not been found to affect the overall outcome thus questioning the need for radical surgery. Surgical treatment alone results in 5 year survival of >95%.

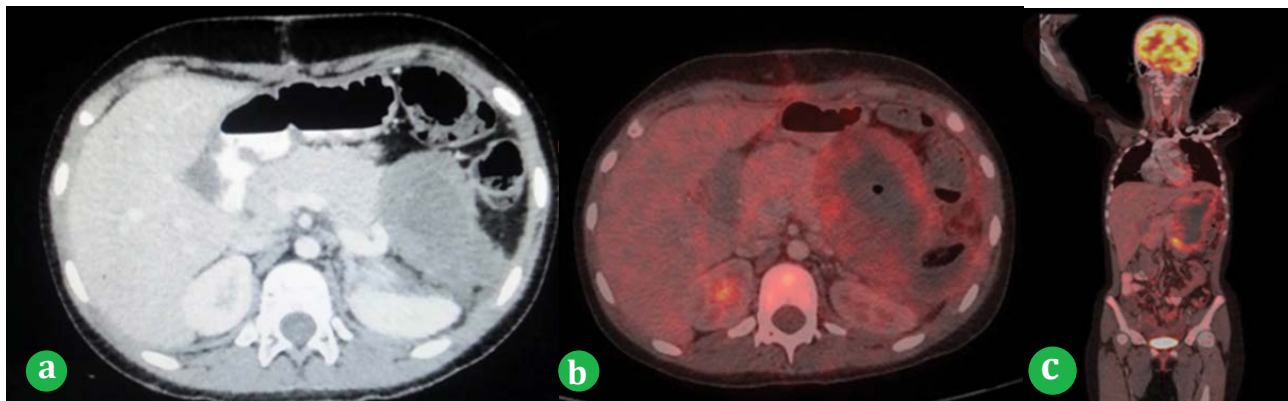
There is very limited literature regarding management of patients with unresectable SPN. There are only isolated

case reports regarding the use of chemotherapy to down-stage the disease (**Table 1**) [13, 14, 15]. Matsuda *et al.* have reported the use of intra-arterial chemoembolization of doxorubicin in a patient with SPN with multiple liver metastases [16]. Subsequent systemic combination chemotherapy (with 5-FU, doxorubicin and mitomycin-c) proved to be far less effective for both the primary and metastatic sites in this patient.

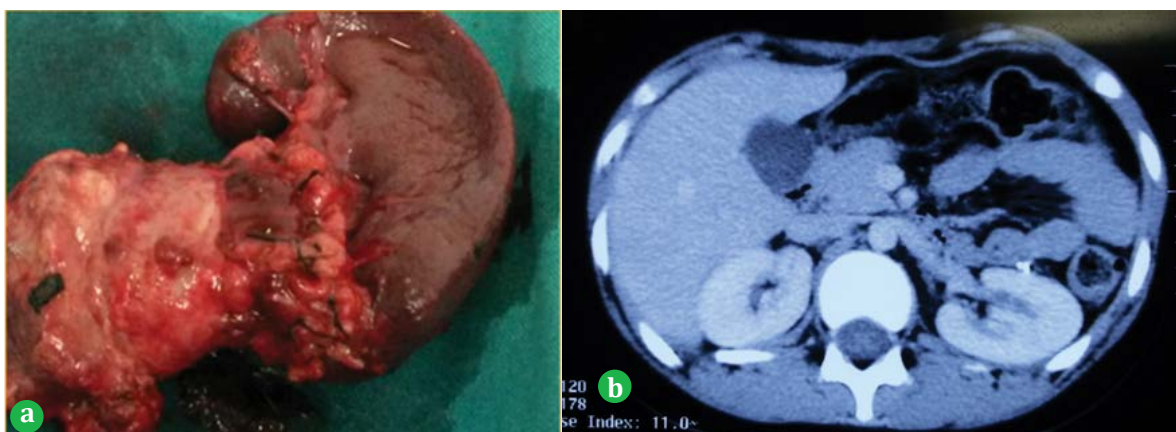
Chemotherapeutic agents that have been tried in the neoadjuvant setting are Cisplatin with 5-Fluoro-Uracil (5-FU) [14], Gemcitabine with Cisplatin [13], Gemcitabine alone [15] and Tamoxifen in hormone receptor positive tumours. Chemotherapy in the adjuvant setting was utilized by Tajima *et al.* [5] who used S-1 for local recurrence and hepatic arterial infusion of Gemcitabine for liver metastases. Rebhandl *et al.* used chemotherapy in view of recurrence which was stable at 6 months [17]. The use of chemotherapy has also been reported by Sperti *et al.* about 32 months after the index surgery for multiple liver metastases resulting in disease control [6].

Radiotherapy (RT) has also been used occasionally in patients with locally advanced SPNs to render them amenable to resection [4, 18, 19, 20]. The reported responses to RT varies from a partial to a complete radiologic remission [4, 18].

To the best of our knowledge, there is no previous report on the management of a patient presenting with a ruptured SPN. In an attempt to reduce the morbidity of



**Figure 4.** (a). CT showing a necrotic lesion in relation to the tail of the pancreas. (b). Peripherally FDG-avid large lobulated lesion in the tail of the pancreas with FDG-avid left pleural effusion and pelvic ascites. (c). Peripherally FDG-avid lesion in the tail of the pancreas.



**Figure 5.** (a) Resected specimen with distal pancreatectomy and splenectomy. (b) Follow-up scan at 12 months shows no recurrence.

**Table 1.** Isolated case reports.

|                            | Intention    | Chemo-radiotherapy                     | Response               | Surgery                                 | Survival |
|----------------------------|--------------|--|------------------------|---|----------|
| Strauss <i>et al.</i> [14] | Down-staging | Chemotherapy (Cisplatin +5-FU)         | Partial response       | Pancreatico-duodenectomy                | -        |
| Das <i>et al.</i> [13]     | Down-staging | Chemotherapy (Gemcitabine + Cisplatin) | Partial response       | Spleen preserving distal pancreatectomy | 13 mo    |
| Maffuz <i>et al.</i> [15]  | Down-staging | Chemotherapy (Gemcitabine)             | Near complete response | Pancreatico-duodenectomy                | -        |

an extended resection the patient was only subjected to partial debulking surgery with haemostasis and biopsy as an initial procedure. Again due to the rarity of this situation in children, there is no previous report of using chemotherapy for SPN [21]. We chose VAC for its efficacy in epithelial tumours and for the absence of any short or long term side effects at the dose used. With use of chemotherapy, surgery with potential for life long morbidity in a young girl was avoided. She was subsequently managed with complete resection of residual mass with a good outcome.

### CONCLUSIONS

SPN may rarely present with rupture with exsanguinating haemorrhage. There may be a role for an initial salvage procedure followed by chemotherapy as a down-staging modality in patients presenting with large or unresectable SPNs. This can be followed then by a complete tumour resection.

### Conflict of Interest

All authors declare no conflict of interests.

### References

1. Klimstra DS, Wenig BM, Hefless CS. Solid-pseudopapillary tumor of the pancreas: a typically cystic carcinoma of low malignant potential. *Semin Diagn Pathol* 2000; 17:66-80. [PMID: 10721808]
2. Hassan I, Celik I, Nies C, Zielke A, Gerdes B, Moll R, Ramaswamy A, et al. Successful treatment of solid-pseudo-papillary tumor of the pancreas with multiple liver metastases. *Pancreatol* 2005; 5:289-94. [PMID: 15855828]
3. Nishihara K, Nagoshi M, Tsuneyoshi M, Yamaguchi K, Hayashi I. Papillary cystic tumors of the pancreas: assessment of their malignant potential. *Cancer* 1993; 71:82-92. [PMID: 8416730]
4. Fried P, Cooper J, Balthazar E, Fazzini E, Newall J. A role for radiotherapy in the treatment of solid and papillary neoplasms of the pancreas. *Cancer* 1985; 56:2783-2785. [PMID: 4052952]
5. Tajima H, Takamura H, Kitagawa H, Nakayam H, Shoji M, Watanabe T, Tsukada T, et al. Multiple liver metastases of pancreatic solid pseudopapillary tumor treated with resection following chemotherapy and transcatheter arterial embolization: A case report. *Oncol Lett* 2015; 9:1733-1738. [PMID: 25789032]
6. Sperti C, Berselli M, Pasquali C, Pastorelli D, Pedrazzoli S. Aggressive behaviour of solid-pseudopapillary tumor of the pancreas in adults: a case report and review of the literature. *World J Gastroenterol* 2008; 14:960-5. [PMID: 18240360]

7. Levy P, Bougaran J, Gayet B. Diffuse peritoneal carcinosis of pseudo-papillary and solid tumor of the pancreas. Role of abdominal injury. *Gastroenterol Clin Biol* 1997; 21:789-793. [PMID: 9587521]
8. Machado C. M, Machado A. M, Bacchella T, Jukemura J, Almeida JL, Cunha JE. Solid pseudopapillary neoplasm of the pancreas: distinct patterns of onset, diagnosis, and prognosis for male versus female patients. *Surgery* 2008; 143:29-34. [PMID: 18154930]
9. Yang F, Jin C, Long J, Yu XJ, Xu J, Di Y, Li J, et al. Solid pseudopapillary tumor of the pancreas: a case series of 26 consecutive patients. *Am J Surg* 2009; 198:210-215. [PMID: 19268906]
10. Goh BK, Tan YM, Cheow PC, Alexander Chung YF, Chow PK, Wong WK, Ooi LL. Solid pseudopapillary neoplasms of the pancreas: An updated experience. *J Surg Oncol* 2007; 95:640-4. [PMID: 17477365]
11. Guo N, Zhou QB, Chen RF, Zou SQ, Li ZH, Lin Q, Wang J, et al. Diagnosis and surgical treatment of solid pseudopapillary neoplasm of pancreas: Analysis of 24 cases. *J Can Chir* 2011; 54:368-74. [PMID: 21939604]
12. Stommer P, Kraus J, Stolte M, Giedl J. Solid and cystic pancreatic tumors: clinical, histochemical, and electron microscopic features in ten cases. *Cancer* 1991; 67:1635-1641. [PMID: 1900454]
13. Das G, Bhuyan C, Das BK, Sharma JD, Saikia BJ, Purkayastha J. Spleen-preserving distal pancreatectomy following neoadjuvant chemotherapy for papillary solid and cystic neoplasm of pancreas. *Indian J Gastroenterol* 2004; 23:188-189. [PMID: 15599007]
14. Strauss JF, Hirsch VJ, Rubey CN, Pollock M. Resection of a solid and papillary epithelial neoplasm of the pancreas following treatment with cisplatin and 5-fluorouracil: a case report. *Med Pediatr Oncol* 1993; 21:365-367. [PMID: 8492753]
15. Maffuz A, Bustamante FT, Silva JA, Torres-Vargas S. Preoperative gemcitabine for unresectable, solid pseudopapillary tumour of the pancreas. *Lancet Oncol* 2005; 6:185-186. [PMID: 15737835]
16. Matsuda Y, Imai Y, Kawata S, Nishikawa M, Miyoshi S, Saito R, Minami Y, Tarui S. Papillary cystic neoplasm of the pancreas with multiple hepatic metastases: a case report. *Gastroenterology Jpn* 1987; 22:379-384. [PMID: 2442059]
17. Rebhandl W, Felberbauer FX, Puig S, Paya K, Hochschorner S, Barlan M, Horcher E. 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. [PMID: 11320522]
18. Zauls JA, Dragun AE, Sharma AK. Intensity-modulated radiation therapy for unresectable solid pseudopapillary tumor of the pancreas. *Am J Clin Oncol* 2006; 29:639-640. [PMID: 17149006]
19. Dales RL, Garcia JC, Davies RS. Papillary-cystic carcinoma of the pancreas. *J Surg Oncol* 1983; 22:115-117. [PMID: 6823124]
20. Matsunou H, Konishi F. Papillary-cystic neoplasm of the pancreas. A clinicopathologic study concerning the tumor aging and malignancy of nine cases. *Cancer* 1990; 65:283-291. [PMID: 2295051]
21. Sclafani LM, Reuter VE, Coit DG, Brennan MF. The malignant nature of papillary and cystic neoplasm of the pancreas. *Cancer* 1991; 68:153-8. [PMID: 2049737]