Journal of Case Reports and Studies
Volume 13 | Issue 1
ISSN: 2348-9820

Case Report Open Access

Solitary Fibrous Tumor of the Pancreas: Case Report

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Citation: Jacqueline Mugnier, Edgar M González, Elga J Vargas et al. (2025) Solitary Fibrous Tumor of the Pancreas: Case Report, J Case Rep Stud 13(1): 101

Received Date: December 06, 2024 **Accepted Date:** January 06, 2025 **Published Date:** January 10, 2025

Abstract

Solitary fibrous tumors (SFTs) are rare mesenchymal neoplasms, with pancreatic involvement being exceptionally uncommon. This case corresponds to a 67-year-old woman who consulted for pancreatic mass causing biliary obstruction and imaging studies suggested pancreatic adenocarcinoma or a neuroendocrine tumor. Histopathological analysis of a biopsy identified spindle-cell proliferation without atypia or necrosis and a characteristic immunohistochemical profile positive for CD34, CD99, BCL-2, and STAT6. These findings confirmed the diagnosis of a solitary fibrous tumor of pancreas. This case highlights the diagnostic reliance on immunohistochemistry, particularly STAT6 and NAB2-STAT6 gene fusion, to differentiate SFTs from other mesenchymal neoplasms. This is the first reported case of a pancreatic SFT in Colombia, adding to the limited global data on this rare entity.

Keywords: Solitary Fibrous Tumors; Pancreas; Mesenchymal Tumors

List of Abbreviations: ALT: alanine aminotransferase; AST: aspartate aminotransferase; Ca: calcium; CDk4: cyclin-dependent kinase 4; CKs: cytokeratin's; CT: computerized tomography; DM: diabetes mellitus; EMA: epithelial membrane antigen; ERCP: Cholangiopancreatography; GIST: gastrointestinal stromal tumor; H&E: hematoxylin & eosin; HPF: high power fields; IHC: immunohistochemical; INR: international normalized ratio; K: potassium; MRI: magnetic resonance imaging; MSA: muscle-specific actin; Na: sodium; PT: prothrombin time; PTT: partial thromboplastin time; SAT: systemic arterial hypertension; SFTs: Solitary fibrous tumors; SMA: smooth muscle actin

Introduction

Mesenchymal neoplasms of the pancreas are exceptionally rare, accounting for only 1% of all tumors of the organ [1], with leiomyosarcomas being the most frequent [2]. Solitary fibrous tumors (SFTs) of the pancreas are categorized as myofibroblastic mesenchymal neoplasms with uncertain to malignant behavior, according to the World Health Organization's classification of tumors [3]. The global incidence is approximately 2.8/100,000 (4), with only 0.6% of these being malignant in the literature [5, 6]. SFTs are most commonly found in the pleura (65% incidence) [1], but they can arise in any anatomical location, including the esophagus, gallbladder, liver, lung, thyroid, kidney, adrenal gland, seminal vesicles, paranasal sinuses, breast, salivary glands, meninges, and the mesentery of the small intestine. They can also appear on the serosal surfaces of the stomach and colon [1, 3, 5], making their occurrence in the pancreas exceedingly rare [7]. The condition is commonly diagnosed in adults aged 50 to 54 [4, 8], with no difference in presentation between sexes [3,4].

SFTs are generally asymptomatic and slow-growing, but they can manifest in various ways, including jaundice, pain, weight loss, or lumbar pain [9, 10]. Additionally, SFTs are known to be associated with hypoglycemia due to the release of insulin-like growth factor II [8]. Macroscopically, SFTs are described as oval to round, grayish masses that may or may not be encapsulated [11]. Histologically, they consist of spindle cells with hypercellular areas grouped in fascicles or scattered without a characteristic pattern (the so-called "pattern less pattern"), embedded in a hyalinized collagen stroma rich in vessels with a deer-antler or hemangiopericytic pattern [1]. Immunohistochemical (IHC) studies show positivity of tumor cells for CD34, vimentin, CD99, and the oncoprotein Bcl-2 (6), while they are negative for cytokeratin's (CKs), smooth muscle actin (SMA), desmin, epithelial membrane antigen (EMA), CD117, S-100, HMB-45, and C-kit [7, 12]. Currently, the nuclear biomarker NAB2-STAT6 is highly sensitive for SFTs [1, 13, 14]. The proliferation index with Ki-67 has been observed to be below 1% in cases described as benign [15].

Case Report

A 67-year-old woman with a history of systemic arterial hypertension (SAT), type 2 diabetes mellitus (DM), and hypothyroidism was referred to FCI due to the finding of an abdominal mass measuring 78 x 79 mm with necrotic areas and a mass effect on the bile duct, causing secondary obstruction of the biliary tract, diagnosed by contrast-enhanced Abdominal CT scan on 15/12/2023. Figure 1. Differential diagnoses included neuroendocrine tumor versus pancreatic adenocarcinoma.

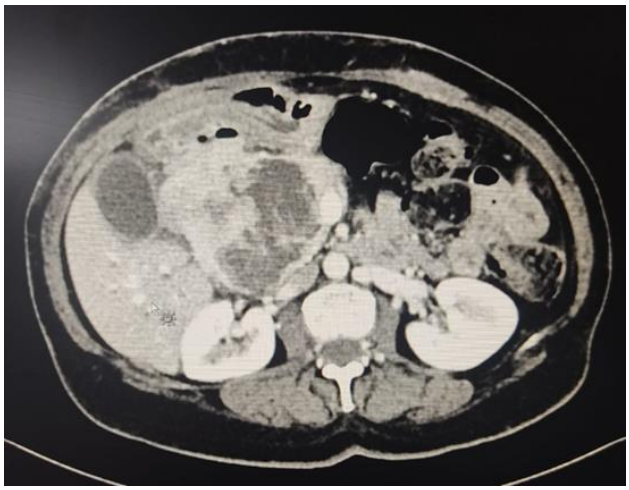

Figure 1: Contrast abdominal tomography. Lesion of neoplastic aspect depending on the head of the pancreas (78 x 79 mm) with necrotic areas inside. The tumor caused secondary dilatation of the intra and extrahepatic biliary tract by extrinsic compression of the middle third of the common bile duct.

The following laboratory tests were ordered: alkaline phosphatase 719 mg/dl, AST 28 U/L, ALT 64 U/L, amylase 29 U/L, total bilirubin 1.2 mg/dl (direct 0.7 mg/dl and indirect 0.5 mg/dl); INR 0.95, PT 13.5 seconds, PTT 27.2 seconds; Na 138 meq/L, K 4.0 meq/L, Mg 1.89 meq/L, Ca 8.8 meq/L, CA19-9 687.58U/ml, carcinoembryonic antigen 2.16 ng/ml, and complementary imaging studies: chest CT and bone scan showed no evidence of lesions or vascular invasion. On 17/12/2023, an abdominal and pelvic magnetic resonance imaging (MRI) documented a focal solid lesion involving the uncinate process, head, and neck of the pancreas with well-defined margins, necrotic areas, and measuring 92 x 97 x 93 mm, with adenocarcinoma being the first diagnostic option and pancreatic neuroendocrine tumor the second. Figure 2.

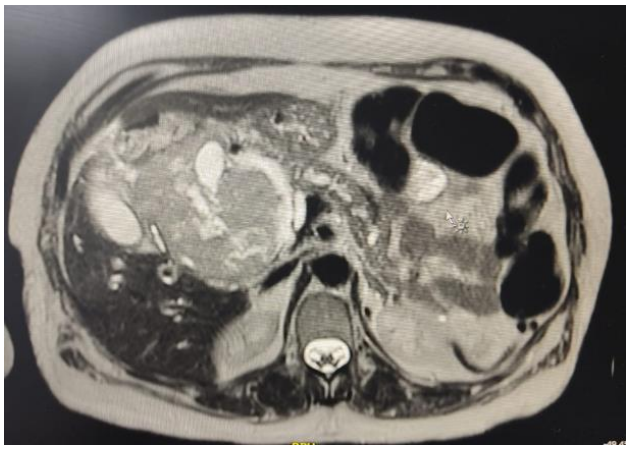

Figure 2: MRI of the abdomen. There is a solid focal lesion involving the uncinate process, head and neck of the pancreas with areas of necrosis measuring 92 x 97 x 93 mm. Invasion of the superior mesenteric vein, confluent and portal hilar vein is observed as well as contact of the tumor with the superior mesenteric artery.

A percutaneous biopsy under ultrasound guidance was performed on 15/12/2023, and the pathology report described a spindle-cell neoplastic lesion consisting of cells with oval nuclei without atypia and scant cytoplasm associated with collagen bands. The mitotic count was less than 1 mitosis in 10 HPF. No areas of necrosis or hemorrhage were observed. Figure 3. IHC studies found positivity for CD34, BCL-2, STAT-6, and negativity for AE1/AE3, DOG-1, smooth muscle actin, CD117, and S100 markers. Figure 4.

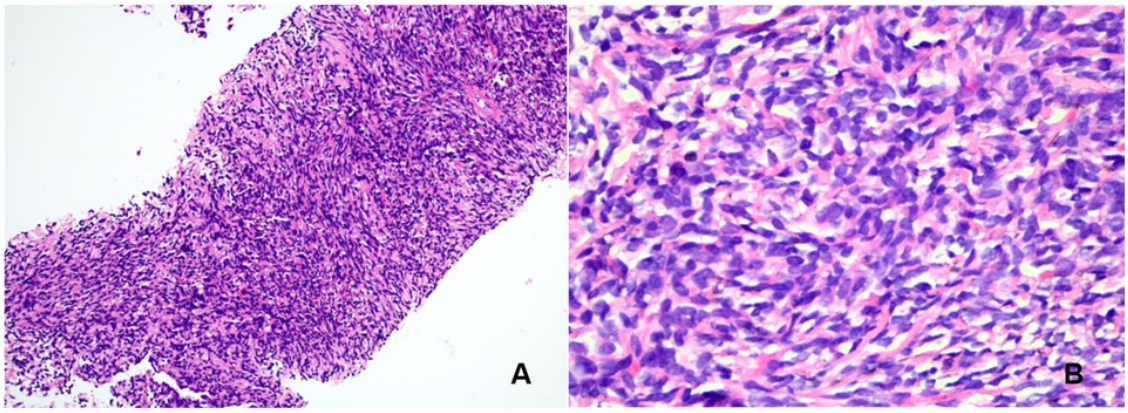

Figure 3: A. Spindle cells embedded in collagenized stroma. H&E, 10x. B Oval nuclei with homogeneous chromatin. H&E, 40x.

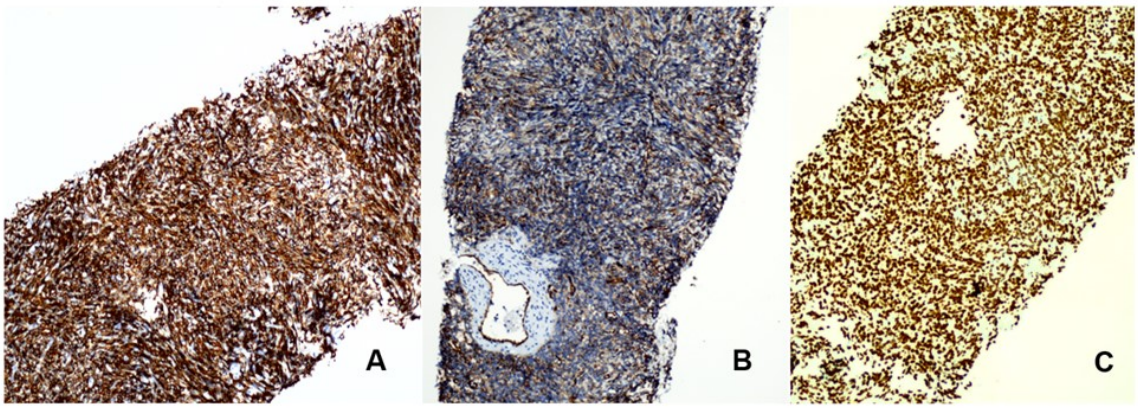

Figure 4: Tumor cells are positive for A. BCL2, B. CD34 and C. NAB2-STAT6. 10x.

Due to the SFT's close relationship with the superior mesenteric vein, confluent portal vein, and superior mesenteric artery, the patient was considered not a candidate for surgical resection due to the high risk of complications. The patient underwent Endoscopic Retrograde Cholangiopancreatography (ERCP) + endoscopic sphincterotomy and insertion of a biliary duct stent for palliative purposes on 19/12/2023. She continues to be hospitalized at home and is followed up by palliative care and oncology.

Discussion

These tumors were first described in the pleura by Klemperer and Rabin in 1931 (5), and in 1999, Lüttges et al. reported the first case in the pancreas [12, 16]. D'Amico FE et al. mention that 29 cases of pancreatic SFT have been documented in the scientific literature [16]. These cases describe tumors ranging from 5 to 18 cm with an average of 5.3 cm [10, 16], with the predominant location being the head (58.6%), followed by the body (31%) and the tail (10.4%) [13, 14, 17].

The differential diagnoses of SFT include other mesenchymal neoplasms such as leiomyosarcoma, fibrosarcoma, dedifferentiated liposarcoma, gastrointestinal stromal tumors (GIST), fibrous histiocytoma, hemangioendothelioma, hemangiopericytoma, hemangioma, lymphangioma, and even non-neoplastic proliferations [6,7,13], with IHC studies being indispensable as they are considered the diagnostic gold standard [13].

Leiomyosarcomas are positive for SMA, muscle-specific actin (MSA), and Desmin, markers that are negative in SFT (18). Both SFT and fibrosarcoma are positive for vimentin and CD34, but fibrosarcoma is reactive for p53 [19]. Dedifferentiated liposarcomas are immunoreactive for cyclin-dependent kinase 4 (CDk4), p16, p53, and SMA (20).

Both SFT and GIST show immunoreactivity for CD34 (82%), but GISTs are positive for CD117 in 95% and SMA in 18% of cases, with these two markers being negative in SFT (21). Fibrous histiocytoma is immunoreactive for SMA (38%) and CD34 in 40% of cases (22).

Vascular tumors such as hemangioendothelioma, hemangiopericytoma, and hemangioma share positivity for CD31, transcription protein ERG, FLI1, and CD34, with the latter being the only marker they share with SFT (23). Lymphangioma differs from SFT by its positive IHC for PROX1, VEGFR, CD31, LYVE1, and SMA (24). Finally, the nuclear biomarker STAT6 is positive in SFTs and negative in reactive processes and most mesenchymal neoplasms (2).

A specific genetic alteration of the TFS, the gene fusion NAB2-STAT6 and mainly its chimeric variant NAB2ex6-STAT6ex16/17 (more frequent in its extra thoracic expressions), is considered a driver mutation with specific clinical and patho-

logical feature (24-26). The detection of this fusion gene ranges from 55-100% of cases of TFS, being negative in most mesenchymal neoplasms (2,24,25,27). For this reason, the nuclear immunoexpression by IHC of STAT6 is a useful marker to demonstrate the presence of the NAB2-STAT6 fusion (27).

Ki-67 expression greater than 1%, histological findings such as atypical mitoses, high cellularity, increased mitotic activity (more than 4 mitoses per 10 HPF), tumor necrosis, infiltrative margins, and tumor size greater than 10 cm (about 3.94 in) are associated with malignancy (6,13).

Table 1: Reported cases of solitary fibrous tumor of the pancreas (1999-2024)

Authors	Findings of Immunohistochemistry	Histologic of malignancy	Diagnosis of malignancy	Recurrence	Evolution
Lüttges et al	CD34, CD99, Bcl2, vimentina	No	No	No	Life
Chatti et al	CD34, CD99, Bcl2, vimentina	No	No	No	Intraoperative death
Gardini et al	CD34, CD99, Bcl2, vimentina, SMA	NA	No	No	Life
Miyamoto et al	CD34, Bcl2	No	No	No	Life
Srinivasan et al	CD34, Bcl2	No	No	No	Life
Kwon et al	CD34, CD99, vimentina	No	No	No	NA
Ishiwatari et al	CD34, Bcl2	Necrosis	No	No	Life
Chetty et al	CD34, CD99, Bcl2	No	No	No	Life
Sugawara et al	CD34	No	No	No	NA
Santos et al	CD34, B/catenin	No	No	No	NA
Tasdemir et al	CD34, Bcl2, B/catenin, vimentina, Ki 67 2%	No	No	No	Life
Azadi et al	CD34, Bcl2, Ki 67 5%	No	No	No	NA
Van der Vorst et al	CD34, CD99, Bcl2	No	No	No	NA
Yamanashi et al	CD34, vimentina, Bcl2	No	Si	Si	Life
Chen et al	CD34, Bcl2, vimentina, CD68, SMA	No	No	No	Life
Hwang et al	CD34, Bcl2, actina, CD10, ER, PR	No	No	No	Life
Han et al	CD34, CD99	No	No	-	No progression
Estrella et al	CD34, Bcl2, CK, p16, p53	Nuclear atypia, necrosis, 17 mitoses/10 HPFs	Si	No	Life
Baxter et al	CD34, Bcl2	NA	No	No	Life
Paramythiotis et al	CD34, CD99, Bcl2, vimentina, S-100	No	No	No	Life

Murakami et al	STAT6, CD34, Bcl2, ACTH, POMC, NSE	No	No	No	Death due to sepsis
Spasevska et al	CD34, vimentina, CD99, Bcl2, B/catenin	No	No	No	Death due to postoperative complication
Clare et al	STAT6, CD34, Bcl2, CD56, CAM5.2, AE1/AE3	6/10 HPFs	Si	No	Life
Sheng et al	CD34, vimentina, SMA, Ki-67 3%	Mild-moderate nuclear pleomorphism, 2-5/10 HPFs, hypercellularity	No	No	Life
D'Amico et al	STAT6, CD34	No	No	No	Life
Oana et al	CD34, Bcl2	No	No	No	Life
Geng et al	STAT6, CD34, Bcl2, CD31, PHH-3, D2-40, Ki-67 10%	4-5/10 HPFs, necrosis	Si	Residual hepático	Life
Qian et al	STAT6, CD34, Bcl2, Ki-67 10%	4-5/10 HPFs, local infarction	Si	Multiples	Life
Taguchi et al	STAT6, CD34, Bcl2, vimentina, AE1/AE3	Hypercellularity 12/10 HPFs, increasing necrosis.	Si	No	Life
This case	CD34, Bcl2, STAT6, Ki-67 40%	1/10 HPFs	No	No	-

Modified table of cases from Taguchi et al article (14), with our specific case and markers. This would be the first SFT's case reported in Colombia

Conclusion

SFTs are rarely documented outside the pleura, and their location in the pancreas is an exceptional finding. To date, 29 cases of pancreatic SFT have been reported in the literature, and this would be the first case reported in Colombia. Table 1. Histologically, they resemble other benign and malignant mesenchymal tumors as well as non-tumoral fibroblastic proliferations, making clinical correlation and IHC studies essential for a definitive diagnosis, with the STAT6 marker being particularly noteworthy. Most SFTs exhibit benign behavior, and surgical resection is the treatment of choice. Our patient did not undergo surgery due to the close relationship of the tumor with vascular structures and the high probability of complications.

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