Solitary Fibrous Tumor of the Pancreas:  
A Case Report and Review of the Literature

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Solitary fibrous tumors (SFTs) are histologically characterized as mesenchymal tumors of probable fibroblastic origin that can arise at pleural and extrapleural sites. SFTs originating in the pancreas are extremely rare. Here, we report a case of pancreatic SFT in a 77-year-old female who presented with jaundice. A malignant neuroendocrine tumor (NET) was suspected based on radiologic findings. However, it is difficult to differentiate SFTs from a NET from radiographs and in this report, we summarize magnetic resonance imaging findings and discuss how to distinguish between SFT and NET using immunohistochemistry. Radical excision is the treatment of choice for SFT; however, in the present case, excision was not possible and close observation showed no changes 10 months after the diagnosis. (Korean J Med 2015;88:293-298)

Keywords: Pancreas; Solitary fibrous tumors

INTRODUCTION

Solitary fibrous tumors (SFTs) are found in pleural and extrapleural sites and histologically characterized as fibroblastic mesenchymal tumors. The pancreas is an extremely rare site of extrapleural SFT [1] and was first described in 1999 with only 13 cases reported to date [1-13]. Thus, there is a lack of awareness about the disease, which is difficult to diagnose. Here, we report a case of inoperable pancreatic SFT and review the diagnostic literature and discuss current treatment strategies.
CASE REPORT

A 77-year-old female presented with a 1-month history of jaundice without other symptoms. Physical examination revealed icteric sclera and yellowish skin. The patient had undergone Roux-en-Y choledochojejunostomy and left hepatectomy with cholecystectomy for intrahepatic stones and common bile duct stones 20 and 13 years before this presentation, respectively. Abnormal laboratory findings included elevated bilirubin levels (total bilirubin = 13.1 mg/dL; direct bilirubin = 8.6 mg/dL) and the alkaline phosphatase level was 709 IU/L. Tumor marker levels, including carcinoembryonic antigen and carbohydrate antigen 19-9, were within the normal range. Abdominal ultrasonography showed a 1.4 × 1.5-cm hypoechoic mass at the pancreatic head without stones. Computed tomography (CT) revealed a well-demarcated mass with enhanced contrast uptake in both arterial and venous phases on the pancreatic head. The surrounding bile duct of the pancreaticoduodenal arcade was dilated. The mass was hypointense on T1-weighted magnetic resonance (MR) images and hyperintense on T2-weighted MR images (Fig. 1). Based on these imaging findings, a malignant neuroendocrine tumor (NET) was suspected. However, fluorodeoxyglucose-positron emission tomography (FDG-PET)/CT showed no significant FDG uptake in the mass.

Ultrasonography-guided needle biopsy of the pancreas was performed. Histologic evaluation showed a lesion composed of alternating hypocellular areas composed of spindle-shaped cells and hypocellular areas with hyalinized, keloid-like fibrous tissue. Hyalinized thick bands separated the alternating hypocellular and hypopcellular areas. No cellular pleomorphism, mitosis, or necrosis was observed. These findings suggested a benign spindle cell proliferation such as SFT, inflammatory myofibroblastic

Figure 1. (A) CT scan reveals a well-demarcated mass with enhanced contrast uptake (white arrow) in arterial phase. (B) CT scan reveals a well-demarcated mass with mild enhanced contrast uptake (white arrow) in venous phase. (C) MR imaging shows a mass (white arrow) that is hypointense on T1-weighted MR images. (D) MR imaging shows a mass (white arrow) that is hyperintense on T2-weighted MR images. CT, computed tomography; MR, magnetic resonance.
tumor, or gastrointestinal stromal tumor (GIST). Immunohistochemistry revealed that CD34 and CD99 were strongly immunopositive, and c-kit and S100 were negative. The histologic and immunohistochemical findings were consistent with an SFT (Fig. 2). In the present case, radical excision was not possible because of the patient’s surgical history. Follow-up ultrasonography showed no metastasis or changes in the size after 10 months (Fig. 3).

**DISCUSSION**

The 2002 World Health Organization classification defines SFT as a ubiquitous mesenchymal tumor of probable fibroblastic type with a prominent haemangiopericytoma-like branching vascular pattern. In 65% of SFT cases, the tumor arises from the pleura, but they can also be found at other sites [1,8], although the pancreas is an extremely rare extrapleural location [3,8]. Symptoms differ according to the location and size of the mass.

Figure 2. Histologic findings of pancreatic needle biopsy. (A) The pancreatic needle biopsy was suggestive of a pancreatic mass. The pancreas (arrow head) shows a lesion (arrow) composed of interlacing spindle cells (hematoxylin-eosin [H-E] stain, ×20). (B) The lesion consisted of proliferating spindle cells arranged in an interlacing pattern (H-E stain, ×100). (C) The spindle cells showed positive immunoreactivity against anti-CD34 antibodies (×100). (D) Immunoreactivity against anti-CD 99 antibodies was also positive (×100).
Figure 3. Abdominal ultrasonography. (A) Abdominal ultrasonography shows a 1.4 × 1.5-cm hypoechoic mass at a pancreatic head without stones. (B) After 10 months, follow-up ultrasonography showed no changes in tumor size.

and include abdominal pain, constipation, jaundice, and weight loss. However, SFT is often asymptomatic because it is a benign neoplasm and slow-growing tumor [1]. In this case, the patient presented with jaundice.

On CT imaging, SFT generally shows a hypervascular, hyper-enhancing mass with contrast enhancement during both arterial and venous phases [1]. These features, which indicate a well-demarcated and well-enhanced lesion in both phases, are similar to those of NET. In contrast, MR imaging of SFT is controversial. Moreover, only pleural SFT, but not pancreatic SFT, has been detected by MR imaging. Tateishi et al. [14] analyzed the MR images of 22 patients with pleural SFT and showed that most pleural SFTs show low-intensity signal on both T1-weighted and T2-weighted images. However, MR findings of pancreatic SFTs have not been reported because this tumor is rare. Table 1 shows the MR findings of the present pancreatic SFT. Unlike pleural SFT, this pancreatic SFT showed low-intensity and high-intensity signal on both T1-weighted and T2-weighted images. High-intensity signal on T2-weighted images correspond to hemorrhage or cystic degeneration. Therefore, a pancreatic SFT showing high-intensity signal on T2-weighted images indicate high vascularity and cellularity. NET show similar signals on MR images. Therefore, SFT sometimes mimic NET [3-5,7,8]. Our radiologic findings initially suggested a NET, similar to those in other cases. However, on FDG-PET/CT, SFT shows no significant FDG uptake, whereas malignant NET is usually characterized by FDG uptake. Immunohistochemistry is useful for differentiating SFT [1]. Pathology findings show proliferating spindle cells in SFT, whereas NET is defined as epithelial neoplasms. Other differential diagnoses of SFTs include several tumors such as GIST, schwannoma, and fibromyxoid sarcoma. Histologically, SFT is characterized by alternating hypocellular and hypercellular areas separated by thick bands. Immunohistochemically, SFT expresses CD34 in 80-90% of cases and CD99 in 70% of cases. They are usually negative for c-kit, S100 protein, desmin, and cytokeratins, which are markers of GIST, schwannoma, and fibromyxoid sarcoma, respectively. CD34 immunoreactivity is also found in GIST, although SFT and GIST differ in the expression of other markers such as c-kit, which is expressed only in GIST. Therefore, in the present study, CD34 and c-kit expression was assessed in the diagnosis of SFT.

Radical excision is the treatment of choice for SFT [1,6-9]. Approximately 10-15% of SFTs are malignant. The criteria for SFT malignancy include large tumor size (> 50 mm), disseminated disease at presentation, infiltrative margins, and histologic features consistent with high cellularity, nuclear pleomorphism, areas of tumor necrosis, and an increased mitotic index (> 4 mitoses/10 high-powered fields). Malignant SFTs have reduced CD34 immunoreactivity. However, in the present case, radical ex-
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CT, computed tomography; MRI, magnetic resonance imaging; M, male; F, female; SPEN, solid pseudopapillary epithelial neoplasm.
cision was not possible and the patient was treated by close follow-up; the lesion showed no changes.

In conclusion, we report a rare case of pancreatic SFT. Radiologic imaging such as CT and MR may aid the diagnosis of pancreatic SFT. However, other types of tumor should be excluded, including NET, and microscopic and immunohistochemical studies are necessary for accurate diagnosis. In addition, all pancreatic SFT has been removed surgically, whereas the present case was followed-up by close observation with no interval change or atypia after 10 months. We recommend clinical follow-up for SFT treatment if the criteria of malignancy are not met.

**REFERENCES**