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# Malignant solitary fibrous tumor of the pancreas with systemic metastasis: A case report and review of the literature

Geng H *et al*. Malignant SFT of the pancreas

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**Abstract**

BACKGROUND

Pancreatic solitary fibrous tumor (SFT) is a rare neoplasm of intermediate biological potential. So far, only 22 cases have been reported since 1999. All the cases, except one, exhibited benign features. Here, we report the first case of malignant pancreatic SFT with typical Doege-Potter syndrome, along with the clinical and pathologic evidence of its systemic metastasis.

CASE SUMMARY

The patient was a 48-year-old man with a 1-year history of pancreatic and liver masses and refractory hypoglycemia. Increased uptake of the tracer fluorodeoxyglucose (FDG) was found in the liver and bones by fluorine-18 FDG positron emission tomography/computed tomography. After multidisciplinary discussion, a distal pancreatectomy procedure was performed, and histological examination showed a lesion composed of abundant heterogeneous spindle cells with localized necrosis. On immunohistochemistry evaluation, STAT6 was found to be diffusely expressed in the tumor. Based on the overall evidence, the patient was diagnosed with malignant pancreatic SFT with liver and bone metastases.

CONCLUSION

The diagnosis of malignant SFT requires comprehensive evidence including clinical, immunohistochemistry, and histological features. This case may be presented as a reference for diagnoses and management of malignant pancreatic SFTs with systemic metastasis.

**Key words:** Solitary fibrous tumor; Pancreas; Malignant; Doege-Potter syndrome; Case report

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**Core tip:** Solitary fibrous tumor is now considered as a fibroblastic mesenchymal neoplasm of intermediate biological potential, and it rarely occurs in the pancreas. Here, we report a case of malignant pancreatic solitary fibrous tumor with systemic metastasis, review the literature, and discuss its biological features, diagnosis, and prognosis evaluations.

**INTRODUCTION**

Solitary fibrous tumor (SFT), first described in 1870, was established as a pleural neoplasm by Klemperer and Rabin in 1931[1]. This tumor is commonly found in serosal membranes, the dura of the meninges, and deep soft tissues. It is now recognized as a type of fibroblastic mesenchymal neoplasm of intermediate biological potential characterized by the pathognomonic *NAB2-STAT6* gene fusion[2]. Only a few reports on malignant pancreatic SFT have been previously published. We present herein the first case of malignant pancreatic SFT with typical Doege-Potter syndrome and, hepatic and bone metastases.

**CASE PRESENTATION**

***Primary complaints***

A 48-year-old man was admitted to our hospital with 1-year history of pancreatic and liver tumors. The tumors were accidentally found when the patient went to a local hospital after a sudden incidence of fainting. It is noteworthy that he reported of recurrent incidences of hypoglycemia, however, there was no history of any endocrine disease.

***History of past illness***

His medical history showed that he had been treated eight times for metastatic liver tumor by transcatheter arterial chemoembolization and once by radioactive seed implantation. Five years before presentation, he had undergone an excision of a tumor of the right pterygopalatine fossa.

***Physical examination***

Physical examination showed no other positive findings except that the liver was enlarged and palpable.

***Laboratory and imaging examinations***

Laboratory investigations showed an abnormal hemogram, including hemoglobin of 123 g/L (reference range: 131-172 g/L), neutrophils 72.8% (reference range: 50%-70%) and lymphocytes 10.8% (reference range: 20%-40%). The results of liver and kidney function were normal. The levels of serum tumor markers (CEA, CA 19-9, CA 12-5, and AFP) were all within normal limits.

Computed tomography (CT) imaging of the abdomen showed a 4.7 cm well-defined mass located in the lower posterior part of the body of the pancreas (Figure 1A). Non-uniform enhancement was observed from the arterial to portal venous phase. Meanwhile, multiple nodules and masses of various sizes were seen in the liver (Figure 1B). The largest one was located in the segment Ⅷ of the liver with a diameter of about 15.9 cm. No obvious dilatation of intrahepatic and extrahepatic bile ducts was observed.

Pancreatic magnetic resonance (MR) imagining also confirmed a hypervascular tumor located in the body of the pancreas and multiple tumors located in the liver. Those tumors were hypointense on T1-weighted MR images and hyperintense on T2-weighted MR images.

For a complete preoperative evaluation, fluorine-18 fluorodeoxyglucose positron emission tomography/CT (18F-FDG PET/CT) was performed. Images from the PET/CT revealed that both the pancreatic and the metastatic liver lesions had an increased uptake of the tracer FDG. Besides, the thoracic and lumbar vertebrae, humerus, femur, scapulae, ribs, sacrum, and pelvis also showed heterogeneous FDG uptake (Figure 2).

***Further diagnostic work-up***

A liver biopsy guided by B-mode ultrasound confirmed that the tumor was an SFT/ hemangiopericytoma (Grade 2).

**FINAL DIAGNOSIS**

The patient was eventually diagnosed with a malignant SFT of the pancreas with Doege-Potter syndrome and metastases to the liver and bone.

**TREATMENT**

In order to improve the quality of life of the patient and control the growth of the mass, a distal pancreatectomy, involving the body and tail and splenectomy, was performed after a multidisciplinary discussion, and the metastatic neoplasm in the left lateral lobe of the liver was also resected.

**OUTCOME AND FOLLOW-UP**

On gross examination, the pancreatic specimen measured 15 cm × 6 cm × 2 cm, which contained two well-circumscribed non-encapsulated masses. The larger lesion measuring 6.5 cm × 5 cm, had a soft fleshy cut surface containing hemorrhagic and necrotic areas (Figure 3A). Another metastatic lesion located in the left lobe of the liver measured 14 cm × 12 cm ×4 cm with a pale-yellow cut surface (Figure 3B). All the resection margins were free of tumor. On histopathological examination, it was found that the tumor was composed of abundant heterogeneous spindle cells (Figure 4A). A localized area of necrosis (Figure 4B) was visualized and there were 4-5 mitotic figures (Figure 4C) per 10 high-power fields (HPFs). Immunohistochemical (IHC) analysis of the resected tumor revealed that the tumor cells were diffusely positive for STAT6 (Figure 4D), CD34, CD31, Bcl-2, cell proliferation marker Ki-67, PHH-3, and D2-40, and negative for glial fibrillary acidic protein, S100, smooth muscle actin (SMA), Desmin, delay of germination 1, CD117, and receptor tyrosine kinase. The proliferation index of Ki-67 was observed to be above 10%. The patient’s postoperative recovery was uneventful. Furthermore, a transcatheter arterial chemoembolization procedure was also performed to eliminate the residual tumor of the right liver, and postoperative follow-up at 6 mo demonstrated good results (Figure 5).

**DISCUSSION**

SFTs are now considered to occur anywhere in the body, but the pancreatic fibrous tumor is still rarely recorded in the literature: only 22 cases have been reported since 1999 (Table 1). The vast majority of the cases presented with benign features, and only one case was defined as being malignant, based on its histological features[3]. The case we present here, to our knowledge, is the first malignant pancreatic SFT with clinical and pathological evidence of liver and bone metastases.

Until now, there is no one comprehensive definition of malignant SFTs. According to the previous literature, abdominal pain is the most common presentation of clinical syndrome in a pancreatic SFT (10/22 cases, 45.45%), followed by an incidental abdominal mass (8/22 cases, 36.36%) and obstructive jaundice. Infrequently, patients may present with paraneoplastic syndromes. The clinical manifestations were commonly refractory and recurrent hypoglycemia, which is the main clinical characteristic of Doege-Potter syndrome. Increased secretion of a pro-hormone form of the insulin-like growth factor Ⅱ has been confirmed to be the primary mechanism of hypoglycemia according to a study[4]. Han *et al*[5] reported that SFTs with Doege-Potter syndrome were often malignant. Our case showed typical features of Doege-Potter syndrome at the onset of disease and also the malignant features. However, these symptoms were non-specific.

Radiologically, homogeneous enhancement of the lesion in the arterial phase to portal venous phase on CT as well as low T1 signal intensity and high T2 signal intensity on magnetic resonance imaging can be observed in most of the cases[2]. The non-typical feature makes it difficult to distinguish SFTs from the other soft tissue tumors[6]. Particularly, it was reported that a malignant and larger tumor may present with hemorrhage, calcifications, cystic areas and so on[2]. A non-uniform enhancement was observed in the image examinations of our case that showed similar features.

Furthermore, although a higher FDG uptake on 18F-FDG PET/CT may be a sign of a malignant SFT, the diagnostic utility is still debatable due to its imperfect sensitivity[7]. However, in our case, the 18F-FDG PET/CT was useful in the differential diagnosis of benign and malignant SFTs and evaluation of clinical significance. Thus, 18F-FDG PET/CT examination is still a recommendation for the full evaluation of suspicious malignant tumors.

Recently, the *NAB2-STAT6* fusion gene was found to express a unique molecular feature in 100% of SFT cases[8]. Thus, compared with other conventional IHC markers like CD34, STAT6 has been proved to be more sensitive (98%) and specific (85%) for SFT. Furthermore, a previous study reported that a higher risk of SFT aggressive behavior may be associated with specific *NAB2-STAT6* fusion variants[9], which could be a biomarker for identifying the distinct molecular feature of malignant SFTs.

For pathologic features, grossly, the pancreatic SFTs range from 2.0-18.5 cm in diameter[10,11]. Tumors are usually well-circumscribed with a fibrous pseudocapsule. The cut surface may show a wide range of patterns from firm, white to tan, and fleshy mass with hemorrhage, necrosis, or calcification usually presented in large or malignant cases[2]. Histologically, a typical “patternless pattern”, *i.e.*, various atypical spindled cells arrayed randomly within the stroma, can be seen in most cases (Table 2). People have defined malignant SFTs based upon its special histologic features: ≥ 4 mitotic figures per 10 HPFs, necrosis or hemorrhage, increased cellularity, nuclear pleomorphism, and a large size (> 10 cm). The histological results of our case meet these criteria and show high-grade malignant manifestations. However, a poor correlation with patients outcomes[12] has been seen as low validity in predicting the biological features of SFTs. Therefore, pathologists treat SFT as a neoplasm of intermediate biological potential. Furthermore, complete surgical resection is the mainstay of treatment for pancreatic SFTs and good results were reported (Table 2). Unfortunately, almost no information has been provided concerning systemic treatments for malignant pancreatic SFTs. For this reason, a multidisciplinary discussion, especially with the participation of pathologists, is recommended before initiation of treatment procedures for patients with an advanced stage of the disease.

**CONCLUSION**

In summary, we present a malignant pancreatic SFT with systemic metastasis and typical Doege-Potter syndrome features. The diagnosis and prognosis evaluation of malignant SFTs rely on more accurate criteria combined with clinical, IHC, and histological evidence. Furthermore, prospective studies are needed to provide greater evidence about the systemic management of malignant pancreatic SFTs.

**REFERENCES**

1 **Klemperer P**, Coleman BR. Primary neoplasms of the pleura. A report of five cases. *Am J Ind Med* 1992; **22**: 1-31 [PMID: 1415270 DOI: 10.1002/ajim.4700220103]

2 **Huang SC**, Huang HY. Solitary fibrous tumor: An evolving and unifying entity with unsettled issues. *Histol Histopathol* 2019; **34**: 313-334 [PMID: 30431144 DOI: 10.14670/HH-18-064]

3 **Estrella JS**, Wang H, Bhosale PR, Evans HL, Abraham SC. Malignant Solitary Fibrous Tumor of the Pancreas. *Pancreas* 2015; **44**: 988-994 [PMID: 26166470 DOI: 10.1097/MPA.0000000000000350]

4 **Fung EC**, Crook MA. Doege-Potter syndrome and 'big-IGF2': a rare cause of hypoglycaemia. *Ann Clin Biochem* 2011; **48**: 95-96 [PMID: 21406561 DOI: 10.1258/acb.2011.011020]

5 **Han G**, Zhang Z, Shen X, Wang K, Zhao Y, He J, Gao Y, Shan X, Xin G, Li C, Liu X. Doege-Potter syndrome: A review of the literature including a new case report. *Medicine (Baltimore)* 2017; **96**: e7417 [PMID: 28682900 DOI: 10.1097/MD.0000000000007417]

6 **Azadi J**, Subhawong A, Durand DJ. F-18 FDG PET/CT and Tc-99m sulfur colloid SPECT imaging in the diagnosis and treatment of a case of dual solitary fibrous tumors of the retroperitoneum and pancreas. *J Radiol Case Rep* 2012; **6**: 32-37 [PMID: 22690289 DOI: 10.3941/jrcr.v6i3.904]

7 **Tazeler Z**, Tan G, Aslan A, Tan S. The utility of 18F-FDG PET/CT in solitary fibrous tumors of the pleura. *Rev Esp Med Nucl Imagen Mol* 2016; **35**: 165-170 [PMID: 26632105 DOI: 10.1016/j.remn.2015.10.003]

8 **Schweizer L**, Koelsche C, Sahm F, Piro RM, Capper D, Reuss DE, Pusch S, Habel A, Meyer J, Göck T, Jones DT, Mawrin C, Schittenhelm J, Becker A, Heim S, Simon M, Herold-Mende C, Mechtersheimer G, Paulus W, König R, Wiestler OD, Pfister SM, von Deimling A. Meningeal hemangiopericytoma and solitary fibrous tumors carry the NAB2-STAT6 fusion and can be diagnosed by nuclear expression of STAT6 protein. *Acta Neuropathol* 2013; **125**: 651-658 [PMID: 23575898 DOI: 10.1007/s00401-013-1117-6]

9 **Barthelmeß S**, Geddert H, Boltze C, Moskalev EA, Bieg M, Sirbu H, Brors B, Wiemann S, Hartmann A, Agaimy A, Haller F. Solitary fibrous tumors/hemangiopericytomas with different variants of the NAB2-STAT6 gene fusion are characterized by specific histomorphology and distinct clinicopathological features. *Am J Pathol* 2014; **184**: 1209-1218 [PMID: 24513261 DOI: 10.1016/j.ajpath.2013.12.016]

10 **Miyamoto H**, Molena DA, Schoeniger LO, Haodong Xu. Solitary fibrous tumor of the pancreas: a case report. *Int J Surg Pathol* 2007; **15**: 311-314 [PMID: 17652547 DOI: 10.1177/1066896907302419]

11 **Tasdemir A**, Soyuer I, Yurci A, Karahanli I, Akyildiz H. A huge solitary fibrous tumor localized in the pancreas: a young women. *JOP* 2012; **13**: 304-307 [PMID: 22572138 DOI: 10.6092/1590-8577/613]

12 **Gold JS**, Antonescu CR, Hajdu C, Ferrone CR, Hussain M, Lewis JJ, Brennan MF, Coit DG. Clinicopathologic correlates of solitary fibrous tumors. *Cancer* 2002; **94**: 1057-1068 [PMID: 11920476 DOI: 10.1002/cncr.10328]

13 **Lüttges J**, Mentzel T, Hübner G, Klöppel G. Solitary fibrous tumour of the pancreas: a new member of the small group of mesenchymal pancreatic tumours. *Virchows Arch* 1999; **435**: 37-42 [PMID: 10431844 DOI: 10.1007/s004280050392]

14 **Chatti K**, Nouira K, Ben Reguigua M, Bedioui H, Oueslati S, Laabidi B, Alaya M, Ben Abdallah N. [Solitary fibrous tumor of the pancreas. A case report]. *Gastroenterol Clin Biol* 2006; **30**: 317-319 [PMID: 16565671 DOI: 10.1016/s0399-8320(06)73174-8]

15 **Gardini A**, Dubini A, Saragoni L, Padovani F, Garcea D. [Benign solitary fibrous tumor of the pancreas: a rare location of extra-pleural fibrous tumor. Single case report and review of the literature]. *Pathologica* 2007; **99**: 15-18 [PMID: 17566307]

16 **Kwon HJ**, Byun JH, Kang J, Park SH, Lee MG. Solitary fibrous tumor of the pancreas: imaging findings. *Korean J Radiol* 2008; **9 Suppl**: S48-S51 [PMID: 18607126 DOI: 10.3348/kjr.2008.9.s.s48]

17 **Srinivasan VD**, Wayne JD, Rao MS, Zynger DL. Solitary fibrous tumor of the pancreas: case report with cytologic and surgical pathology correlation and review of the literature. *JOP* 2008; **9**: 526-530 [PMID: 18648147]

18 **Chetty R**, Jain R, Serra S. Solitary fibrous tumor of the pancreas. *Ann Diagn Pathol* 2009; **13**: 339-343 [PMID: 19751911 DOI: 10.1016/j.anndiagpath.2009.02.006]

19 **Ishiwatari H**, Hayashi T, Yoshida M, Kuroiwa G, Sato Y, Kobune M, Takimoto R, Kimura Y, Hasegawa T, Hirata K, Kato J. [A case of solitary fibrous tumor of the pancreas]. *Nihon Shokakibyo Gakkai Zasshi* 2009; **106**: 1078-1085 [PMID: 19578317 DOI: 10.11405/nisshoshi.106.1078]

20 **Sugawara Y**, Sakai S, Aono S, Takahashi T, Inoue T, Ohta K, Tanada M, Teramoto N. Solitary fibrous tumor of the pancreas. *Jpn J Radiol* 2010; **28**: 479-482 [PMID: 20661701 DOI: 10.1007/s11604-010-0453-x]

21 **Santos LA**, Santos VM, Oliveira OC, De Marco M. Solitary fibrous tumour of the pancreas: a case report. *An Sist Sanit Navar* 2012; **35**: 133-136 [PMID: 22552135 DOI: 10.4321/s1137-66272012000100013]

22 **van der Vorst JR**, Vahrmeijer AL, Hutteman M, Bosse T, Smit VT, van de Velde CJ, Frangioni JV, Bonsing BA. Near-infrared fluorescence imaging of a solitary fibrous tumor of the pancreas using methylene blue. *World J Gastrointest Surg* 2012; **4**: 180-184 [PMID: 22905287 DOI: 10.4240/wjgs.v4.i7.180]

23 **Chen JW**, Lü T, Liu HB, Tong SX, Ai ZL, Suo T, Ji Y. A solitary fibrous tumor in the pancreas. *Chin Med J (Engl)* 2013; **126**: 1388-1389 [PMID: 23557579]

24 **Hwang JD,** Kim JW, Chang JC. Imaging Findings of a Solitary Fibrous Tumor in Pancreas: A Case Report. *J Korean Soc Radiol* 2014; 70 [DOI: 10.3348/jksr.2014.70.1.53]

25 **Baxter AR**, Newman E, Hajdu CH. Solitary fibrous tumor of the pancreas. *J Surg Case Rep* 2015; **2015** [PMID: 26628714 DOI: 10.1093/jscr/rjv144]

26 **Han SH,** Baek YH, Han S-y, Lee SW, Jeong JS, Cho JH, Kwon H-J. Solitary Fibrous Tumor of the Pancreas: A Case Report and Review of the Literature. *Korean J Intern Med* 2015; 88 [DOI: 10.3904/kjm.2015.88.3.293]

27 **Murakami K**, Nakamura Y, Felizola SJ, Morimoto R, Satoh F, Takanami K, Katakami H, Hirota S, Takeda Y, Meguro-Horike M, Horike S, Unno M, Sasano H. Pancreatic solitary fibrous tumor causing ectopic adrenocorticotropic hormone syndrome. *Mol Cell Endocrinol* 2016; **436**: 268-273 [PMID: 27585487 DOI: 10.1016/j.mce.2016.08.044]

28 **Paramythiotis D**, Kofina K, Bangeas P, Tsiompanou F, Karayannopoulou G, Basdanis G. Solitary fibrous tumor of the pancreas: Case report and review of the literature. *World J Gastrointest Surg* 2016; **8**: 461-466 [PMID: 27358679 DOI: 10.4240/wjgs.v8.i6.461]

29 **Spasevska L**, Janevska V, Janevski V, Noveska B, Zhivadinovik J. Solitary Fibrous Tumor of the Pancreas: A Case Report and Review of the Literature. *Pril (Makedon Akad Nauk Umet Odd Med Nauki)* 2016; **37**: 115-120 [PMID: 27883325 DOI: 10.1515/prilozi-2016-0024]

30 **Oana S**, Matsuda N, Sibata S, Ishida K, Sugai T, Matsumoto T. A case of a "wandering" mobile solitary fibrous tumor occurring in the pancreas. *Clin J Gastroenterol* 2017; **10**: 535-540 [PMID: 28956313 DOI: 10.1007/s12328-017-0774-8]

**Footnotes**

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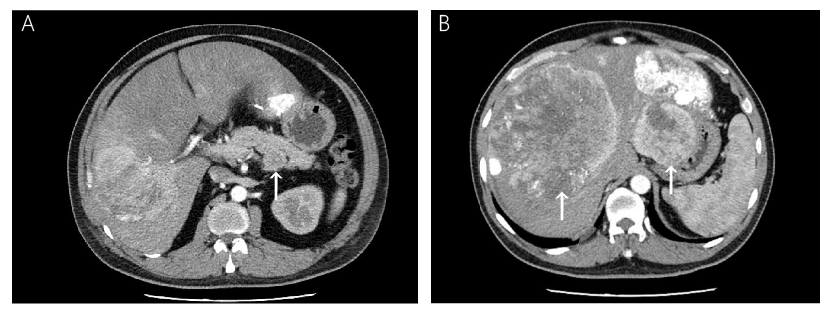
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**Figure Legends**



**Figure 1 Computed tomography imaging of the abdomen.** A: A 4.7 cm × 4.4 cm mass (white arrow) located in the body of pancreas. Non-uniform enhancement was observed from the arterial phase [computed tomography (CT) value = 68 Hu] to portal venous phase (CT value = 59 Hu); B: Numerous liver metastatic tumors (white arrows). Enhanced scanning showed irregular enhancement and the largest one located in segment Ⅷ measured 15.9 cm.

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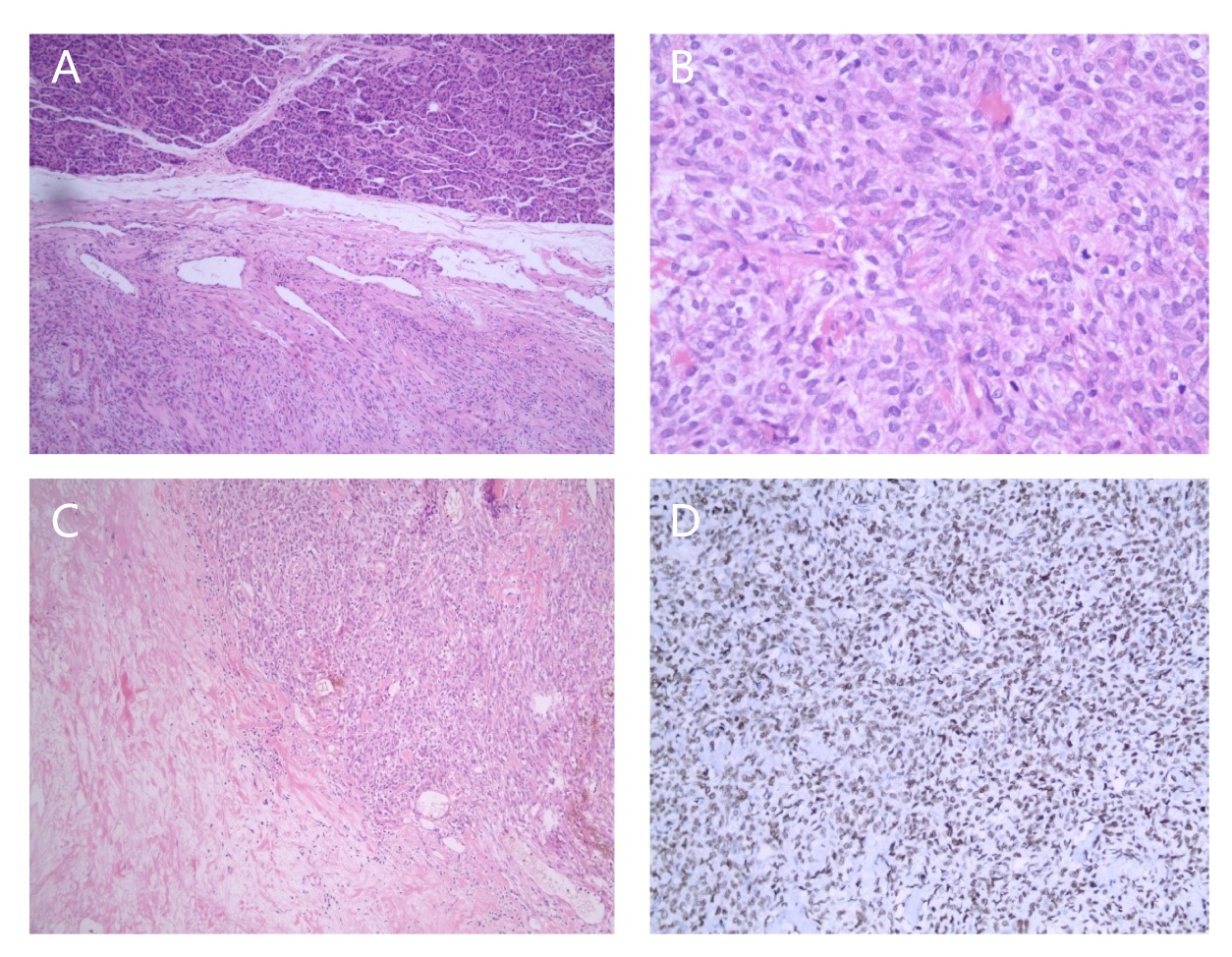
**Figure 2 Systemic fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography scan.** Thepositron emission tomography/computed tomography scan revealed an increased uptake of fluorodeoxyglucose in the liver and heterogeneous fluorodeoxyglucose uptake in multiple bones.

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**Figure 3 Photographs of cut surface of surgical specimens.** A: A soft mass located in the body of pancreas. Hemorrhage and necrosis changes can be seen in the cut surface; B: The pale-yellow cut surface of the hepatic metastasis.

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**Figure 4 Photomicrographs of histologic and immunohistochemical staining.** A: Various atypical spindled cells irregularly arranged in the stroma [hematoxylin-eosin (HE) staining; magnification: ×50]; B: Histologic demonstration of mitotic activity (HE staining; magnification: ×200); C: Presence of necrosis (HE staining; magnification: ×100); D: Immunohistochemical staining for STAT6 showed diffused positivity in tumor cells (magnification: ×100).

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**Figure 5 Timeline.** A brief summary of the patient’s medical history is presented.TACE: Transcatheter arterial chemoembolization.

**Table 1 Clinical features of pancreatic solitary fibrous tumors**

|  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Ref.** | **Age/Sex** | **Chief complaint(s)** | **Size (cm)** | **Location** | **Arterial-CT** | **Venous-CT** | **T1-MRI** | **T2-MRI** | **Tumor marker** |
| Lüttges *et al*[13] | 50/female | Incidental | 5.5 | Body | Enhanced | Enhanced | NA | NA | Negative |
| Chatti *et al*[14] | 41/male | Abdominal pain | 13.0 | Body | Enhanced | Enhanced | Hypointense | Hyperintense | Negative |
| Gardini *et al*[15] | 62/female | Abdominal pain | 3.0 | Head | Enhanced | Enhanced | NA | NA | Negative |
| Miyamoto *et al*[10] | 41/female | Abdominal pain | 2.0 | Head-body | Enhanced | Enhanced | NA | NA | Negative |
| Kwon *et al*[16] | 54/male | Incidental | 4.5 | Body | Enhanced | Enhanced | Hypointense | Hyperintense | Negative |
| Srinivasan *et al*[17] | 78/female | Back pain, weight loss | 5.0 | Body | Enhanced | Enhanced | NA | NA | Negative |
| Chetty *et al*[18] | 67/female | Incidental | 2.6 | Head | Enhanced | Enhanced | NA | NA | Negative |
| Ishiwatari *et al*[19] | 58/female | Incidental | 3.0 | Head | Enhanced | Enhanced | Hypointense | Hyperintense | Negative |
| Sugawara *et al*[20] | 55/female | Incidental | 7.0 | Head | Enhanced | Enhanced | Hypointense | Hyperintense | Negative |
| Azadi *et al*[6] | 57/male | Incidental | 3.1 | Tail | Enhanced | Enhanced | Hypointense | Hyperintense |  |
| Santos *et al*[21] | 40/male | Incidental | 3.0 | Body | NA | NA | NA | NA | Negative |
| Tasdemir *et al*[11] | 24/female | Epigastric pain | 18.5 | Head | Enhanced | Enhanced | NA | NA | Negative |
| van der Vorst *et al*[22] | 67/female | Abdominal pain | 2.8 | Head | Enhanced | NA | NA | NA | Negative |
| Chen *et al*[23] | 49/female | Abdominal pain | 13.0 | Head | Enhanced | Enhanced | NA | NA | Negative |
| Hwang *et al*[24] | 53/female | Incidental | 5.2 | Head | Enhanced | Enhanced | Hypointense | Hyperintense | Negative |
| Baxter *et al*[25] | 54/female | Abdominal pain | 3.5 | Head | NA | N.A | NA | NA | CEA, CA19-9 |
| Estrella *et al*[3] | 52/female | Obstructive jaundice | 15.0 | Head | Heterogeneous | Heterogeneous | NA | NA | Negative |
| Han *et al*[26] | 77/female | Jaundice | 1.5 | Head | Enhanced | Enhanced | Hypointense | Hyperintense | Negative |
| Murakami *et al*[27] | 82/male | Hypokalemia, hypertension, edema | 6.0 | Tail | Heterogeneous | Heterogeneous | Hypointense | Hyperintense | Negative |
| Paramythiotis *et al*[28] | 55/male | Abdominal pain | 3.6 | Body | Enhanced | Enhanced | Hypointense | Hyperintense | Negative |
| Spasevska *et al*[29] | 47/male | Epigastric pain and jaundice | 3.5 | Head | Enhanced | Enhanced | N.A | N.A | CA19-9 |
| Oana *et al*[30] | 73/male | Abdominal discomfort | 7.5 | Head | Enhanced | Enhanced | Hypointense | Hyperintense | Negative |
| Current case | 48/male | Hypoglycemia | 6.5 | Body | Enhanced | Enhanced | Hypointense | Hyperintense | Negative |

CT: Computed tomography; MRI: Magnetic resonance imaging; NA: Not applicable.

**Table 2 Immunohistochemical and histological features along with outcomes of pancreatic solitary fibrous tumors**

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| **Ref.** | **Immunohistochemistry (+)** | **Histology** | **Risk assessment** | **Treatment** | **Follow-up** |
| Lüttges *et al*[13] | CD34, CD99, Bcl-2, vimentin | No necrosis or mitoses | Benign | Distal pancreatectomy | Alive and well (20 mo) |
| Chatti *et al*[14] | CD34, CD99, Bcl-2, vimentin | “Regular spindle cells” | Benign | Enucleation | Died 3 d postoperatively due to complications |
| Gardini *et al*[15] | CD34, CD99, Bcl-2, vimentin, smooth muscle actin (focal) | NA | Benign | Traverso-longmire | Alive and well (16 mo) |
| Miyamoto *et al*[10] | CD34, Bcl-2 | No necrosis or mitoses | Benign | Laparoscopic enucleation | Alive and well (7 mo) |
| Kwon *et al*[16] | CD34, CD99, vimentin | “Typical bland spindle cells” | Benign | Median segmentectomy | NA |
| Srinivasan *et al*[17] | CD34, Bcl-2 | <1 mitoses/10 HPFs, no necrosis | Benign | Distal pancreatectomy | Alive and well (7 mo) |
| Chetty *et al*[18] | CD34, CD99, Bcl-2 | No necrosis or mitoses | Benign | Whipple | Alive and well (6 mo) |
| Ishiwatari*et al*[19] | CD34, Bcl-2 | Necrosis, no mitoses | Benign | Pancreaticoduodenectomy | Alive and well (42 mo) |
| Sugawara *et al*[20] | CD34 | No necrosis or mitoses | Benign | Pancreaticoduodenectomy | NA |
| Azadi *et al*[6] | CD34, Bcl-2, Ki67 < 5% | No malignant features | Benign | Distal pancreatectomy | NA |
| Santos *et al*[21] | CD34, beta-catenin | No necrosis or mitoses | Benign | Partial pancreatectomy | NA |
| Tasdemir *et al*[11] | CD34, Bcl-2, beta-catenin, vimentin, Ki67 < 2% | 1-2 mitoses/10 HPFs | Benign | Enucleation | Alive and well (3 mo) |
| van der Vorst *et al*[22] | CD34, CD99, Bcl-2 | No necrosis or mitoses | Benign | Enucleation | NA |
| Chen *et al*[23] | CD34, Bcl-2, vimentin, CD68, muscle-specific actin | Necrosis, no mitoses | Benign | Whipple | Alive and well (30 mo) |
| Hwang *et al*[24] | CD34, Bcl-2, muscle-specific actin, CD10, ER, PR | “Spindle shaped cell with patternless cell deposition” | Benign | Duodenal preserving partial pancreatic head resection | Alive and well (30 mo) |
| Baxter *et al*[25] | CD34, Bcl-2 | NA | Benign | Whipple | NA |
| Estrella *et al*[3] | CD34, Bcl-2, keratin (rare), p16, p53 | Nuclear atypia, 17 mitoses/10 HPFs, necrosis | Malignant | Pancreaticoduodenectomy | Alive and well (40 mo) |
| Han *et al*[26] | CD34, CD99 | No necrosis or mitoses | Benign | Ultrasonography-guided needle biopsy | No metastasis or changes in the size after 10 mo |
| Murakami *et al*[27] | CD34, Bcl-2, STAT6, ACTH (focal), POMC (focal), NSE (focal) | “Spindle neoplastic cells in fascicular arrangement” | Benign | Distal pancreatectomy | Died 4 mo postoperatively due to sepsis |
| Paramythiotis *et al*[28] | CD34, CD99, Bcl-2, vimentin, S100 (focal) | No mitoses | Benign | Distal pancreatectomy | Alive and well (40 mo) |
| Spasevska *et al*[29] | CD34, vimentin, CD99, Bcl-2 (focal), nuclear beta-catenin (focal) | No necrosis or mitoses | Benign | Whipple | Died 1 wk postoperatively due to complications |
| Oana *et al*[30] | CD34, Bcl-2 | No necrosis or mitoses | Benign | Partial pancreatectomy | Alive and well (36 mo) |
| Current case | CD34, Bcl-2, STAT6, CD31, PHH-3, D2-40 and Ki67 > 10% | Necrosis, 4-5 mitoses/10 HPFs | Malignant | Distal pancreatectomy and hepatic tumor resection | Alive and well (6 mo) |

NA: Not applicable.