

Solid pseudopapillary tumor of the pancreas: Report of five cases

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ABSTRACT

Introduction: Solid pseudopapillary tumor of the pancreas (SPT) is a rare pancreatic tumor with an unclear pathogenesis and good prognosis after resection. We report our experience with the diagnostic and therapeutic management of these tumors over 10 years. **Case Series:** A retrospective chart review was performed for all patients undergoing pancreatic resection at our institution from 2001-2011. Patients with final pathology demonstrating SPT were identified. Of 617 pancreatic resections performed at our institution, five patients (0.8%) were found to have solid pseudopapillary tumor of the pancreas. All patients were female with a mean age of 39.2 years. Abdominal pain was the presenting symptom in four patients. MRI demonstrated pancreatic head involvement in three patients and pancreatic tail involvement in one patient. CT scan was used to diagnose one tumor located in the body of the pancreas. Mean tumor size was 8.0 cm. **Surgical**

treatment consisted of two open pancreaticoduodenectomies, one laparoscopic pancreaticoduodenectomy, one laparoscopic subtotal pancreatectomy and one laparoscopic distal pancreatectomy. Surgical pathology revealed no evidence of vascular invasion in any of the tumors and an R0 resection complete resection with no microscopic residual tumor was obtained in all patient. No patients had involved lymph nodes with a mean of 16 lymph nodes inspected. No complications from any pancreatic resection were observed. **Conclusion:** Solid pseudopapillary tumors of the pancreas are a rare but treatable pancreatic tumor. Complete surgical excision is the treatment of choice and can be achieved through an open or minimal access technique.

Keywords: Solid pseudopapillary tumor, Pancreas, Pancreaticoduodenectomy

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INTRODUCTION

First described by Franz in 1959, solid pseudopapillary tumor of the pancreas (SPT) is a rare, low-grade malignant tumor of unknown etiology accounting for 0.2-2.7% of all primary pancreatic

tumors [1–3]. It is also known as solid cystic papillary epithelial tumor, papillary cystic tumor, solid and cystic tumor, papillary-cystic neoplasm, Hamoudi or Franz tumor. SPT is most prevalent in women of younger age [1, 4–9]. 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 [2, 6, 10–13].

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 [12].

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 [14]. Surgical resection is the treatment of choice for affected patients and is associated with an overall good prognosis.

CASE SERIES

Case 1: A 21-year-old female presented to our institution with persistent abdominal pain for three years after a palliative cholecystojejunostomy for an “inflammatory myofibroblastic pseudotumor of the pancreas” followed by 16 cycles of adjuvant chemotherapy with doxorubicin and ifosfamide. Her symptoms included epigastric pain, jaundice, and decreased appetite. Laboratory analysis was notable only for slight elevation of transaminases (ALT 42 IU/L, AST 30 IU/L). MRI showed a well circumscribed, oval heterogeneous mass 8.9x8.3 cm, in the head of the pancreas, displacing the common bile duct, portal vein and duodenum (Figure 1). Obliteration of the SMV with collateral formation and dilation of the pancreatic duct were noted. An exploratory laparotomy with pancreaticoduodenectomy was performed with resection of SMV and re-implantation into the IVC. Pathology showed a 9.5 cm pancreatic SPT without invasion. No lymph nodes were involved in tumor and immunohistochemical staining was positive for vimentin with patchy staining for synaptophysin and chromogranin. Her postoperative course was uncomplicated and she was discharged home on postoperative day 7. At three months follow up she was without complaints. She was unfortunately lost to follow up after as she returned home to Puerto Rico (Table 1).

Case 2: A 17-year-old female presented with episodic postprandial right upper quadrant pain. Cholecystitis was suspected and an ultrasound was performed showing cholelithiasis and an incidental

pancreatic tail mass eight cm in size. MRI was subsequently performed showing an 8.8x5.8 cm mass in the pancreatic tail with involvement of the splenic artery causing splenomegaly and gastric varices. Endoscopic ultrasound with fine needle aspiration was performed finding a solid and cystic mass in the pancreatic tail consistent with SPT. Laparoscopic subtotal pancreatectomy, splenectomy and cholecystectomy were performed demonstrating a 12.5 cm SPT with extrapancreatic extension. Immunostains were notable for diffusely positive vimentin, CD10 and progesterone, with focal positivity for cytokeratin and synaptophysin. No lymph nodes were involved with the tumor. Her postoperative course was uncomplicated and she was discharged home on postoperative day-9. She has done well at nine months follow up with no signs of tumor recurrence (Table 1).

Case 3: A 52-year-old female with a four month history of postprandial discomfort and nausea with emesis was found to have an incidental 1.6 cm pancreatic body mass on CT scan performed for elevated liver function tests. Endoscopic ultrasound with biopsy was performed showing a 1.6 cm hypoechoic mass in the mid-body of the pancreas with central calcification. Fine needle aspiration was suggestive of SPT. Laparoscopic distal pancreatectomy with en-bloc splenectomy was performed demonstrating a two cm SPT without lymphovascular invasion. Immunostains were strongly positive for CD 10, vimentin and progesterone, with weak positivity for synaptophysin. No immediate complications were observed and she was discharged on postoperative day-2. No further therapy was required and she was without evidence of recurrence at two year follow up (Table 1).

Case 4: A 75-year-old female presented with a one year history of a palpable mass in her right upper quadrant associated with occasional stabbing abdominal pain. She had a history of an open cholecystectomy 30 years prior with the incidental finding of an unresectable “calcified tumor” between the pancreas and the liver. CT scan was performed showing a 12 cm cystic and calcified tumor near the pancreatic head (Figure 2). An ultrasound guided biopsy was performed suggestive of a malignant neuroendocrine tumor. MRI at our institution demonstrated a 16.3x13.9 cm pancreatic head mass with multiple cystic areas and foci of hemorrhage. Displacement of the portal vein and common bile duct was seen without evidence of invasion. An exploratory laparotomy and pancreaticoduodenectomy was performed demonstrating a 15 cm pancreatic head mass consistent with SPT. Immunostains were strongly positive for vimentin and neuron specific enolase with focal reactivity for chromogranin and synaptophysin, carcinoembryonic antigen and CAM5.2. No lymph nodes were involved with tumor. No immediate complications were observed and she was discharged on postoperative day-8. At five year follow up she has required no additional therapy and is without evidence of recurrent disease (Table 1).

Case 5: A 30-year-old female presented to her

outside physician with abdominal pain after exposure to a family member with mononucleosis. Further questioning revealed diarrhea and a 20 pound weight loss over 1–2 months. An ultrasound was performed showing splenic calcifications and subsequently a CT scan noting a 1.1 cm mass in the pancreatic head. CT-guided biopsy was performed with tumor staining positive for alpha-1-antitrypsin, beta-catenin, CD10, vimentin, progesterone receptor and CD56. This was consistent with a diagnosis of SPT. She was referred to our institution for resection. MRI was performed for preoperative planning which confirmed a 1.2 cm lesion in the pancreatic head with high signal intensity on T2 and contrast enhancement on delayed images consistent with her outside diagnosis. A replaced right hepatic artery was incidentally identified without evidence of additional disease. A laparoscopic pancreaticoduodenectomy was performed, finding a 1.2 cm pancreatic head SPT with 0/7 involved lymph nodes

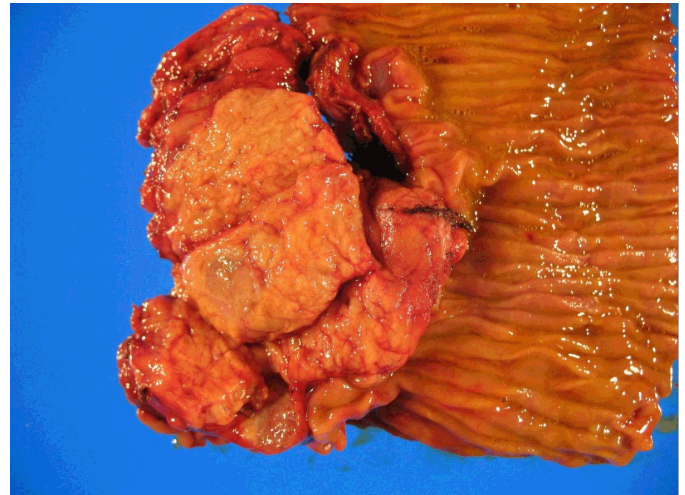


Figure 3: Patient 5 gross specimen demonstrating a well-circumscribed 1.2 cm nodule. Microscopy showed a fibrous capsule, histologically with mixed solid and cystic components consistent with SPT.



Figure 1: Abdominal MRI coronal view of patient 1 demonstrating a heterogeneous, well circumscribed 8.3x7.7x8.9 cm mass in the head of the pancreas with internal cystic structure and high T1 signal intensity suggesting hemorrhagic debris, consistent with SPT.

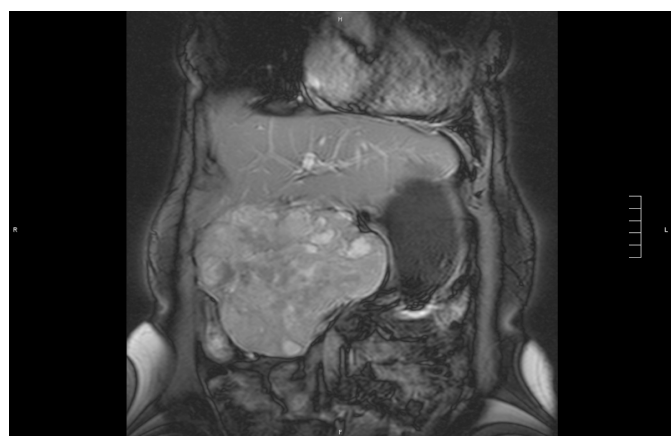


Figure 2: Patient 4 abdominal MRI coronal view demonstrating a 16.3x10.2x13.9 cm lobulated, heterogeneous, but well-circumscribed mass at the level of the pancreatic head containing multiple cystic areas and discrete foci of hemorrhage consistent with SPT.

(Figure 3). No immediate complications were observed and she was discharged on postoperative day 6. At one month follow up she is progressing well, with plans to undergo a six month follow up MRI (table 1).

DISCUSSION

Solid pseudopapillary tumor (SPT) of the pancreas is a rare neoplasm with a generally good prognosis after complete surgical excision. Earlier detection and increased awareness of these neoplasms has led to their increased prevalence in recent years [15]. SPT has a particularly high prevalence in females younger than 35 years of age. In the largest retrospective review conducted to date including 718 patients with SPT, more than 90% of patients were female with an average age of 22 years [6–4]. Female predominance has been attributed to the proximity of primordial pancreatic cells to the ovarian ridge during development [16]. 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 [10, 17].

Radiographic features include a well encapsulated, hypodense mass with various solid and cystic components on CT scan [12]. Ultrasound demonstrates a homogenous or heterogenous encapsulated mass composed of both solid echogenic and hypoechogenic components [18-19]. MRI features include a well defined lesion with high or low signal intensity on T1 and high signal intensity on T2 [12]. MRI is preferable to CT scan for demonstrating the presence of a capsule, hemorrhage, or cystic degeneration [12].

The tumor may involve any portion of the pancreas but the head and tail are most common [3, 6]. Adjacent

Table 1: Patient characteristics of all five patients.

Pt	Sex	Age	Symptoms	Radiographic findings	Operation performed	Tumor size (cm)	LN	Immunostains
1	F	21	Abdominal pain	MRI pancreatic head mass with CBD and PV displacement, SMV obliteration	Open PD with SMV resection and reimplantation	9.5	0/20	vimentin+, synaptophysin+, chromogranin+,
2	F	17	Postprandial abdominal pain	MRI pancreatic tail mass draping over splenic artery	Lap SP, splenectomy, cholecystectomy	12.5	0/29	vimentin+, CD 10, + PR+
3	F	53	Abdominal pain, elevated LFTs	CT midbody pancreatic mass	Lap DP, en bloc splenectomy	2.0	0/16	vimentin+, CD10+, PR+
4	F	75	Incidentally found during cholecystectomy	MRI pancreatic head mass with PV and CBD displacement	Open PD	15	0/7	keratin+, NSE+
5	F	30	Abdominal pain, diarrhea, weight loss	CT/MRI pancreatic head mass, replaced right hepatic artery	Lap PD	1.2	0/7	vimentin+, PR, + CD 10+, CD 56+, β -catenin+, α 1AT+

Abbreviations: LN - Lymph node, α 1AT - alpha-1 antitrypsin, CBD - common bile duct, CD - cluster of differentiation, DP - distal pancreatectomy, Lap - laparoscopic, LFTs - liver function tests, PD - pancreaticoduodenectomy, NSE - neuron specific enolase, PR - progesterone receptor, PV - portal vein, SMV - superior mesenteric vein, SP - subtotal pancreatectomy.

structures such as the mesenteric vessels, stomach, and duodenum may also be involved [10]. In the above study of 718 patients with SPT, pancreatic tail involvement was seen in 35.9% of patients, 34% of tumors were found in the pancreatic head, and 10.3% were found in the body and tail [6]. Extrapancreatic tumors were noted in approximately 1% of patients [6].

Grossly, the tumor is demarcated from adjacent pancreatic tissue by the presence of a fibrous capsule [10, 12]. 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 [10]. Characteristic findings include the presence of solid areas alternating with pseudopapillary formations, foamy histiocytes, nuclear grooves and cytoplasmic globules [15].

Tumor markers observed in SPT including APT/ β -catenin, cyclin D1 and D3, vimentin, antitrypsin, NSE, and progesterone [12, 16, 20–24]. In a study by Kosmahl et al. the most consistent tumor markers present in 90% of their patients with SPT were vimentin, alpha-1-antitrypsin, NSE and progesterone [16]. Other authors have noted that immunoreactivity to β -catenin is found in the cytoplasm and nuclei of almost all SPT's [23–24].

Malignant transformation occurs in 15% of adults and 13% of children, with a greater risk of malignant transformation noted in males and the elderly [12, 25]. Angioinvasion, perineural invasion and deep invasion of pancreatic tissue characterize malignancy [12]. Tumor recurrence has been found in 5–7% of patients after surgical resection [6, 15]. Metastases are most common

to liver, lymph nodes, and peritoneum and are found in 10–15% of patients [12, 15, 17, 26]. Poor prognostic factors include the presence of vascular invasion, tumor size greater than five cm and low nuclear grade [15, 26, 27]. Five year survival after complete resection is 94–100% [6, 15, 17].

Surgery is the only definitive treatment for SPT with a cure rate of greater than 95% with complete resection [28–34]. In contrast to other pancreatic tumors, aggressive surgical resection is warranted even in the presence of local invasion, recurrence, or limited metastases [27, 32–33, 35–36]. En bloc resection of tumor with formal lymphadenectomy can be undertaken in addition to resection of synchronous or metachronous metastases [14]. Surgical debulking can be effective for metastatic disease involving the liver [28–29, 37–39]. In contrast to other pancreatic tumors, invasion of the portal vein or superior mesenteric artery does not indicate tumor unresectability [40]. Pancreatic fistula is the most common complication after complete resection seen in approximately 7% of patients [11]. 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 [41–44]. The tumors' high rate of resectability limits the need for adjuvant therapy [41].

CONCLUSION

Solid pseudopapillary tumors of the pancreas are a

rare but treatable pancreatic tumor. While clinical signs and symptoms are relatively non-specific, characteristic findings on imaging and histology separate these tumors from the more malignant pancreatic tumors. Complete surgical excision is the treatment of choice and can be achieved through an open or minimal access technique.

Author Contributions

Cameron D Adkisson – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Adam S Harris – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Mellena D Bridges – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Justin H Nguyen – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Horacio J Asbun – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

John A Stauffer – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of Submission.

Conflict of Interest

The authors declare no conflict of interest.

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