


## CASE REPORT

# Ileum preserving expanded jejunectomy and pancreaticoduodenectomy with combined resection of the superior mesenteric artery for huge retroperitoneal solitary fibrous tumor

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## Introduction

Solitary fibrous tumors (SFTs) are rare soft tissue tumors, previously regarded as hemangiopericytomas and initially described in 1942 [1]. Although usually benign, but, 10–15% of SFTs have a malignant course, characterized by metastases to other organs and recurrent disease [2]. Dedifferentiated SFTs, which have transitioned from conventional to high-grade sarcoma, are especially aggressive lesions [3]. Like other soft tissue tumors, SFTs can occur at all anastomotic sites. Bone and soft tissue sarcoma SFTs have been classified by the World Health Organization (WHO) as typical or malignant based on the number of mitoses, cellular atypia, and the presence of necrosis and hypercellularity [4]. In general, radical resection is required

### Key Clinical Message

We encountered a patient with a large retroperitoneal solitary fibrous tumor, in whom we could preserve approximately 150 cm of the ileum even after pancreaticoduodenectomy combined with resection of the superior mesenteric artery, thus preventing short bowel syndrome.

### Keywords

Ileum preserving expanded jejunectomy, pancreaticoduodenectomy, solitary fibrous tumor, superior mesenteric artery resection, two-staged operation.

for cure, because chemotherapy is ineffective in most patients. However, it may be difficult to resect these tumors completely because of their anatomical location or extent.

This report describes a woman with an extremely large retroperitoneal tumor occupying the upper abdominal cavity, compressing the pancreas, duodenum, and common bile duct. She underwent expanded jejunectomy and pancreaticoduodenectomy, combined with resection of the superior mesenteric artery (SMA), but preserving the ileum. Although extended jejunectomy and ileectomy are considered necessary after resection of an SMA, we could preserve about 150 cm of the ileum, preventing short bowel syndrome after resection. This two-staged operation, consisting of resection and reconstruction, was safe and effective in this patient.

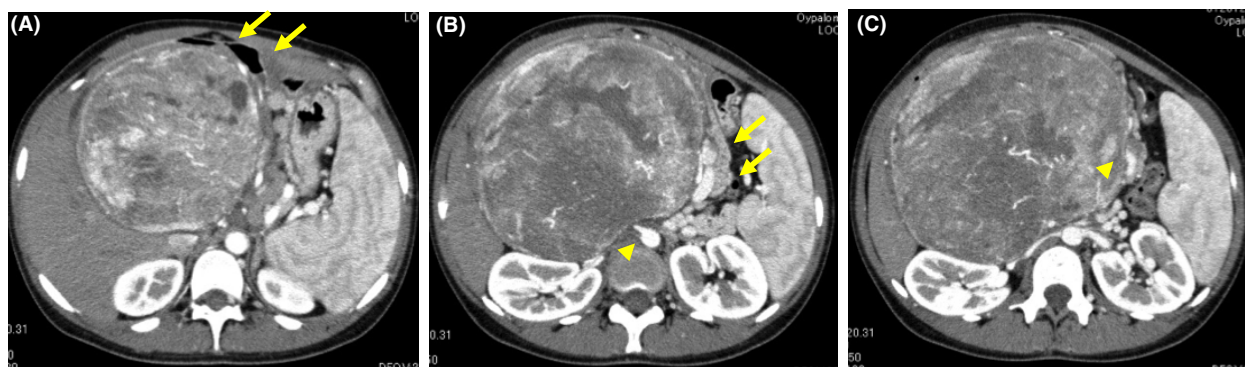
## Case and Technique

An 18-year-old woman with a 3-year history of an enlarging abdominal mass was admitted to hospital for worsened constipation, the abdominal mass, and splenomegaly. She had no relevant past medical or family history. Laboratory findings were unremarkable, except for leukocytopenia (2850/ $\mu\text{L}$ ), anemia (hemoglobin 11.7 g/dL), and thrombocytopenia ( $6.6 \times 10^4/\mu\text{L}$ ). Computed tomography (CT) revealed a heterogeneous, deeply stained huge abdominal mass compressing the stomach and duodenum anteriorly and the pancreas and main branch of the portal vein to the left side. The tumor also involved the SMA and the superior mesenteric vein (SMV) (Fig. 1A–C). T1-weighted magnetic resonance imaging (MRI) showed that the tumor was of low intensity, similar to that of the muscles, with some deep staining. Angiography revealed that the tumor was mainly fed by the gastroduodenal artery (GDA) and inferior pancreaticoduodenal artery (IPDA) and partially fed by the right subphrenic artery and anastomosis from the inferior mesenteric artery (IMA) (Fig. 2A–C). Fluorodeoxyglucose positron emission tomography (FDG PET) showed a low level of FDG uptake (maximum standardized uptake value [SUVmax] 1.6) by the lesion.

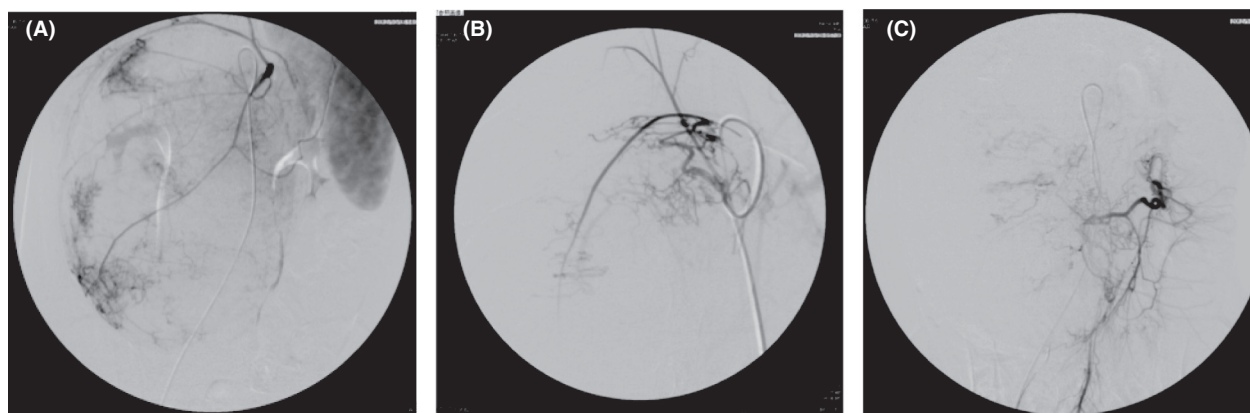
Although the tumor could not be diagnosed pathologically, as it could not be biopsied, the mass was resected because a malignancy, such as a liposarcoma, leiomyosarcoma, angiomyolipoma, gastrointestinal stromal tumor (GIST), or desmoid tumor, could not be ruled out. The tumor was very large and showed hypervascularity, suggesting that considerable bleeding would occur during surgery. Therefore, preoperative selective arterial embolization was performed, including embolization of the feeding arteries via the IPDA, GDA, right subphrenic artery, and IMA.

The tumor had compressed the pancreas, duodenum (Fig. 4A), and common bile duct. The SMA was circularly surrounded by the tumor, about 2 cm distal to the root of SMA where the artery branched from the aorta. Although the tumor invasion was not apparent, the SMV was ligated and cut at almost the same level as the SMA. Eventually, pancreaticoduodenectomy with expanded jejunectomy combined with total ligation of the SMA was required for the radical resection of a tumor of this size (Fig. 3A). Because the entire operation would generate excessive surgical stress, such as a very long operating time (17 h) and considerable blood loss (10,500 g), surgery was performed in stages. Moreover, although embolization may be effective, neovascularization due to slow progression and robust adhesion to surrounding tissue would cause considerable intraoperative bleeding and require a long operation time. The first operation included PD, expanded jejunectomy, and total ligation and resection of the SMA, along with anastomosis of the pancreatic duct with the stomach, tube gastrostomy, ileostomy, and tube-stomy of the common bile duct (Fig. 3B). Careful examination of the remnant ileum after resection of the SMA showed that the wall of the intestine was highly vascular; indicating that the remnant ileum could be preserved. Ileostomy was performed to assess the viability of the ileal stump after surgery; had the remnant ileum showed necrosis, it would have been removed.

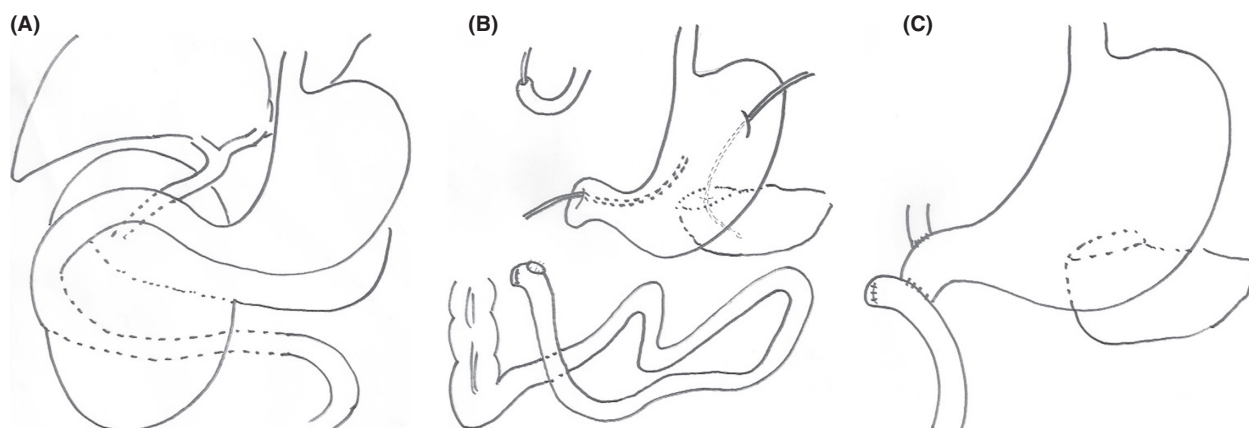
The weight of the tumor was 4.4 kg (Fig. 4B and C). Histologically, the tumor showed proliferation of spindle, oval-to-round-shaped cells with mild nuclear atypia and eosinophilic cytoplasm arranged in an unpatterned architecture, accompanied by hemangiopericytoma-like branching vessels, fibro-collagenous or myxoid stroma and cystic degeneration (Fig. 5A). Mitotic figures were observed occasionally (1/20 high-powered fields [HPF]). Immunohistochemically, the tumor cells were positive for



**Figure 1.** Contrast-enhanced CT showing (A, B) a heterogeneously stained huge abdominal mass displacing the (A) stomach and duodenum anteriorly (arrow) and (B) the pancreas and main branch of the portal vein to the left side (arrow), (B, C) as well as showing involvement of the (B) SMA (arrowhead) and (C) SMV (arrowhead).



**Figure 2.** Angiographic results showing that the tumor was fed (A) mainly by the gastroduodenal artery and (B, C) also fed by the (B) inferior pancreaticoduodenal artery and (C) right subphrenic artery and an anastomosis from the inferior mesenteric artery.



**Figure 3.** Surgical course in this patient. (A) View showing that the tumor compressed the pancreas, duodenum, and common bile duct and involved the SMA. (B) Procedure of the first operation, which included pancreaticoduodenectomy, expanded jejunectomy, and total ligation and resection of the SMA, along with anastomosis of the pancreatic duct with the stomach, tube gastrostomy, ileostomy, and tube-stomy of the common bile duct. (C) Procedure of the second operation, which included anastomosis of the common bile duct with the duodenum and anastomosis of the duodenum with the ileum.

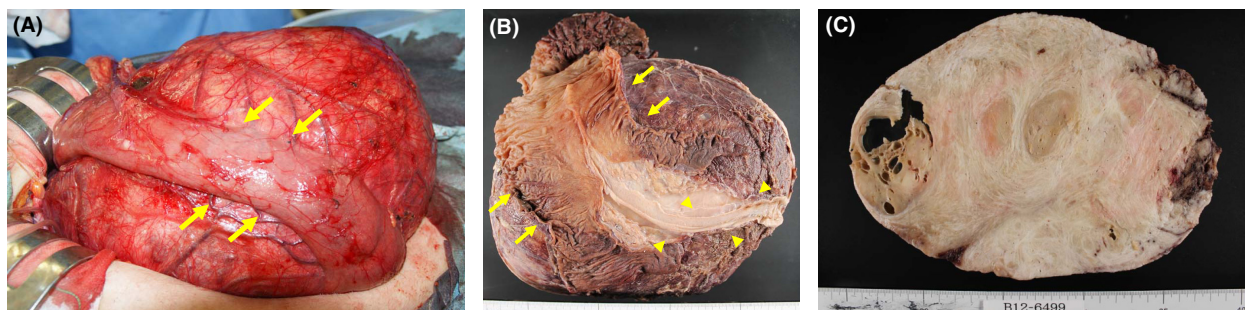
CD34 (Fig. 5B) and bcl-2, weakly positive for CD99, and negative for EMA, alpha-smooth muscle actin, and muscle-specific actin. These features indicate a solitary fibrous tumor.

The postoperative course after the first operation was uneventful, with no findings of ischemia or necrosis in the remnant ileum. A second operation, involving reconstruction of the biliary and gastrointestinal tracts, was therefore performed 5 weeks after the first operation. During the second operation, the common bile duct was anastomosed with the duodenum and the duodenum was anastomosed with the ileum (Fig. 3C). The postoperative course after the second operation was also uneventful, except for stenosis of the biliary tract and cholangitis; these were treated by insertion of temporary plastic tube stents into the anastomotic site of the bile duct and

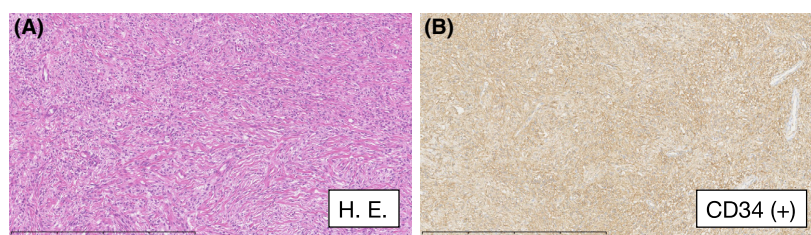
duodenum and by administration of antibiotics, respectively. The patient was discharged from the hospital 10 weeks after the second operation. Although she required treatment for stenosis between the common bile duct and duodenum, the patient has remained healthy, without any recurrence, 2 years after the operation. The plastic tube stents were removed after dilatation of that site.

## Discussion

The WHO classification of bone and soft tissue sarcomas regards SFT as typical or malignant based on the number of mitoses, cellular atypia, and the presence of necrosis and hypercellularity [4]. About 85–90% of SFTs are benign, whereas 10–15% are malignant with possible



**Figure 4.** Pathological findings. (A) Intraoperative finding, showing that the horizontal portion of the duodenum was dislocated anteriorly by the tumor. (B) View of the resected specimen, showing that it measured 21 × 18 × 15 cm in size and weighed 4400 g. The duodenum (arrow) and common bile duct (arrow head) were extremely distended. (C) Cross-sectional view of the resected specimen, showing a solid tumor with a grayish-white cut surface, a cystic component, and a hemorrhagic focus.



**Figure 5.** Postoperative findings. (A) Histopathological findings, showing oval- or spindle-shaped cells in a random growth pattern with a collagenous matrix. (B) Immunohistochemical findings, showing that the tumor was positive for CD34.

metastases to other organs and recurrent disease [2]. SFTs can occur at any anastomotic sites, but are rare in the retroperitoneal space. Although radical resection is required for curative treatment, SFTs may be difficult to resect completely because of their anatomical position and size.

The large tumor in our patient was located in the retroperitoneal space of the right upper abdomen, near the pancreas and duodenum, with the tumor causing extreme distension of the duodenum and common bile duct, dislocation of the portal vein, and involvement of the SMA and SMV. Prognostic factors for local recurrence and metastasis of SFT include positive surgical margins, tumor size >10 cm, and histological findings of malignancy, including mitosis, nuclear pleomorphism, cellularity, necrosis, and the presence of a malignant component [5]. Tumor resectability was found to be the single most important indicator of clinical outcome [6]. Complete cure requires complete tumor resection, including PD and resection of the SMA.

In general, extended jejunectomy and ileectomy are required after ligation or resection of the SMA, as in a patient who has undergone acute superior mesenteric arterial embolization. PD with SMA reconstruction can be performed safely for malignancies; however, this technique is quite complicated, and prognosis is not good

[7]. PD in a patient with stenosis of the SMA included an anastomosis between the SMA and IMA, such as Riolan's arch [8], providing retrograde blood flow from the IMA sufficient to maintain the viability of the jejunum and ileum [9]. Generally, acute occlusion of the SMA will lead to intestinal necrosis, with tumor growth resulting in the development of collateral vessels from the IMA to the SMA. Therefore, the ileum could be preserved. As one of the arteries feeding the tumor was found to flow from a branch of the IMA, blood flow to the remnant ileum could be supplied by the IMA through a retrograde pathway.

Furthermore, we found that a two-staged operation was effective in this patient. Two-staged operations have been performed on high-risk patients with esophageal cancer, including patients with severe liver dysfunction and pulmonary disease [10]. Two-staged pancreaticoduodenectomy has also been performed on some patients, including those undergoing concomitant major resection of other organs and patients with severe comorbidities [11].

Evaluating the malignant potential of an SFT is difficult. A high mitotic index or MIB1 index is suggestive of low malignant potential. This patient has remained healthy and without any tumor recurrence 2 years after the operation. However, late recurrence – including

relapse 10 years after initial diagnosis – has been reported not only in patients with malignant SFT but in those with typical SFT [12]. Careful periodic follow-up is therefore important.

In conclusion, we described the successful surgical treatment of a patient with a large SFT, by ileum-preserving-expanded jejunectomy and pancreaticoduodenectomy with combined resection of the SMA. Preservation of part of the ileum (about 150 cm in length) can prevent short bowel syndrome even after resection of the SMA.

## Authorship

AE: involved in making the study conception and design, collection of data, and writing the manuscript. YI: involved in making the study conception and design. MM, MY, and YT: involved in critical review of the article. KT: involved in pathological analysis. NK, ET, and KM: involved in collection of the data.

## Conflict of Interest

All authors have no conflict of interest to declare.

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