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Solid pseudopapillary tumor of the pancreas: Experience at a tertiary care centre of Northern India

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ABSTRACT

INTRODUCTION: Solid pseudopapillary tumor (SPT) of the pancreas is rare, accounting for 0.13–2.7% of all pancreatic tumors. It is unique, has low malignant potential and predominantly affects young women. Radiological and pathological studies have revealed that the tumor is quite different from other pancreatic tumors. But the cell origin of SPT and tumorigenesis are still enigmatic. Abdominal mass is the most common presenting symptom. Due to the paucity of the number of cases, the natural history of the disease is not fully understood. This study was undertaken to examine the clinico-pathological characteristics of the disease and to evaluate the outcome of surgical intervention in a tertiary referral care centre.

MATERIALS AND METHODS: A retrospective analysis of all patients diagnosed and treated for SPN in our hospital over a period of 10 years (2005–2015) was carried out. A database of the characteristics of these patients was developed. In all, 11 patients were identified. A CT scan of the abdomen was performed in all the patients and the findings revealed a mass in the pancreas. The investigations performed included routine blood investigations, chest X-ray, CA-19-9 level and either an ultrasound or a CT Scan of the abdomen.

RESULTS: During the time period of 10 years, of 349 patients with pancreatic malignancy admitted to our department, only 11 were diagnosed as having SPN (3.15%). Ten patients were women (90%) and one patient was a man (10%). The patients had a median age of 27.6 years (range 17–41). The most common symptoms were abdominal pain and dullness. Eight patients (72.7%) presented with abdominal pain or abdominal dullness and three patient (27%) were asymptomatic. All the 11 patients were taken up for surgery. Three patients underwent distal pancreatectomy with splenectomy, three patients underwent the total mass excision and one patient underwent total pancreatic resection. Three required extended distal pancreatectomy with splenectomy. One underwent spleen-preserving distal pancreatectomy.

CONCLUSION: SPT is rare, but treatable pancreatic tumor. While clinical signs and symptoms are relatively nonspecific, characteristic findings on imaging and histology separate these tumors from the more malignant pancreatic tumors. The prognosis is favorable even in the presence of distant metastasis. Although surgical resection is generally curative, a close follow-up is advised in order to diagnose a local recurrence or distant metastasis.

1. Introduction

First described by Franz in 1959, Solid pseudopapillary tumor (SPT) of the pancreas is rare, accounting for 0.13–2.7% of all pancreatic tumors [1]. It is unique, has low malignant potential and predominantly affects young women [1,2]. Until it was defined by the World Health Organization (WHO) in 1996 as 'solid pseudopapillary tumor' of the pancreas, this tumor was described by using various names including 'solid cystic tumor', 'papillary cystic tumor', 'papillary epithelial neoplasia', 'solid and papillary epithelial neoplasia', 'papillary epithelial tumor' and 'Frantz's tumor', 'solid and papillary tumor', 'solid-cysticpapillary epithelial neoplasm', 'benign or malignant papillary tumor of the pancreas' [3].

Radiological and pathological studies have revealed that the tumor is quite different from other pancreatic tumors. But the cell origin of SPT and tumorigenesis are still enigmatic. The pathogenesis is thought to result from cells of the endocrine pancreas though some investigators have postulated origin from the exocrine pan-

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creas. These tumours have a long asymptomatic period and are usually detected when they have grown to a large size [4–8].

Abdominal mass is the most common presenting symptom, with dyspepsia, early satiety, nausea, or vomiting being less common presenting symptoms. Up to 20% of patients are asymptomatic with tumors identified either incidentally on imaging or at operation for unrelated pathology [9,10]. Grossly, SPTs are identified as well demarcated, encapsulated tumors with extrapancreatic growth. Mixed solid and cystic components are evident with internal necrotic or hemorrhagic debris and lobulated, solid tissue at the periphery. Characteristic radiographic features include the presence of an encapsulated mass with solid and cystic components on either CT scan or MRI, with MRI notably better for identification of certain tumor characteristics such as the presence of a capsule, hemorrhage or cystic degeneration [10]. SPT should be added to the differential diagnosis in any patient with a solid and partly cystic mass of the pancreas especially in females under 35 years of age. Surgical resection is the treatment of choice for affected patients and is associated with an overall good prognosis [11].

Due to the paucity of the number of cases seen, the natural history of the disease is not fully understood. This study was undertaken to examine the clinico-pathological characteristics of the disease and to evaluate the outcome of surgical intervention in a tertiary referral cancer centre.

2. Material and methods

A retrospective analysis of all patients diagnosed and treated for SPN in our hospital over the past 10 years was carried out (2005–2015). A database of the characteristics of these patients was developed, including age, gender, tumor location (data were derived from radiological investigations or surgical records) and size (data were derived from radiological investigations or surgical records and finally confirmed by pathology), treatment (data were derived from the medical records, including the types of surgery), and histopathological and immunohistochemical features. In all, 11 patients were identified. A CT scan of the abdomen was performed in all the patients and the findings revealed a mass in the pancreas. Pre-operative fine needle aspiration cytology (FNAC) was performed in 2/11 patients. All the patients who underwent resection were followed up every 6 months. The investigations performed included routine blood investigations, chest X-ray, CA-19-9 level and either an ultrasound or a CT Scan of the abdomen.

3. Results

During the time period of 10 years, of 349 patients with pancreatic malignancy (which does not include ampulla vateri, distal choledocal and duodenal tumor) admitted to our department, only 11 were diagnosed as having SPN (3.15%). Ten patients were women (90%) and one patient was a man (10%). The patients had a median age of 27.6 years (range 17–41). The most common symptoms were abdominal pain and dullness. Eight patients (72.7%) presented with abdominal pain or abdominal dullness and three patient (27%) were asymptomatic with the diagnosis made by an incidental finding on routine examination. Abdominal CT and/or magnetic resonance imaging (MRI) showed the typical features of solid pseudopapillary neoplasm in seven (63.6%) of the patients (Fig. 1). Usually, the tumors appeared as, well circumscribed lesions with a mixed cystic and solid component but were almost entirely solid or else cystic with thick walls. In two patients the tumor was located in the pancreatic head (18%), in three patients in the body (27%) and in the remaining six patients in the tail (54%).

All the 11 patients were taken up for surgery. Three patients underwent distal pancreatectomy with splenectomy, three patients underwent the total mass excision and one patient underwent total pancreatic resection. Three required extended distal pancreatectomy with splenectomy. One underwent spleen-preserving distal pancreatectomy. The mean diameter of the tumor was 6.9 cm (range 3–12 cm). Patient characteristics are summarized in Table 1. In seven cases lymph node dissection was done in a number between 5 and 14, whereas no dissection was needed for four patients. No lymph node metastasis was present in any patient.

At histopathological examination, tumor mass separated from pancreas with a fibrous capsule was seen. Pseudopapillary, cystic and solid growth patterns were seen in the tumor mass. Tumor cells had an ovally shaped, small and centrally localized nucleus and large eosinophilic cytoplasm. Tumors consisted of pseudopapillary structures made of cells aligned around fine vessels, solid areas, hemorrhagic areas and cystic areas of different size (Fig. 2). No mitosis was seen in eight cases, whereas minimal mitosis was present in one case (2/10 per high powered field) and multiple mitosis were present in two cases (20/10 per high powered field) (Table 2). The immunohistochemistry profiles are summarized in Table 3 showh in Fig. 3 Capsular invasion was present in three cases (case numbers 2, 6 and 10), spleen invasion wasalso present in case number 2. These three cases were considered as malignant SPN and treated with six courses of gemcitabine + cis-platinum chemotherapy. Multiple liver and omentum metastases developed in case number 2 at the seventh postoperative month; this patient died at the ninth month. Multiple liver and omentum metastases developed in case number 6 at the 20th postoperative month and she died at the 24th month. The other eight cases have been followed up closely and no recurrence or metastasis has been seen. The average postoperative hospital stay was 10.3 days.

4. Discussion

Solid pseudopapillary tumor of the pancreas (SPT) is an uncommon and enigmatic pancreatic neoplasm first described by Frantz in 1959 [1]. This lesion usually has a low malignant potential [2]. The tumor has been identified by a number of synonyms including solid and cystic tumor, solid and papillary epithelial neoplasm, papillary cystic neoplasm, papillary cystic epithelial neoplasm, papillary cystic tumor, and Franz tumor. In 1996, the World Health Organization (WHO) renamed this tumor as SPT for the international histologic classification of tumor of the exocrine pancreas [3]. This uncommon, typically benign tumor is found mainly in young non-Caucasian women between the 2nd and 3rd decades of life. It seems to have a predilection for Asian and African- American women, although rare cases have been reported in children and men [12]. Female predominance has been attributed to the proximity of primordial pancreatic cells to the ovarian ridge during development [13].

The differential diagnosis of suspicious neoplasms should include microcystic adenoma, mucinous cystic neoplasm, nonfunctioning islet cell tumor, pancreatic adenocarcinoma, pancreaticoblastoma, cystic degeneration of solid neoplasm and calcified hemorrhagic pseudocyst [14]. Abdominal discomfort or vague pain is the most common symptom, followed by a gradually enlarging mass and compression signs induced by the tumor. Some patients are completely asymptomatic, with the tumor detected incidentally by imaging studies or routine physical examination [1]. CT scan, ultrasonography (US) and endosonography (EUS) have been used with variable success in diagnosing SPN. CT scan and EUS are more sensitive and specific and have shown more accuracy in diagnosing SPN [15]. Magnetic resonance imaging (MRI) can be diagnostic. Typically, a large, well-defined, encapsulated lesion with heterogeneous high or low signal intensity on T1-weighted,

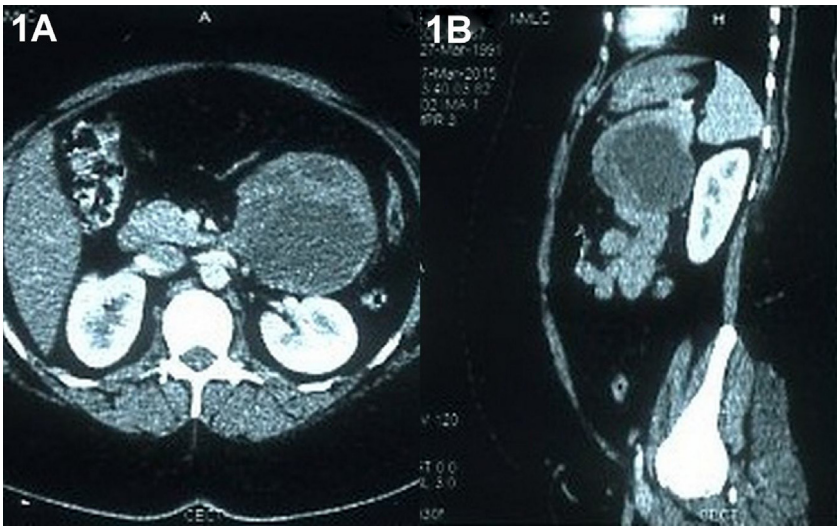

Fig. 1. (A&B) CECT abdomen revealed a 9.4 × 9.2 × 10.2 cm lesion arising from body and tail of pancreas.

Table 1
PATIENT CHARACTERISTICS.

CASE	AGE/SEX	SURGERY	LOCATION	SIZE (CMS)	INVASION	NODAL STATUS	FOLLOW UP
1	17/F	DPS	BODY	9 × 6 × 3	–	0/7	HEALTHY
2	19/F	DPS	TAIL	11 × 7 × 3	CAPSULE AND SPLEEN	0/14	HEALTHY
3	21/F	DPS	HEAD	12 × 6 × 3	–	0/11	HEALTHY
4	27/F	TME	TAIL	7 × 5 × 3	–	–	HEALTHY
5	41/F	TP	TAIL	6 × 3 × 1	–	–	HEALTHY
6	17/F	SPDP	BODY + TAIL	4 × 3 × 3	CAPSULE	0/7	OMENTAL METS
7	34/F	SDPS	HEAD + TAIL	11 × 7 × 3	–	0/6	OMENTAL METS
8	28/M	SDPS	TAIL	4 × 3 × 2	–	0/12	OMENTAL METS
9	33/F	TME	BODY + TAIL	3 × 3 × 2	–	0/5	HEALTHY
10	40/F	TME	TAIL	4 × 3 × 2	CAPSULE	–	HEALTLY
11	27/F	SDPS	TAIL	5 × 2 × 1.5	–	–	HEALTHY

DPS: DISTAL PANCREACTOMY AND SPLENCTOMY; TME: TOTAL MASS EXCISION; TP: TOTAL PANCREACTOMY; SPDP: SPLEEN PRESERVING DISTAL PANNCREACTOMY; SDPS: SUBTOTAL DISTAL PANCREACTOMY WITH SPLENECTOMY.

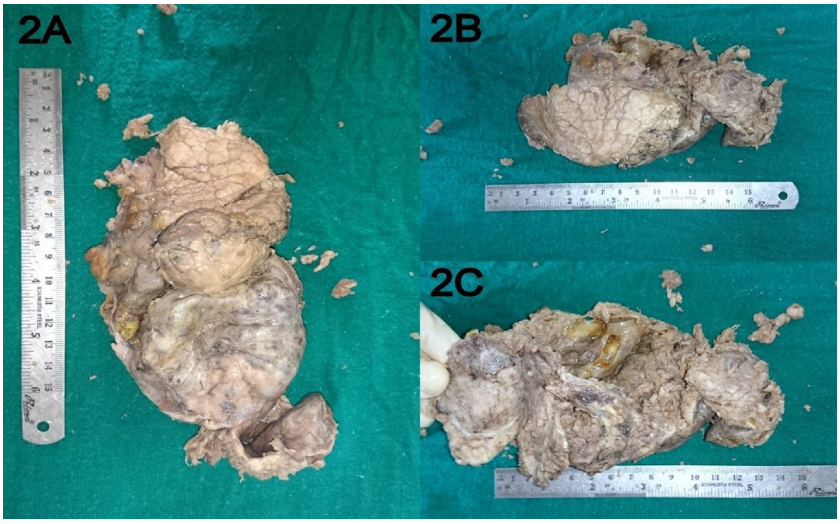

Fig. 2. (A, B & C) ON GROSS: an encapsulated, well demarcated tumor in body and tail of pancreas.

heterogeneous high signal intensity on T2 weighted, and early peripheral heterogeneous enhancement with progressive fill-in is found on gadolinium-enhanced dynamic MRI. These features help differentiate this rare tumour from other pancreatic neoplasms [16]. It exhibits the typical features on CT scan, including regular shape, well-defined margins and homogeneous appearance corresponding to the solid and cystic texture [2,4,6]. These radiological features are very different from those of ductal adenocarnoma, as the latter is always infiltrative with poorly defined margin and is invasive to the surrounding tissues. The tumor may involve any portion of the pancreas but the head and tail are most common.

Table 2
HISTOPATHOLOGICAL FEATURES.

CASES	PATTERN	NECROSIS	MITOSIS	PLEOMORPHISM
1	SOLID, CYSTIC, PAPILLARY	+	–	–
2	CYSTIC, PAPILLARY	–	–	–
3	SOLID, CYSTIC, PAPILLARY	–	–	MINIMAL
4	SOLID, CYSTIC, PAPILLARY	+	–	MINIMAL
5	SOLID, CYSTIC, PAPILLARY	–	–	+
6	SOLID, CYSTIC, PAPILLARY	–	–	+
7	SOLID, CYSTIC, PAPILLARY	–	–	+
8	SOLID, CYSTIC, PAPILLARY	+	2/10 HPF	+
9	CYSTIC, PAPILLARY	+	20/10 HPF	+
10	CYSTIC, PAPILLARY	–	20/10 HPF	+
11	CYSTIC, PAPILLARY	+	–	+

Table 3
IMMUNOHISTOCHEMISTRY RESULTS.

CASE	CK	CEA	VIMENTIN	CHROMO	NSE	CD 10	CD 56	SYNAPTO	P53	KI 67	PR	EMA
1	+	–	+	–	–	+	–	+	–	–	+	+
2	+	–	+	–	+	+	+	–	–	–	+	+
3	+	–	+	–	+	+	+	+	–	–	+	+
4	+	–	+	–	+	+	+	+	–	–	+	–
5	+	–	+	–	–	+	+	+	–	–	–	+
6	–	–	+	–	+	+	–	+	–	–	–	+
7	+	–	+	–	+	+	+	+	–	–	+	+
8	–	–	+	–	+	+	+	+	–	–	+	+
9	–	–	+	–	+	+	+	–	–	–	–	+
10	–	–	+	–	+	+	–	–	–	–	+	–
11	+	–	+	–	+	+	+	+	–	–	+	+

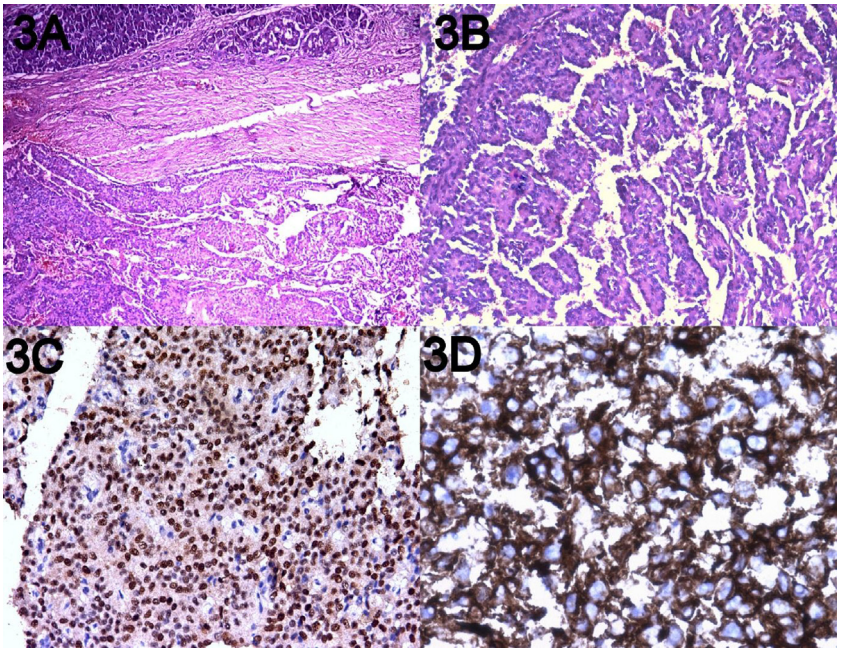

Fig. 3. (A&B) On Microscopy: a well circumscribed tumor mass separated from pancreas by a fibrous capsule. The tumor cells were arranged in sheets, nests, pseudopillary formations. (C) The tumor cells were positive for PR (progesterone receptor). (D) CD10 & CD20.

Adjacent structures such as the mesenteric vessels, stomach, and duodenum may also be involved [17].

FNAC has been used for the preoperative cytological diagnosis of SPN. The cytology specimen is usually highly cellular and is characterized by the presence of epitheloid cells that present singly or in aggregates containing fibrovascular cores. No evidence of pleomorphism or mitotic activity is seen in the cells. The most conclusive criterion for identification of SPN is the pseudopapillary arrangement with bland appearing tumour cells. EUS-guided FNAC has been reported, and this can help in correctly diagnosing SPN pre-operatively [15]. Grossly, the tumor is demarcated from adjacent pancreatic tissue by the presence of a fibrous capsule [10]. On cut section it shows solid and cystic areas with necrotic and haemorrhagic patches. Microscopic examination yields a mixed solid and cystic mass with hemorrhagic or necrotic cellular material in its center with lobules of solid tissue at its periphery. Characteristic findings include the presence of solid areas alternating with pseudopapillary formations, foamy histiocytes, nuclear grooves and cytoplasmic globules [18]. Theories of histogenesis are controversial but have generally been divided into three main groups:

pancreatic duct cell origin, acinar cell origin, or primitive cell origin. Furthermore, sex hormones may play a role in the pathogenesis or growth of SPTs: Nearly all studies demonstrate no evidence of estrogen receptors; however, progesterone receptors are present in many cases [17]. Morales et al. described the temporal relationship between a fast-growing SPT and pregnancy in a young woman and also demonstrated the presence of a progesterone receptor in the tumor tissue. The growth rate of the SPT of the pancreas seemed to be enhanced by the concurrence of pregnancy. Many investigators favor the theory that SPTs originate from multipotent primordial cells, whereas others suggest an extrapancreatic origin from genital ridge angle-related cells [19]. The cystic appearance on gross examination may present a diagnositic dilemma with cystic neoplasms of the pancreas.

Immunophenotyping has been used to differentiate these tumours from other pancreatic neoplasms. Solid-pseudopapillary tumours test positive for vimentin, neuron-specific enolase, α1-antitrypsin, and α1- antichymotrypsin and are negative for chromogranin, epithelial membrane antigen, and cytokeratin, insulin and glucagon. Neuroendocrine tumors, especially the well-differentiated ones, are the most important entities in the differential diagnosis of SPT, because they may display similar light microscopic features, and neuroendocrine markers are variably expressed in SPT [1,2]. Except for the consistently negative results for chromogranin A, expressions of other neuroendocrine markers such as synaptophysin, neuron-specific enolase and CD56 at various levels were demonstrated [1,2]. Recently, the nuclear type of β-catenin has been regarded as an unique immunohistochemical feature of SPT as it underlies the genetic mutation of catenin found in more than 90% of SPT [7,8]. Recently, β-catenin and Wnt signaling pathway has been found to play an important role in SPT tumorigenesis [7]. SPT almost consistently harbours β-catenin gene (CTNNB1) mutations in exon 3 resulting in the activation of the Wnt-signaling pathway [20].

Although most SPTs exhibit benign behavior, malignant degeneration does occur. The malignant pancreatic tumors were often older at presentation and had a male predilection [17]. According to the WHO classification scheme, SPTs with clear criteria of malignancy (vascular and nerve sheath invasion or lymph node and liver metastases) are designated as solid-pseudopapillary carcinomas [3]. Nishihara et al. indicated that venous invasion, high nuclear grade, and prominent "necrobiotic nests" help detect the malignant potential of papillary cystic tumors [21].

In approximately 85% of the patients, SPN is limited to the pancreas, while in 10% to 15% of tumors, and have already metastasized at the time of presentation [22]. The most common sites for metastasis are the liver, regional lymph nodes, mesentery, omentum and peritoneum. Once the diagnosis of SPN is made, surgery is the first choice of treatment. Conservative resection with preservation of as much pancreatic tissue as possible is the treatment of choice. According to the location of the tumor, distal pancreatectomy with or without splenectomy, pylorous preserving pancreatoduodenectomy, Whipple operation or enucleation can be performed [10]. Extensive lymphatic dissection or more radical approaches are not indicated when the disease is localized. Local invasion and metastases are not contraindications for resection. Portal vein resection is advocated when there is evidence of tumor invasion. For the metastases, surgical debulking should be performed, in contrast to other pancreatic malignancies. Metastases can be removed with enucleations or lobectomies and some patients with unresectable SPN may also have a long term survival [22]. The overall five-year survival rate of patients with SPN is about 95% [1]. Malignant SPN, designated as a solid-pseudopapillary carcinoma, occurs in 15% of adult patients. According to the WHO classification system, these are: 1) solidpseudopapillary neoplasms with borderline malignancy potential; and 2) solid-pseudopapillary carcinomas. Criteria which distinguish potentially malignant tumors and which are classified as 'SP carcinoma' are: 1) angioinvasion; 2) perineural invasion; and 3) deep invasion of the surrounding pancreatic parenchyma. A recent study showed that some histological features, such as extensive necrosis, nuclear atypia, high mitotic rate, immunohistochemistry findings of expression of Ki-67 and sarcomatoid areas may be associated with aggressive behavior [23].

Tumor recurrence has been found in 5–7% of patients after surgical resection [1]. In contrast to other pancreatic tumors, aggressive surgical resection is warranted even in the presence of local invasion, recurrence, or limited metastases [17,21,22]. In contrast to other pancreatic tumors, invasion of the portal vein or superior mesenteric artery does not indicate tumor unresectability [24]. Pancreatic fistula is the most common complication after complete resection seen in approximately 7% of patients [9]. The role of adjuvant therapy in treatment of SPTs is unclear, with few studies demonstrating a role for gemcitabine and radiotherapy to downsize large tumor(s) or treat the rare case of unresectable disease. The tumors' high rate of resectability limits the need for adjuvant therapy [25–27].

5. Conclusion

To conclude, SPT is rare, but treatable pancreatic tumor. While clinical signs and symptoms are relatively nonspecific, characteristic findings on imaging and histology separate these tumors from the more malignant pancreatic tumors. The identification of a large bulky pancreatic tumour in a child or woman should raise suspicions of solid pseudopapillary tumour of the pancreas. Because of the indolent nature of these tumours and the low malignant potential, aggressive attempts at complete surgical resection are warranted. Large tumours are usually resectable and size does not predict outcome. Surgical excision offers the best chance for cure and should always be attempted irrespective of the magnitude of resection involved. Patients with SPT have an excellent prognosis after surgical excision. The prognosis is favorable even in the presence of distant metastasis. Although surgical resection is generally curative, a close follow-up is advised in order to diagnose a local recurrence or distant metastasis and choose the proper therapeutic option for the patient.

We state that the work has been reported in line with the SCARE criteria [28].

We declare that there is no conflict of interests amongst the authors.

We also state that there was no source of funding for carrying out this study.

Conflicts of interest

There is no conflict of interest amongst the authors.

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Ethical approval

Not applicable as it is a case series.

Consent

The consent has been taken from the parents of the child for publication of this case series.

Author contribution

Namita Bhutani – Reviewed the literature and wrote the article.
Pradeep Kajal – Superwised the article and did the final editing.
Sham Singla – Operated upon the patients.
Vijender Sangwan – Managed the investigative part including the ultrasonography and CT scan of abdomen and provided the images.

Guarantor

Kamal N Rattan.

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