

## Solitary fibrous tumour of the pancreas: a case report

### *Tumor fibroso solitario de páncreas: a propósito de un caso*

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#### ABSTRACT

The solitary fibrous tumour is an uncommon neoplasm of mesenchymal origin. It occasionally develops in extra pleural locations, and is very infrequent in the pancreas. A 40-year-old woman had an unsuspected solitary fibrous tumour detected by chance in the body of the pancreas. Detection of the mass occurred with imaging studies. CT and MRI images showed the well-circumscribed benign lesion, with 3 cm on its greatest diameter. Treatment consisted of partial pancreatectomy, and her postoperative recovery was uneventful.

**Key words.** Diagnosis. Pancreas. Pancreatic neoplasma. Solitary fibrous tumor.

#### RESUMEN

El tumor fibroso solitario es una neoplasia poco común, de origen mesenquimal, que ocasionalmente se desarrolla en sitios extrapleurales, y es muy raro en el páncreas. Se presenta el caso de una mujer de 40 años, que presentó un tumor fibroso solitario descubierto por casualidad en el cuerpo del páncreas. La masa se observó con estudios de imagen. La lesión benigna, bien delimitada, con 3 centímetros de diámetro mayor, se descubrió en imágenes de CT y MRI. El tratamiento consistió en pancreatectomía parcial, y su recuperación postoperatoria no tuvo complicaciones.

**Palabras clave.** Diagnóstico. Páncreas. Neoplasma pancreático. Tumor fibroso solitario.

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## INTRODUCTION

Solitary fibrous tumor (SFT) is a very rare neoplasm of mesenchymal origin, characterized by the proliferation of fibroblast-like spindle cells and a variable component of hyalinized collagen<sup>1-9</sup>. Hypo and hyper cellular areas develop with anarchical arrangement of tumor cells in fascicles; while the fibrocollagen stroma shows variable hyalinization and may present numerous vessels<sup>1,3</sup>. Extra abdominal SFT is reported in the pleura (65%), lung, mediastinum, pericardium, meninges, orbit, oral cavity, salivary glands, nose, paranasal sinus, thyroid, diaphragm, breast, spinal cord, musculoskeletal system, vulva and vagina<sup>5,7</sup>. Intra abdominal STF has been described in the peritoneum, mesentery, adrenals, urogenital tract, stomach, and liver, but pancreatic location is exceeding rare<sup>1-9</sup>. All tumors had benign histological features. There were four tumor resections, three pancreatoduodenectomies, and two partial pancreatectomies<sup>1-9</sup>. Diagnosis cannot be established exclusively with base on clinical and imaging data, and suggestive histopathology features must be confirmed by immunoistochemical studies. SFTs are classically positive for CD34, bcl-2, vimentin and CD99, and negative for cytokeratins, smooth muscle actin, desmin, epithelial membrane antigen, S-100, HMB-45, as well as c-kit<sup>1,3,5-9</sup>. Case studies can contribute to increase the awareness about this scarcely reported condition.

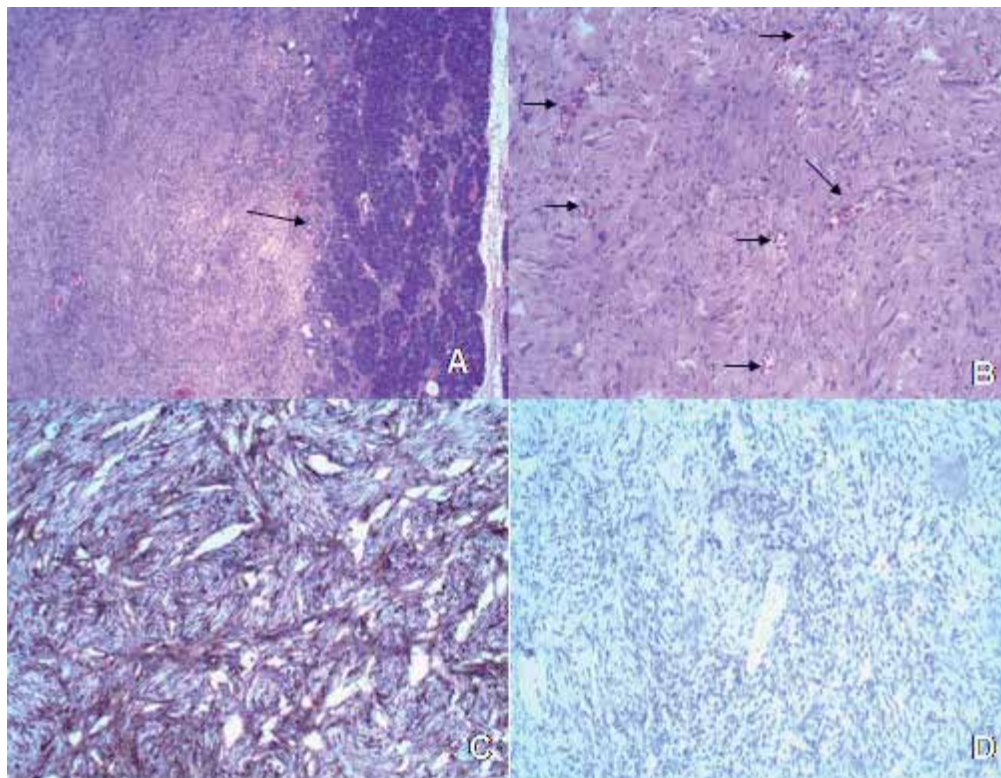
## CASE REPORT

A 40-year-old woman came to Hospital because of a gynecological affection. Her physical examination and routine laboratory data were within normal limits, including the tumor markers. She was submitted to CT and MRI pelvic and abdominal imaging studies, which incidentally revealed a rounded solid mass in the body of the pancreas. This unsuspected mass appeared well circumscribed, with 3 cm in its greatest diameter. Worth of note, no other remarkable change was detected in images of the abdominal cavity, and regional lymphadenopathy was not observed. The samples obtained from fine needle aspiration were not sufficient to establish the diagnosis. The patient was then submitted to a partial

pancreatectomy, with uneventful postoperative course. Grossly, the tumor was solid with some cystic areas, pseudocapsulated and well delimited, surrounded by normal pancreas tissue. The histopathology analysis revealed that the boundaries between the SFT and normal pancreas tissue was clearly demarcated (Fig. 1A), and the focally hyalinized fusocellular growth showed a highly vascularized stroma (Fig. 1B). Neither cytonuclear atypias nor mitoses, or necrotic areas were observed, and the mass was considered of benign nature. On immunohistochemistry studies, the majority of tumor spindle cells appeared strongly positive for CD34 (Fig. 1C), and moderately for beta-catenin. Moreover, they were negative for pancytokeratins AE1 and AE3, EMA, CD117, desmin, smooth muscle actin, and S-100 protein, with no immunoexpression of c-kit (Fig. 1D).

## DISCUSSION

The 40-old-woman here reported had an asymptomatic benign SFT, with 3 cm of diameter, in the pancreatic body. The tumor cells were positive for CD34 and beta-catenin, and negative for c-kit. The diagnosis of SFT relied upon histological features plus immunodetection of CD34<sup>1-9</sup>, and absence of immunoexpression of c-kit. These data ruled out the hypothesis of gastrointestinal stromal tumor<sup>6-8</sup>. Rakheja et al also described positive staining for beta-catenin in twelve cases of non-pancreatic SFT (one was malignant), and suggested that the relationship between immunoistochemical features and clinical behavior of these tumors should be cleared<sup>10</sup>. Similar to the present case, they found a pure membranous staining pattern of beta-catenin in two (17%) of the benign SFTs, which were located in the eyelid and omentum<sup>10</sup>. Although the staining patterns of beta-catenin are variable in SFTs, both cytoplasmic and membranous locations are normal, while the nuclear accumulation is abnormal<sup>10</sup>. Differential diagnosis of SFT includes non-epithelial tumors: leiomyosarcoma, liposarcoma, fibrosarcoma, fibrous histiocytoma, heman-gioendothelioma, hemangiopericytoma, hemangioma, lymphangioma, islet cell tumor, paraganglioma, neuroectodermal tumor, and schwannoma<sup>3,5,6,8,9</sup>.



**Figure 1A.** Clear limits (arrow) between the fibrous tumor and the normal pancreas (HE, 40x).

**Figure 1B.** Hyalinized fusocellular tumor showing a vascularized (arrows) stroma (HE, 100x).

**Figure 1C.** Immunoexpression of CD34.

**Figure 1D.** Lack of immunoexpression of c-kit (CD117).

In this patient, the finding of typical pathological features of SFT of the pancreas ruled out those hypotheses. The solid tumor was well demarcated from the normal pancreas, showing a patternless architecture composed by bland spindle cells arranged in fascicles alternating with dense hyalinized areas. Dilated vascular channels mimicking hemangiopericytoma were focally distributed<sup>1-9</sup>. Cellular atypias, mitosis and necrosis were absent, and normal acini were surrounding the tumor.

Statistical data about SFT of the pancreas showed few changes after including findings from the present case study (Table 1).

Nine out of ten patients were women (90%), and 60% were asymptomatic. The mean age of the patients was 54.6 ( $\pm$  12.3) years, and the median age was 54.5 (40 to 78) years. More often (60% of cases), the mass was in the body of the pancreas, with the greatest diameter ranging between 2 and 13 cm [mean of 4.9 ( $\pm$ 3.4) cm, and a median of 3 cm]. Although all these SFTs of the pancreas had benign characteristics, larger series of cases with longstanding follow-up are necessary to better clear their eventual malignant potential<sup>9</sup>. Additionally, the clinical significance of b-catenin staining in SFTs is not totally understood<sup>10</sup>.

**Table 1.** Comparative data of ten patients with solitary fibrous tumors of the pancreas

Authors (year)	Gender/age	Symptoms	Site	Diam	Immunostaining (+)
Lüttges <sup>6</sup> (1999)	F/ 50 years	Absent	Body	5.5 cm	CD34, CD99, bcl-2, vim
Chatti <sup>1</sup> (2006)	M/ 41 years	Abd pain	Body	13 cm	CD34, CD99, bcl-2, vim
Gardini <sup>3</sup> (2007)	F/ 62 years	Abd pain	Head	2.5 cm	CD34, CD99, bcl-2, vim
Miyamoto <sup>7</sup> (2007)	F/ 41 years	Abd pain	Body	2 cm	CD34, bcl-2
Kwon <sup>5</sup> (2008)	F/ 54 years	Absent	Body	7.6 cm	CD34, CD99, vim
Srinivasan <sup>8</sup> (2008)	F/ 78 years	Back pain	Body	4.5 cm	CD34, CD99, bcl-2, vim
Chetty <sup>2</sup> (2009)	F/ 67 years	Absent	Head	2.6 cm	CD34, CD99, bcl-2
Ishiwatari <sup>4</sup> (2009)	F/ 58 years	Absent	Head	3 cm	CD34, bcl-2
Sugawara <sup>9</sup> (2010)	F/ 55 years	Absent	Head	7 cm	CD34
Santos LAM (*)	F/ 40 years	Absent	Body	3 cm	CD34, $\beta$ -catenin

F: female; M: male; Abd: abdominal; Diam: greatest diameter; vim: vimentin.

\*This case study.

We believe that case studies contribute to better awareness about scarcely reported conditions.

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