Solitary Fibrous Tumor of the Pancreas

Analysis of 9 Cases With Literature Review

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Abstract: Solitary fibrous tumor (SFT) has been increasingly reported in various anatomic sites. However, it is still extremely rare in the pancreas. Herein, we present the first series of primary pancreatic SFTs. Nine cases of primary pancreatic SFTs were analyzed. The mean age was 60 years (36 to 76 y) with no sex predilection. Six tumors were in the head, 3 were in the tail. On imaging studies, tumors were described as a hypervascular mass, 2 revealed cystic areas, and 3 were favored to be neuroendocrine tumors. On biopsy, 2 cases were diagnosed as atypical spindle cell tumor; one was misdiagnosed as suspicious for sarcoma, and another case as metastatic renal cell carcinoma. Two were diagnosed as low-grade sarcoma and low-grade stromal tumor on frozen sections. Grossly, tumors were well-demarcated with a median size of 4 cm (0.9 to 15 cm). Microscopically, they were composed of ovoid to spindle tumor cells with no significant mitotic activity and were arranged in alternating hypercellular and hypocellular areas. Staghorn-like vessels and entrapped pancreatic parenchyma were also detected within all tumors. Tumor cells revealed diffuse/strong nuclear STAT6 expression in 7 of 8, CD34 in 7 of 9, and bcl-2 in 4 of 4 tested cases. One tested tumor harbored *NAB2-STAT6* fusion. Eight patients with available follow-up data were free of disease at a mean follow-up of 76 months (3 to 189 mo). SFT should be considered in the differential diagnoses of mesenchymal neoplasms of the pancreas. Immunohistochemical nuclear STAT6 expression is a characteristic feature of SFT. Primary pancreatic SFTs seem to have favorable biological behavior in our series.

Key Words: solitary fibrous tumor, pancreas, pancreatic mesenchymal tumor

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Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm with variable histology and biological behavior. Even though pleura is the most site of common origin, SFT may arise in any anatomic site including abdominal organs. Extrapleural SFTs occur in adults without sex predilection. They are either discovered incidentally or present with nonspecific symptoms.[1,2] Rarely, they cause *paraneoplastic hypoinsulinaemic hypoglycemia* due to insulin-like growth factor production (Doege-Potter syndrome).[3–8] Macroscopically, these tumors are often well-circumscribed, multinodular, and firm. Morphologically, they are characterized with patternless distribution of ovoid to spindle-shaped tumor cells, stromal hyalinization, and hemangiopericytoma-like vessels.[1,2] They harbor *NAB2-STAT6* gene fusion,[9,10] which leads to highly specific nuclear STAT6 expression by immunohistochemical staining.[11] Although the vast majority of SFTs have a benign prognosis, about 10% may behave aggressively with local recurrences or distant metastasis. As per the current (2019) WHO, features related with aggressive behavior include older patient age, larger tumor size, high cellularity, cytologic atypia, >4 mitosis/2 mm^2, hemorrhage, necrosis, and sarcomatous transformation.[2]

Pancreas is an exceedingly rare localization for SFT. The first primary pancreatic SFT case was described in 1999 by Lüttges et al.[12] Since then, only single case reports have been described in the literature.[12–44] In this study, we present the first series of primary pancreatic SFTs, with

the aim of further defining their clinicopathologic features and challenging differential diagnoses.

MATERIALS AND METHODS

Surgical pathology databases of the authors' institutions were searched for cases with a diagnosis of primary pancreatic SFT. Available gross photographs, descriptions, and histologic sections were reevaluated to confirm the diagnosis and further characterize the morphologic and immunohistochemical findings. Available medical records including radiology reports were reviewed to obtain clinical data including age, sex, presenting symptoms, tumor location, presence of prior biopsy or frozen section, surgical procedure type, and outcome.

For all cases, the following histopathologic information was recorded: tumor size; lymph node status; surgical margin status; growth pattern (as infiltrative or expansile); degree of cellularity (as low, moderate, high) and pleomorphism (as low, moderate, high); mitotic rate (by counting 10 fields at ×400 on an Olympus microscope = 0.45 mm^2); presence of necrosis, hemorrhage, myxoid changes, and entrapped pancreatic parenchyma. Prognostic risk stratifications were determined by applying 3 different classification system: WHO 2019 criteria, Pasquali's recurrence risk model (Supplemental Table 1, Supplemental Digital Content 1, http://links.lww.com/PAS/B611), and Demicco's modified 4-variable risk stratification model for the development of metastasis (Supplemental Table 2, Supplemental Digital Content 2, http://links.lww.com/PAS/B612).[2,45,46]

A representative formalin-fixed paraffin-embedded tissue section of the cases, for which a paraffin block was available, was immunolabeled using the standard avidin-biotin-peroxidase method with STAT6 (EP325; Cell Marque) antibody and nuclear staining was accepted as a positive result. Results of other immunohistochemical and as well as electron microscopy studies were recorded from the original pathology reports.

One case, for which additional material was available, was subjected to a custom targeted, RNA-based panel (MSK-Fusion) that utilizes Archer Anchored Multiplex PCR technology and next-generation sequencing to detect gene fusions in 62 genes (including *NAB2* and *STAT6*) known to be involved in chromosomal rearrangements.[47–49] This custom assay has been validated and approved for clinical use at Memorial Sloan Kettering Cancer Center by the New York State Department of Health Clinical Laboratory Evaluation Program.

RESULTS

We identified 9 cases all of which were surgical resections.

Clinical Features

The mean patient age was 60 years (range, 36 to 76 y). Five patients were male and 4 were female. Four patients presented with abdominal pain, one of these patients also had weight loss, and another patient presented with back pain. The other 3 tumors were detected incidentally during workup for other conditions. None of the patients presented with hypoglycemia. One patient had experienced acute pancreatitis 6 years before the diagnosis of SFT.

An incidental intestinal phenotype ampullary adenocarcinoma (1 cm in greatest dimension, pT1) was found synchronously in 1 patient. Another patient had 2 separate incidental well-differentiated WHO 2019-Grade 1 pancreatic neuroendocrine tumors (2 and 0.8 cm in greatest dimensions), and a neuroendocrine microadenoma (0.2 cm). Three patients had a history of other neoplastic conditions including meningioma, renal cell

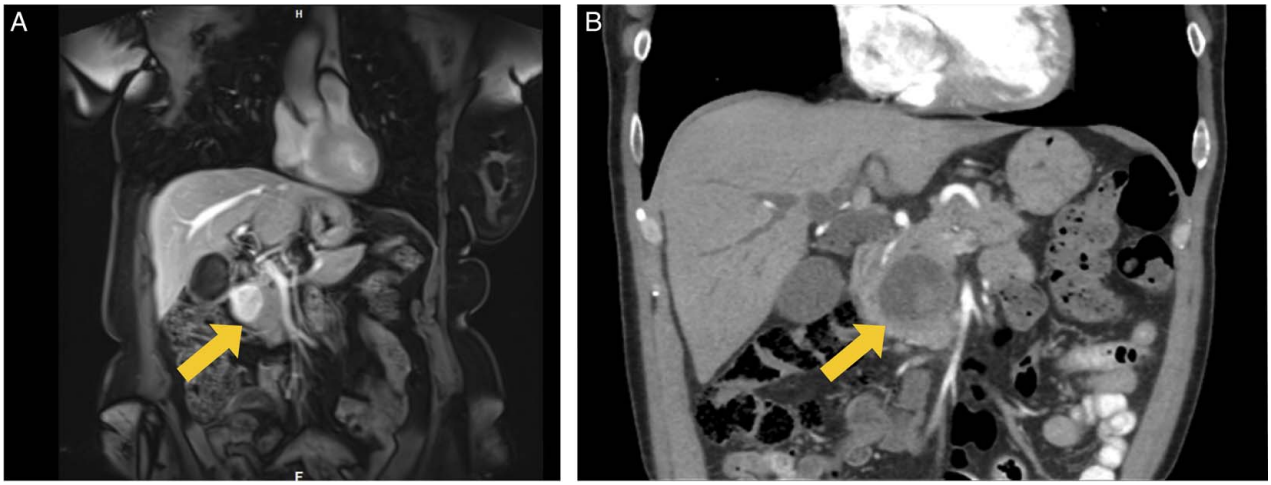

FIGURE 1. Coronal magnetic resonance imaging of case #3 demonstrated a hypointense solid mass (arrow) centered in the head of the pancreas. No dilatation of the main pancreatic duct was noted. A neuroendocrine tumor was favored (A). Coronal computed tomography image of case #4 revealed a heterogeneous cystic lesion (arrow) with central solid component in the pancreatic head, with biliary ductal dilatation. The lesion was reported as "consistent with a cystic malignancy" (B).

carcinoma, breast carcinoma, and cutaneous basal cell carcinoma.

Two of the 8 tumors with available imaging results were described as a cystic lesion, while the others were described as well-circumscribed solid mass. Tumors were described as a hypervascular mass showing persistent enhancement on delayed phase imaging of dynamic computed tomography or magnetic resonance imaging studies (Fig. 1). Three were favored to be neuroendocrine tumors. Other radiologic differential diagnoses included solid pseudopapillary neoplasm, inflammatory conditions, and cystic malignancies.

One case had a biopsy diagnosis of "atypical spindle cell tumor, suspicious for sarcoma" and it was diagnosed as "spindle cell proliferation, favor low-grade sarcoma" on frozen section. Another case was favored to be a "low-grade stromal tumor" on frozen section. Fine needle aspiration of another case was interpreted as metastatic renal carcinoma based on immunohistochemical PAX8 expression and patient's history of renal cell carcinoma.

None of the patients received neoadjuvant chemotherapy. All patients underwent surgical resection (6 pancreaticoduodenectomy, 3 distal pancreatectomy).

Clinical features of the patients are summarized in Table 1.

Pathologic Features

Six tumors were in the head of the pancreas, and 3 were in the tail. Grossly, tumor size varied from 0.9 to 15 cm (median, 4 cm). Tumors were described as well-demarcated, firm, solid lesions with white-tan cut surfaces. Three tumors revealed cystic degeneration (Fig. 2).

Microscopically, sections revealed relatively well-circumscribed tumors with lobular appearance and expansile borders (Fig. 3), although, in some areas, extension to adjacent pancreatic parenchyma was noted. Moreover, on close examination entrapped pancreatic parenchyma was seen both at the periphery and within the tumor in all cases (Fig. 4). In general, the tumors were characterized by so-called patternless pattern and

TABLE 1. Clinicopathologic Features of Our Cases

Case no.	Age, sex	Symptom (s)	Radiologic findings/differential diagnosis	Location	Size (cm)	Cellularity, pleomorphism, necrosis, mitotic count	Immunohistochemical findings	Outcomes, follow-up (mo)
1	72, M	Abdominal pain	Solid hypervascular mass/ PanNET, focal pancreatitis	Head	5	Low, low, no, 3/10 HPF	*Positive:* STAT6, bcl-2, vimentin *Negative:* CD34, CD117, ALK, SMA, CMA, desmin, S100, AE1/AE3, CAM5.2, ER, PR, inhibin	NED, 17
2	76, F	Back pain	Solid hypervascular mass/ PanNET, less likely a thrombosed splenic artery aneurysm	Tail	4	Moderate, low, no, 0/10 HPF	*Positive:* STAT6, bcl-2, focal SMA, focal CMA, focal ER, focal inhibin *Negative:* CD34, desmin, PR	NED, 138
3	65, F	Incidental	Solid hypervascular mass/ PanNET	Head	4	Low, low, no, 0/10 HPF	*Positive:* STAT6, CD34 *Negative:* SMA, desmin, S100, ERG, CD31, CDK4, MDM2, pancytokeratin	NED, 42
4	56, M	Incidental	Cystic mass with solid component/cystic malignancies	Head	3.7	Low, low, no, 0/10 HPF	*Positive:* STAT6, CD34 *Negative:* CD117, CDK4, MDM2, ER, PR	NED, 47
5	36, F	Abdominal pain	Complex cystic mass/ inflammatory conditions, SPN, mucinous neoplasms	Tail	2.7	Low, low, no, 0/10 HPF	*Positive:* STAT6, CD34, SMA *Negative:* Desmin, S100	NED, 189
6	55, F	Abdominal pain	Solid hypervascular mass/ nonductal neoplasms	Head	3	Low, low, no, 0/10 HPF	*Positive:* STAT6, CD34, calponin *Negative:* CD117, DOG1, ALK, SMA, MSA, desmin, S100	NED, 107
7	57, M	Incidental	Solid hypervascular mass/ NA	Head	0.9	Low, low, no, 0/10 HPF	*Positive:* CD34, PAX8, PAX2 *Negative:* CD117, DOG1, S100, pancytokeratin, synaptophysin, chromogranin A	NED, 3
8	56, M	Abdominal pain, weight loss	Solid hypervascular mass/ NA	Head	15	Moderate, low, focal, 1/10 HPF	*Positive:* CD34, bcl-2 *Negative:* CD117, DOG1, SMA, CMA, desmin, S100, CD99, AE1/AE3 *Noncontributory:* STAT6	NED, 62
9	70, M	NA	NA	Tail	13	Low, low, no, <1/10 HPF	*Positive:* STAT6, CD34, bcl-2, CD99 *Negative:* CD117, DOG1, SMA, desmin, S100, HMB45, CD31, ERG, EMA, Claudin, MUC4, synaptophysin, chromogranin A, ER, inhibin, WT1, calretinin	NA

CMA indicates common muscle actin; DP, distal pancreatectomy; EMA, epithelial membrane antigen; ER, estrogen receptor; F, female; M, male; MSA, muscle-specific actin; NA, not available; NED, no evidence of disease; PanNET, pancreatic neuroendocrine tumor; PD, pancreaticoduodenectomy/Whipple procedure; PR, progesterone receptor; SPN, solid pseudopapillary tumor.

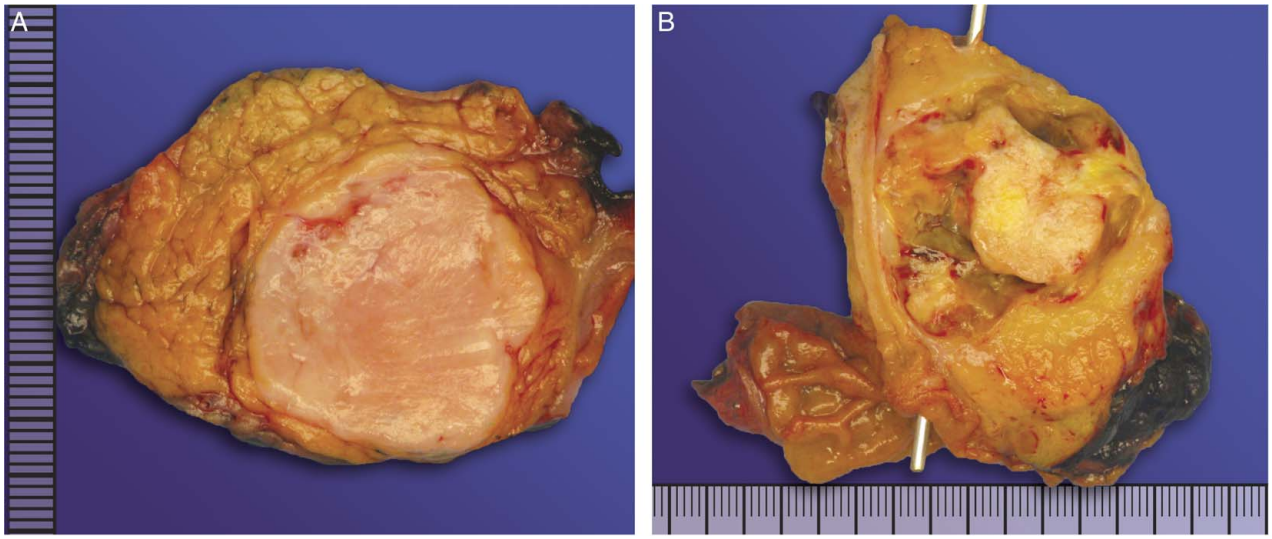

FIGURE 2. The tumors were well-circumscribed, white/tan, solid nodules (A), one of the reasons why these tumors were diagnosed as neuroendocrine tumors or solid pseudopapillary tumor on imaging. Two tumors also showed cystic degeneration (B).

staghorn-like vessels (Fig. 5). Some areas were extremely hypocellular, others were hypercellular and abrupt transition between these hypocellular and hypercellular areas were common (Fig. 6). Loose stroma and myxoid change were seen in all cases. One case had histiocyte accumulations in degenerative areas. Only 1 case (case #8) revealed focal necrosis. On high magnification, the tumors were composed of cytologically bland, ovoid to spindle-shaped tumor cells (Fig. 7). Significant cytologic atypia was not identified in any of the cases and none of the tumors were mitotically very active (no mitosis or ≤ 3 mitotic figures/10 HPFs in any cases).

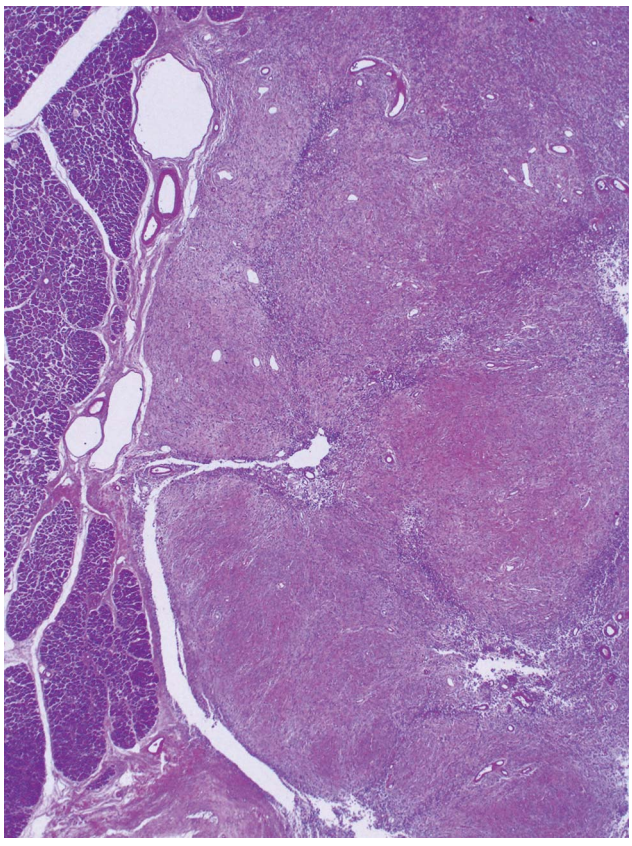

FIGURE 3. The tumors are relatively well-circumscribed with lobular appearance and expansile borders.

Surgical margins were free of the tumor in all cases. No lymph node metastasis was detected, although the case with synchronous ampullary adenocarcinoma revealed metastatic ampullary adenocarcinoma in 1 of 5 lymph nodes (pN1).

Immunohistochemically, 7 of 8 tested tumors revealed nuclear STAT6 expression (Fig. 8A); in the eighth case, STAT6 staining was noncontributory as the tumor was no longer present on the immunohistochemistry slide. However, the tumor had classic morphologic features of SFT and labeled with bcl-2 and CD34, and was negative for AE1/AE3, CD117, DOG1, smooth muscle actin (SMA), desmin, S100, and CD99. Seven (7/9, 78%) tumors were positive for CD34 (Fig. 8B) and 4 (4/4, 100%) tumors were positive for bcl-2.

Electron microscopy reported to demonstrate fusiform and spindle-shaped fibroblast-like tumor cells, with a moderately developed rough endoplasmic reticulum, surrounded by collagen fibers in the only tumor tested (case #1), supporting the diagnosis of SFT.

MSK-Fusion assay revealed *NAB2-STAT6* fusion (*NAB2* exon 4 [NM_005967] and *STAT6* exon 2 [NM_001178078]) in the only tumor tested (case #3).

Pathologic features of the tumors are summarized in Table 1.

Risk Stratification

None of the cases were considered as malignant:

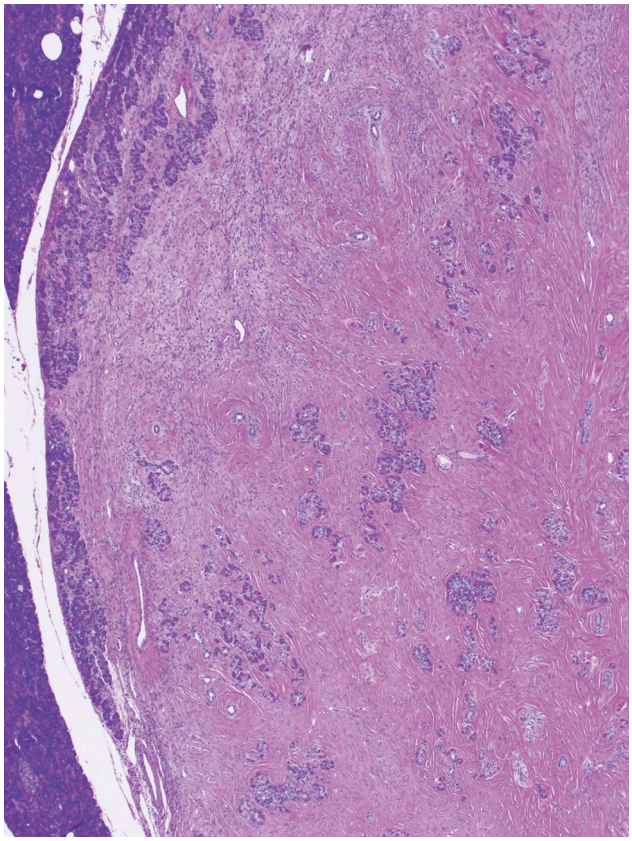

FIGURE 4. Although SFT has expansile borders, there is entrapped pancreatic parenchyma both at the periphery of the tumor and within the tumor.

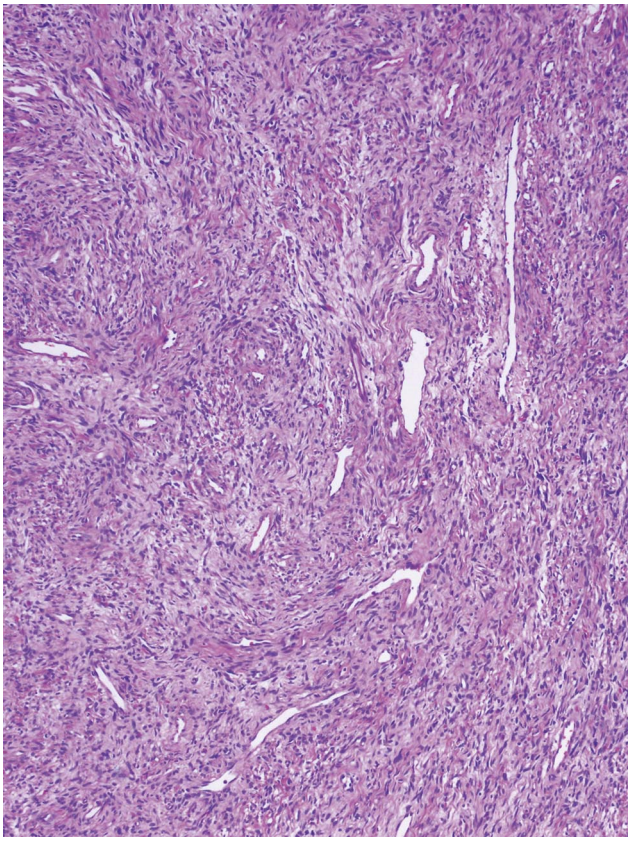

FIGURE 5. SFTs reveal so-called patternles pattern and staghorn-like vessels.

(1) Based on the current (2019) WHO criteria, the tumors are not expected to behave aggressively as none revealed high cellularity, cytologic atypia, >4 mitosis/2 mm^2, hemorrhage, or sarcomatous transformation. Only 1 case (case #8) revealed focal necrosis.[2]
(2) As per Pasquali's SFT recurrence risk model (Supplemental Table 1, Supplemental Digital Content 1, http://links.lww.com/PAS/B611), 7 cases are classified as having very low risk for recurrence, and 2 (case #2 and case #8) were classified as having low risk due to moderate cellularity.[45]
(3) As per Demicco's modified 4-variable risk stratification model (Supplemental Table 2, Supplemental Digital Content 2, http://links.lww.com/PAS/B612), 8 cases are classified as having low risk of metastasis, and 1 case (case #8) is classified as having intermediate risk due to ≥55 age and ≥15 cm tumor size.[46]

Clinical Outcomes

Follow-up information was available for 8 patients (89%). None of the patients received adjuvant therapy and all patients with available survival data are alive with no evidence of disease, with a mean follow-up of 76 months (median, 55 mo; range, 3 to 189 mo).

Literature Analysis

When all the cases in the literature are combined with our cohort (n = 44), the following clinicopathologic characteristics are elucidated: All patients were adults except for 1 pediatric patient who was diagnosed at the age of 14 months. The mean age was 55 years (range, 14 mo to 82 y). There was no sex predilection (female/male = 1). Most of the patients were asymptomatic (n = 19, 44%) or presented with abdominal pain (n = 15, 35%); other symptoms/findings include jaundice (n = 4, 9%), abdominal discomfort (n = 3, 7%), back pain (n = 2, 5%), weight loss (n = 2, 5%), and hypoglycemia (n = 1, 2%). The tumor sizes ranged from 0.9 to 18.5 cm (median, 4 cm). Tumors involved head/neck/uncinate process of the pancreas (n = 25, 57%) more frequently than body/tail (n = 19, 43%). Due to their demarcated round nature, radiologically, the first differential diagnosis was of nonductal tumors, especially pancreatic well-differentiated neuroendocrine tumor, followed by solid pseudopapillary neoplasm and less likely gastrointestinal stromal tumor (GIST). All but 1 patient were surgically treated with different procedures based on tumor location, including enucleation, pancreaticoduodenectomy, or distal pancreatectomy. For 1 patient, surgery was not possible due to comorbidities. The clinicopathologic features of all reported cases are summarized in Tables 2 and 3.

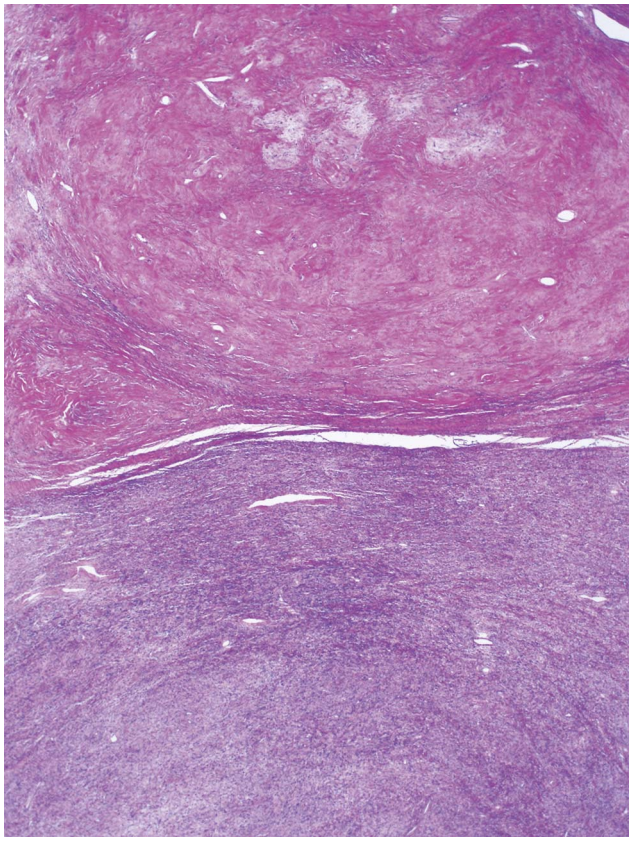

FIGURE 6. Some foci are extremely hypocellular, others are hypercellular and abrupt transition between these hypocellular and hypercellular areas is common.

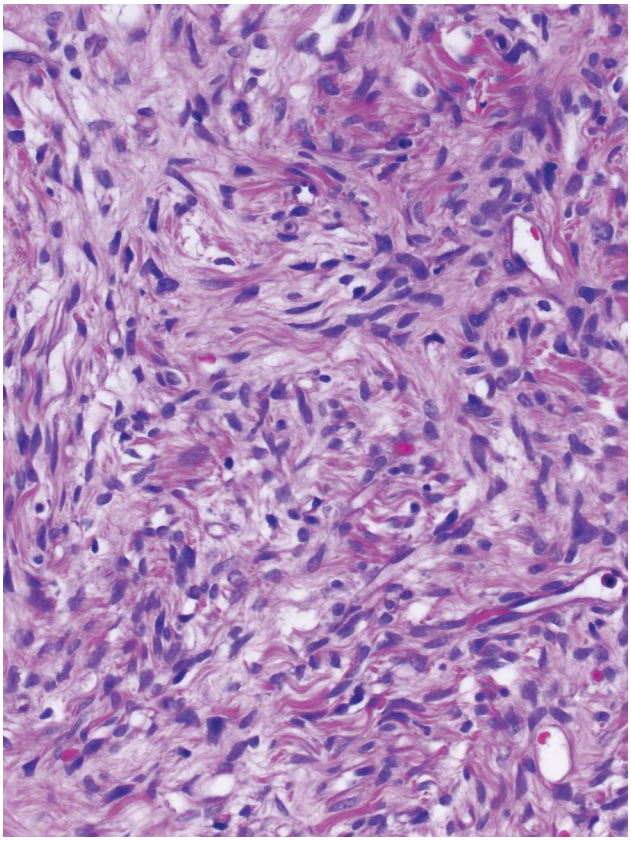

FIGURE 7. On high magnification, the tumors are composed of cytologically bland, ovoid to spindle-shaped cells.

DISCUSSION

Even though primary mesenchymal tumors of the pancreas are rare, various mesenchymal tumor types arising from pancreatic stromal tissue have been reported, including inflammatory myofibroblastic tumor, desmoid tumor, GIST, lymphangioma, cavernous hemangioma, angiomyolipoma, schwannoma, ganglioneuroma, leiomyosarcoma, rhabdomyosarcoma, liposarcoma, angiosarcoma, Ewing sarcoma/primitive neuroectodermal tumor, undifferentiated/unclassified sarcoma.[28,34,50] However, experience with SFT is very limited. Our current understanding of this tumor is

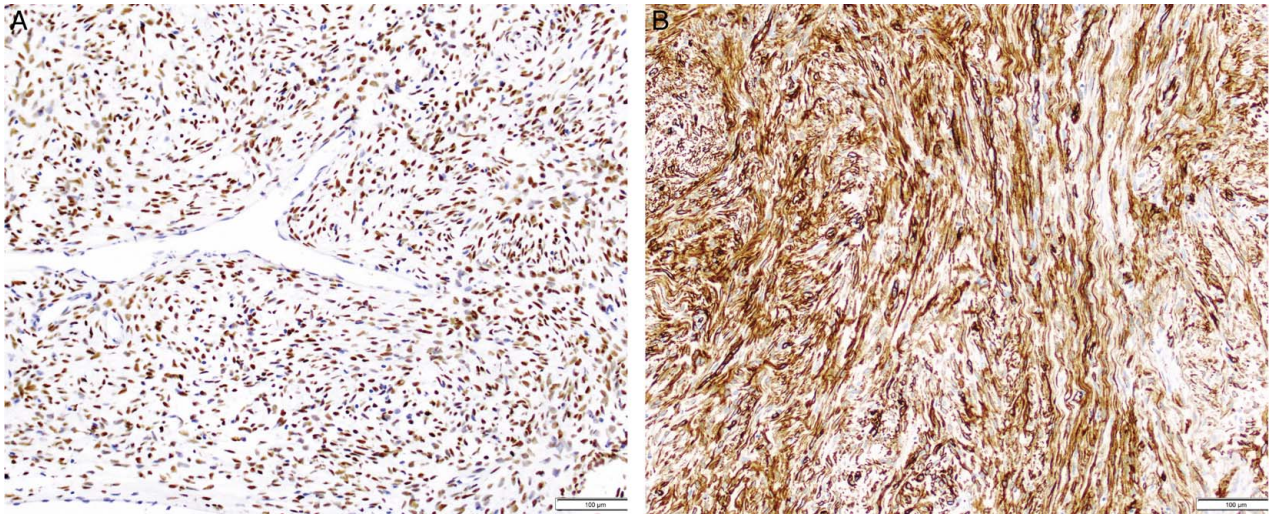

FIGURE 8. Immunohistochemically, all tested tumors revealed STAT6 (nuclear) (A) and seven of nine tumors revealed CD34 (B) expression.

TABLE 2. Summary of Primary Pancreatic SFTs Reported in the Literature

References	Age, sex	Symptom(s)	Radiologic/ clinic differential diagnoses	Location, procedure	Size (cm)	Reported malignancy criteria	Reported diagnosis	Immunohistochemical findings	Outcomes, Follow-up (mo)
Lüttges et al[12]	50, F	Incidental	PanNET	Body, DP	5.5	Moderate pleomorphism, no mitosis or necrosis	SFT	*Positive*: CD34, bcl-2, CD99, vimentin *Negative:* SMA, S100, NSE, cytokeratin, synaptophysin, chromogranin A, insulin, p53	Alive, RF, 20
Chatti et al[13]	41, M	Abdominal pain	PanNET	Body, enucleation	13	Necrosis, no atypia or mitosis	SFT	*Positive*: CD34, bcl-2, focal CD99, focal CD117, focal SMA, vimentin *Negative*: S100, cytokeratin, EMA	Death on postoperative third day due to complications
Gardini et al[14]	62, F	Abdominal pain	PanNET	Head, PD	3	NA	SFT	*Positive*: CD34, bcl-2, CD99, focal SMA, vimentin *Negative*: CD117, desmin, S100	Alive, RF, 16
Miyamoto et al[15]	41, F	Abdominal pain	PanNET	Neck, enucleation	2	No atypia, mitosis, or necrosis	SFT	*Positive*: CD34, bcl-2 *Negative*: CD117, SMA, desmin, S100, AE1/AE3, CAM5.2	Alive, RF, 7
Kwon et al[16]	54, M	Incidental	PanNET, SPN, GIST, neurogenic tumor	Body, median segmentectomy	7.6	No atypia	SFT	*Positive:* CD34, CD99, vimentin *Negative:* CD117, S100, cytokeratin	Alive, RF, 88
Srinivasan et al[17]	78, F	Back pain, weight loss	PanNET	Body, DP	5	No atypia or necrosis, <1/10 HPF mitosis	SFT	*Positive:* focal CD34, bcl-2, CD99, vimentin *Negative:* CD117, CD10, SMA, desmin, S100, CAM5.2, synaptophysin, chromogranin A	Alive, RF, 7
Chetty et al[18]	67, F	Incidental	PanNET	Uncinate process, PD	2.6	No atypia, mitosis, or necrosis	SFT	*Positive:* CD34, bcl-2, CD99 *Negative:* CD117, SMA, desmin, MSA, S100, AE1/AE3, CAM5.2, synaptophysin, chromogranin A, beta-catenin	Alive, RF, 6
Ishiwatari et al[19]	58, F	Incidental	PanNET with cystic changes	Head, PD	3	Atypia (low), 0/10 HPF mitosis, focal necrosis	SFT	*Positive*: CD34, bcl-2 *Negative*: CD99, CD117, SMA, S100, AE1/AE3, EMA, synaptophysin, chromogranin A	Alive, RF, 42
Sugawara et al[20]	55, F	Incidental	NA	Head, PD	7	NA	SFT	*Positive:* CD34 *Negative:* CD117, SMA, S100, ALK, cytokeratin	NA
Azadi et al[21]	57, M	Incidental	NA	Tail, DP	3.1	No atypia or necrosis, low mitotic index	SFT	*Positive*: CD34, bcl-2 *Negative*: CD117, desmin, myogenin, AE1/AE3	NA
dos Santos et al[22]	40, F	Incidental	NA	Body, partial pancreatectomy	3	No atypia, mitosis, or necrosis	SFT	*Positive:* CD34, beta-catenin *Negative*: CD117, SMA, desmin, S100 cytokeratin, EMA	NA
Tasdemir et al[23]	24, F	Epigastric pain	Mesenchymal tumor	Head, enucleation	18.5	1-2/10 HPF mitoses	SFT	*Positive:* CD34, focal bcl-2, vimentin, beta-catenin *Negative*: Desmin, S100, cytokeratin	Alive, RF, 3
van der Vorst et al[24]	67, F	Abdominal pain	PanNET	Uncinate process, enucleation	2.8	No atypia, mitosis, or necrosis	SFT	*Positive*: CD34, bcl-2, CD99 *Negative*: CD117, beta-catenin	NA
Yamanashi et al[25]	50, M	Incidental	PanNEC	Tail, DP	10, 3.5, 3	Intrapancreatic metastases, mild atypia, >2/HPF mitoses, necrosis, hypercellularity	Malignant SFT	*Positive:* CD34, bcl-2, vimentin *Negative:* CD117, cytokeratin, synaptophysin, chromogranin A	Alive, Local recurrence after postoperative 21 mo, 32
Chen et al[26]	49, F	Abdominal pain, distension	PanNET, SPN	Head, PD	13	Necrosis	SFT	*Positive:* CD34, bcl-2, CD68, MSA *Negative*: CD117, CD99, SMA, desmin, S100, cytokeratin	Alive, RF, 30
Hwang et al[27]	53, F	Incidental	PanNET, SPN	Head, partial head resection	5.2	NA	SFT	*Positive:* CD34, bcl-2, CD99, CD10, SMA, ER, PR *Negative*: CD117, DOG1, caldesmon, desmin, S100, AE1/AE3, EMA	Alive, RF, 6
Kim et al[28]	52, F	Incidental	PanNET, SPN, pancreatic cancer	Tail, DP	2	No atypia	SFT		Alive, RF, 12

TABLE 2. (*continued*)

References	Age, sex	Symptom(s)	Radiologic/ clinic differential diagnoses	Location, procedure	Size (cm)	Reported malignancy criteria	Reported diagnosis	Immunohistochemical findings	Outcomes, Follow-up (mo)
Han et al[29]	77, F	Jaundice	PanNET	Head, any surgery was not performed due to patient's surgical history	1.5	No atypia, mitosis, or necrosis	SFT	*Positive:* Focal CD34, bcl-2 *Negative*: CD117, SMA, desmin, S100 *Positive:* CD34, CD99 *Negative*: CD117, S100	Alive, PF with residual tumor, 10
Baxter et al[30]	58, F	Abdominal pain	PanNET, GIST, sarcoma, SPN, SFT, mass forming pancreatitis	Head, PD	3.5	No mitosis or necrosis	SFT	*Positive:* CD34, bcl-2, focal beta-catenin, focal CD99 *Negative*: CD117, SMA, desmin, S100, Melan-A, HMB45, AE1/AE3, CAM5.2, EMA, synaptophysin, chromogranin A, CD56, PR	Alive, RF, 24
Estrella et al[31]	52, F	Jaundice	PanNET	Head, PD	15	Sarcomatous component, nuclear pleomorphism, marked cellularity, up to 17/ 10 HPF mitoses, necrosis	Malignant SFT	*Positive:* CD34, bcl-2, and p53 and p16 in malignant areas *Negative:* CD117, CD99, synaptophysin, chromogranin A	Alive, RF, 40
Murakami et al[32]	82, M	Hypertension, hypercortisolism, hypokalemia, edema	PanNET with ectopic secretion of ACTH	Tail, DP	6	NA	SFT	*Positive:* STAT6, CD34, bcl-2, focal NSE, focal ACTH, focal POMC *Negative:* synaptophysin, chromogranin A	Death after postoperative fourth month due to sepsis
Spasevska et al[33]	47, M	Epigastric pain, jaundice	Cystadenocarcinoma	Head, PD	3.5	Minimal pleomorphism and atypia, 1-2/10 HPF mitoses, no necrosis	SFT	*Positive:* CD34, bcl-2, CD99, vimentin, focal beta-catenin, focal actin *Negative:* CD117, caldesmon, desmin, S100, cytokeratin, EMA	Death after postoperative first week due to complications
Zhang et al[34]	63, M	Incidental	PanNET	Body/tail, DP	3	Marked focal atypia, necrosis, obvious mitosis, hypercellularity	Malignant SFT	*Positive:* CD34, CD99, CD117, SMA, NSE, vimentin. Ki67 index > 30%. *Negative*: F-VII, AE1/AE3, EMA, synaptophysin, chromogranin A, gastrin, VIP, somatostatin	Multiple metastases within postoperative 6 mo, death after postoperative 10th month
	46, M	Abdominal pain	Chronic pancreatitis	Body/tail, DP	4	NA	SFT	*Positive:* CD34, bcl-2, CD99, SMA, focal CD68 *Negative*: CD117, DOG1, caldesmon, desmin, S100, ALK, CD31, CD21, CD35, AE1/AE3	Alive, RF, NA
	43, M	Abdominal discomfort	Castleman's disease	Body/tail, excision	4	NA	SFT	*Positive:* Focal CD34 *Negative*: CD117, SMA, desmin, S100	Alive, RF, NA
Paramythiotis et al[35]	55, M	Incidental	PanNET, SPN, GIST, SFT	Body, resection	3.6	Rare mitosis, necrosis	SFT	*Positive:* CD34, CD99, Bcl-2, vimentin, focal S100 *Negative*: CD117, SMA, desmin, cytokeratin, EMA	Alive, RF, 12
Clare et al[36]	39, F	Incidental	PanNET, SPN	Head, PD	2.2	6/10 HPF mitoses, no necrosis	Malignant SFT	*Positive:* STAT6, CD34, bcl-2, variable expression of AE1/AE3, CAM5.2 *Negative*: CD117, SMA, desmin, S100, MNF116, CK7, CK20, CDX2, synaptophysin, chromogranin A, ER	Alive, RF, 1
D'Amico et al[37]	52, M	Incidental	PanNET	Body, enucleation	2	1/10 HPF mitosis	SFT	*Positive:* STAT6, CD34 *Negative*: NA	Alive, RF, 24
Oana et al[38]	73, M	Abdominal discomfort	PanNET, GIST, ACC	Head, partial pancreatectomy	6.5	No mitosis or necrosis	SFT	*Positive:* CD34, bcl-2 *Negative*: CD117, SMA, desmin, S100, cytokeratin	Alive, RF, 36
Sheng et al[39]	14 mo, M	Obstructive jaundice	NA	Head, PD	2	Duodenal invasion, mild to moderate pleomorphism, 2-5/ 10 HPF mitoses in hypercellular area, no necrosis	SFT with low-grade malignancy	*Positive:* CD34, focal SMA, vimentin *Negative:* Bcl-2, CD117, CD99, desmin, myogenin, S100, ALK, cytokeratin, EMA,	Alive, RF, 12

TABLE 2. (*continued*)

References	Age, sex	Symptom(s)	Radiologic/ clinic differential diagnoses	Location, procedure	Size (cm)	Reported malignancy criteria	Reported diagnosis	Immunohistochemical findings	Outcomes, Follow-up (mo)
								synaptophysin, chromogranin A, CD56	
Afzal et al[40]	43, M	Incidental	PanNET, GIST	Head/neck, PD	12.7	Moderate pleomorphism in hypercellular area, no increased mitosis, no necrosis	SFT	*Positive*: STAT6, focal CD34, bcl-2, CD99, CD117 *Negative*: DOG1, SMA, desmin, S100, SOX10, ERG, CD31, pancytokeratin, synaptophysin, beta-catenin	NA
Geng et al[41]	48, M	Hypoglycemia	NA	Body, DP, liver metastasectomy	6.5	Liver metastases, pleomorphism, 4-5/ 10 HPF mitoses, focal necrosis	Malignant SFT	*Positive:* STAT6, CD34, bcl-2, CD31, D2-40 *Negative*: CD117, SMA, desmin, S100, GFAP	Alive, PF with residual liver tumor, 6
Li et al[42]	61, M	Abdominal pain	Pancreatic cancer, GIST	Body, DP	11	Splenic vein invasion, 25/10 HPF mitoses in hypercellular area	Malignant SFT	*Positive:* STAT6, CD34, focal cytokeratin, focal EMA. Ki67 index; 20%. *Negative:* CD117, DOG1, desmin, S100, TLE-1, beta-catenin, CD21, CD35, CD23	Alive, RF, 4
Taguchi et al[43]	60, M	Palpable mass	PanNEC, GIST, SFT	Head, PD	8	Venous invasion, duodenal invasion, 12/ 10 HPF mitoses, necrosis, hypercellularity	Malignant SFT	*Positive:* STAT6 (weak), CD34, bcl-2, focal AE1/AE3, vimentin *Negative*: CD117, DOG1, SMA, desmin, S100, synaptophysin, chromogranin A	Alive, RF, 12
Liu et al[44]	54, F	Incidental	Benign or low-grade malignant pancreatic tumor	Head, duodenum-preserving pancreatic head resection	3.1	No atypia or necrosis, 0-2/10 HPF mitosis	SFT	*Positive:* STAT6, CD34, CD99 *Negative:* CD117, SMA, desmin, S100, EMA, cyclin-D1	Alive, RF, 6

ACC indicates acinar cell carcinoma; ACTH, adrenocorticotropic hormone; DP, distal pancreatectomy; EMA, epithelial membrane antigen; F, female; GFAP, glial fibrillary acidic protein; M, male; MSA, muscle-specific actin; NA, not available; NSE, neuron-specific enolase; PanNEC, pancreatic neuroendocrine carcinoma; PanNET, pancreatic neuroendocrine tumor; PD, pancreaticoduodenectomy/Whipple procedure; PF, progression-free; POMC, proopiomelanocortin; RF, recurrence-free; SPN, solid pseudopapillary tumor; VIP, vasoactive intestinal polypeptide.

mainly based on individual case reports.[12–44] In this study, we analyzed 9 cases.

Our findings, in combination with the previously published cases, reveal that most primary pancreatic SFTs occur in older adults (mean age, 55 y) without any sex predilection. The patients are either asymptomatic or present with nonspecific findings such as abdominal pain or back pain. Jaundice is rare. The tumors are usually located in the head/neck with a median size of 4 cm. Due to their well-circumscribed nature, clinically they are frequently diagnosed as pancreatic well-differentiated neuroendocrine tumors.

Gross appearance of mesenchymal tumors is not distinctive either and mimic not only nonductal epithelial tumors but also each other. Microscopically, SFTs may have variable histology. Therefore, distinguishing SFT from other mesenchymal tumors, which show spindle cell morphology could be even more challenging. Main microscopic differential diagnoses include desmoid tumor/ fibromatosis, inflammatory myofibroblastic tumor, schwannoma, GIST, and leiomyoma. Needless to mention, melanoma should always be considered in the differential diagnoses of spindle cell lesions.[2] Of note, although pancreatic well-differentiated neuroendocrine tumor is considered as the main presurgical differential of SFT, microscopically they are usually easier to distinguish due to their more cellular epithelial nature and unique chromatin pattern. When in doubt, immunohistochemical staining with neuroendocrine markers such as chromogranin A and synaptophysin would be helpful.[51]

In contrast to the well-circumscribed nature of the SFTs, desmoid tumors are infiltrative and are characterized with long fascicles of the spindle or stellate cells. Although prominent vasculature is present, there are no staghorn-like vessels. Similarly, inflammatory myofibroblastic tumors are composed of loose fascicles of uniform, plump spindle cells. There is also inflammatory infiltrate, predominantly composed of lymphocytes and plasma cells, a feature not common in SFTs. Schwannomas reveal loose fascicles of spindle cells with eosinophilic cytoplasm and tapering nuclei. When present, characteristic Antoni A and B areas are helpful for establishing the right diagnosis. GISTs seen in the pancreas are usually spindle cell type, closely mimicking SFTs. Similarly, leiomyomas are characterized with long intersecting fascicles of spindle cells.

Fortunately, immunohistochemical studies are helpful for tumor classification. Although bcl-2, CD34, and CD99 antibodies have been widely used for SFT diagnosis, these nonspecific stains are not only positive in other tumors but can also be negative in SFTs. In our series, while bcl-2

TABLE 3. Comparison of Our Series and the Literature

	Our series	Literature
Mean age (y)	60	54
Male/female	5/4	17/18
Symptoms	Abdominal pain (n = 4), back pain (n = 1), weight loss (n = 1)	Abdominal/epigastric pain (n = 10), jaundice (n = 4), abdominal discomfort/distension (n = 3), back pain (n = 1), weight loss (n = 1), palpable mass (n = 1)
Head-neck/body-tail	6/3	19/16
Median tumor size (cm)	4	4
Benign/malignant	9/0	28/7
Mean follow-up (mo)	76	17
Status	All are alive with NED	23 are alive with NED 2 are alive with residual disease 1 is alive with local recurrence 2 died due to postoperative complications 1 died due to malignant metastatic SFT 1 died due to sepsis

NED indicates no evidence of disease.

expression was observed in all stained cases, CD34 was negative in 22%. However, after the discovery of *NAB2-STAT6* fusion as the hallmark of SFTs,[9,10] subsequent studies have demonstrated that STAT6 is a reliable immunohistochemical marker for detecting this genetic alteration with very high sensitivity and specificity regardless of anatomic site and morphologic features.[52–56] Like these studies, all our cases revealed nuclear STAT6 expression, including the 2 CD34 negative cases. Of course, it should be kept in mind that STAT6 may be positive in a small subset of dedifferentiated liposarcomas, most likely due to the close location of *STAT6* and *MDM2* on chromosome 12.[52,54] However, the presence of well-differentiated liposarcoma component and positive MDM2 and CDK4 staining can be helpful to distinct dedifferentiated liposarcoma from SFT.

In contrast to SFTs, desmoid tumors are characterized with nuclear β-catenin expression due to *CTNNB1* gene activating mutations and may reveal SMA and/or desmin labeling.[57] Inflammatory myofibroblastic tumors harbor *ALK* or *ROS1* fusions and are characterized with corresponding ALK or ROS1 expression.[58] They are also usually positive for SMA, desmin, and less frequently keratin. Although schwannomas may express CD34, unlike SFTs, they are also positive for S100, glial fibrillary acidic protein, and nestin.[59] Similarly, GISTs may express CD34. However, since most GISTs harbor activating mutations of *KIT* or *PDGFR*, they also label with CD117 and/or DOG1.[60] Leiomyomas are positive for SMA, desmin, caldesmon, and calponin.[61] Finally, melanomas are positive for S100, HMB45, Melan-A, and SOX10.[62]

In addition to these well-known entities, the most challenging differential diagnosis in the pancreas is sclerosing epithelioid mesenchymal tumor, a recently described novel entity specific to the pancreas.[63] Similar to SFTs, these tumors are also well-circumscribed and solid, and the density of neoplastic cells is significantly different throughout the tumor. The neoplastic cells also exhibit variable morphology. Spindle cells with irregular, hyperchromatic nuclei closely mimic SFT. However, presence of epithelioid cells containing scant cytoplasm and round to oval nuclei with open chromatin, and lack of staghorn-like vessels are helpful features. More importantly, sclerosing epithelioid mesenchymal tumors are only positive for vimentin, CD99, keratin (CK18) and extensive molecular testing failed to identify any specific mutation or fusion in these tumors, although they have a distinct methylation profile.[63]

While most of SFTs have favorable course, about 10% of the cases reported to act aggressively with local recurrence or distant metastasis.[1,2] Although various risk stratification models have been proposed to predict the behavior of SFTs, there is still uncertainty for the most useful assessment method in daily practice. The most used parameters include the patient's age, anatomic site, tumor size, mitotic count, cellularity, pleomorphism, hemorrhage/necrosis, surgical margins status, and presence of sarcomatous transformation/dedifferentiation,[45,46,64–71]. Subjectivity, lack of reproducibility, and absence of absolute cutoff values for the assessment of cellularity, cellular atypia, and necrosis cause difficulties to determine malignancy potential of tumor. Also, tumors originated from different anatomic sites and the use of different outcome measures have led to inconsistencies between studies.[3,45,46,72,73] In the current (2019) WHO Classification, older patient age, larger tumor size, presence of hypercellularity, cytologic atypia, mitotic count >4 per 2 mm^2, hemorrhage, necrosis, and sarcomatous transformation are described as the features related with aggressive behavior.[2] In the largest study including 243 extrapleural and nonmeningeal SFTs, Pasquali et al[45] reported that mitotic activity, hypercellularity, and pleomorphism correlate with recurrence and interestingly, larger tumor size is a better prognostic feature. Based on these, they developed a SFT recurrence scoring system that has very low-risk, low-risk, intermediate-risk, and high-risk categories (with 100%, 88.9%, 65.9%, and 52.6% disease-free survival rates at 5 y, respectively).[45] Subsequently, Demicco et al[46] by scoring patient age, tumor size, mitotic count, and tumor necrosis but not cellularity

and pleomorphism, categorized nonmeningeal SFTs as having low, intermediate, and high risks (with no metastasis at 10 y, 10% risk of metastasis at 10 y, and 73% risk of metastasis at 5 y, respectively). None of our 9 SFTs were considered malignant based on WHO malignancy criteria, while 8 revealed low risk and 1 (case #8) revealed intermediate-risk for metastasis as per Demicco's model. The patient with intermediate-risk SFT is alive with no evidence of disease at 62 months' follow-up. However, among previously published primary pancreatic SFTs, there are 7 cases that were reported as a malignant SFT.[25,31,34,36,41–43] These tumors were characterized with marked cellularity (reported in 5 of 7 cases), nuclear pleomorphism and/or atypia (reported in 4 of 7 cases), necrosis (reported in 5 of 7 cases) and frequent (up to 25/10 HPF) mitoses (reported in all 7 cases). Only one of these patients developed multiple metastases within 6 months and succumbed to the disease 10 months after surgery. However, this patient's tumor is reported to be positive for CD34 and CD117.[34] Therefore, the diagnosis appears to be questionable. One patient presented with multiple liver and bone metastases at diagnosis and is alive with disease at 6 months' follow-up[41]; another one with intrapancreatic metastases at the time of the diagnosis developed local recurrence after postoperative 21 months is alive at 32 months' follow-up.[25] The other 4 patients, including one with sarcomatous component in the tumor, are alive with no evidence of disease at 1, 4, 12, and 40 months' follow-up.[31,36,42,43]

The clinicopathologic features of all the reported cases are summarized in Tables 2 and 3.

Of note, recently, it has been suggested that hypoglycemia is also a high risk for unfavorable prognosis.[3] In our study, none of the cases were found hypoglycemic. However, one of the known primary pancreatic SFTs presented with recurrent incidences of hypoglycemia without a history of any endocrine disease.[41] As mentioned above, the patient was eventually diagnosed with a malignant SFT of the pancreas with metastases to the liver and bone as well as Doege-Potter syndrome.

In addition, recent studies suggest that the presence of *TERT* promoter mutation and/or *p53* mutation in SFTs may be associated with malignant behavior.[74–77] Demicco and colleagues also reported that *TERT* promoter mutations correlate with older age, larger tumor size, presence of necrosis, and development of metastasis, but they have no impact on overall survival and disease-specific survival. As such, *TERT* promoter mutation status provides no additional information to predict of tumor behavior in low-risk and high-risk tumors. However, they may be used to identify the cases that have a higher risk of metastasis in the intermediate-risk group.[78] At any rate, there is no TERT mutation studies in primary pancreatic SFTs in the literature.

In conclusion, SFTs form demarcated round tumors in the pancreas creating the impression of nonductal tumors, especially well-differentiated neuroendocrine tumors radiologically. Microscopically, SFT should be considered in the differential diagnosis of spindle cell neoplasms identified in the pancreas. The alternating cellularity, staghorn-like vessels, the intermixing of tumor cells with entrapped pancreatic parenchyma, and characteristic immunophenotype (nuclear STAT6 expression) are helpful features. Although primary pancreatic SFTs tend to have a favorable prognosis, marked cellularity, nuclear pleomorphism, necrosis, and high mitotic activity are risk factors for aggressive behavior.

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