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Single Case

Rapidly Growing Desmoid-Type Fibromatosis of the Mesentery of the Small Intestine after Distal Gastrectomy for Gastric Cancer

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Keywords
Chylous ascites · Gastric cancer · Total gastrectomy · Roux-en-Y reconstruction · Internal hernia · Mesenteric defect · Laparoscopic surgery

Abstract
We report the case of a 55-year-old man with a surgical history of distal gastrectomy with Roux-en-Y reconstruction performed 3 years prior to the present episode. During the follow-up, a newly developed, rapidly growing intraabdominal mass was detected in the mesentery of the small intestine. Although the patient had been asymptomatic, surgical resection was planned with the suspicion of malignancy, especially lymph node recurrence of the gastric cancer, owing to its rapid growth. Laparotomy showed that the tumor was located in the mesentery of the small intestine near the Roux-en-Y limb, and due to the involvement of the feeding vessels to the Roux-en-Y limb, the anastomotic site was resected en bloc with the tumor, and the whole Roux-en-Y limb was reconstructed. The histopathological finding was compatible with desmoid-type fibromatosis of the mesentery of the small intestine. Here we report our case and discuss the previously reported literature, especially related to gastric cancer.

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Introduction

Desmoid-type fibromatosis (DF) is a rare slow-growing soft tissue tumor that shows local invasiveness without the ability to metastasize [1]. Herein, we report a case of DF developed from the mesentery of the small intestine neighboring the Roux-en-Y limb in a patient who had a surgical history of distal gastrectomy with Roux-en-Y reconstruction. Roux-en-Y reconstruction is a method of gastrointestinal reconstruction by gastrojejunostomy and creates a Roux-en-Y limb, which is a jejunojejunal anastomosis to establish intestinal continuity. We successfully resected the tumor and neighboring Roux-en-Y limb en bloc and reconstructed the whole Roux-en-Y limb. DF following gastrectomy for gastric cancer is very rare, and few cases have been reported thus far. We report our case with a review of the relevant literature.

Case Presentation

A 55-year-old man with a previous surgical history of distal gastrectomy with antecolic Roux-en Y reconstruction performed 3 years previously for gastric cancer (tub2, T1, N0, M0, pStage IA) presented with a rapidly growing intraabdominal mass that had developed during the postoperative course. The patient had not undergone adjuvant chemotherapy and had no medical or surgical history other than that of gastric cancer.

On admission, his height was 167 cm, weight was 49 kg, blood pressure was 118/73 mm Hg, heart rate was 67 beats/min, and body temperature was 36.6°C. The abdomen was soft and flat with a midline incision scar. A mobile, elastic, hard mass was palpable on the left side of the abdomen. His complete blood count and serum chemistry results were normal, and all of the examined tumor markers (carbohydrate antigen 19-9, carcinoembryonic antigen, α-fetoprotein, neuron-specific enolase, cytokeratin 19 fragment, and soluble interleukin-2 receptor) were within their normal limits.

Abdominal contrast-enhanced computed tomography (CT) initially detected a newly appearing small mass measuring 14 mm, neighboring the Roux-en-Y limb anastomosis without invasion into the small bowel 22 months previously (although this was confirmed only via retrospective observation). The mass had increased in size from 33 mm (6 months before) to 82 mm at the time of the operation (Fig. 1). On the preoperative CT, the tumor was 82 × 75 mm in size, and it appeared to originate from the mesentery of the small intestine. We determined the most likely differential diagnosis of gastrointestinal stromal tumor and lymph node recurrence of the gastric cancer, and DF was not included in the list. ^{18}F-2-fluoro-2-deoxyglucose positron emission tomography (PET) revealed a heterogeneously enhanced uptake on the lesion (SUV_{max} = 4.40) in the lower abdomen. The mobility of the tumor from the previous CTs also supported the origin of the mesentery. Apart from this, there were no significant findings that indicated metastasis or any other lesions (Fig. 2). Biopsy was considered because chemotherapy, not surgery, may be needed in case of recurrence of the gastric cancer. However, this strategy was abandoned owing to the lack of a suitable route for a percutaneous or endoscopy-assisted approach.

Overall, we suspected that a malignant lesion was located in the mesentery near the Roux-en-Y limb that was responsible for its rapid growth. There was no other lesion elsewhere; therefore, we decided to perform resection for both diagnosis and treatment.

Laparotomy showed that the tumor was located in the mesentery of the small intestine near the Roux-en-Y limb. The feeding vessels for the Roux-en-Y limb anastomotic site were partly involved by the tumor; therefore, the anastomotic site was resected en bloc with the

tumor, and the whole Roux-en-Y limb was reconstructed with functional end-to-end anastomosis using a linear stapler. No intraperitoneal dissemination or distant metastasis was observed during the surgery.

Macroscopically, the resected specimen was a 10 × 9 × 8 cm-sized solid lesion with no necrosis or bleeding (Fig. 3). No exposure to the mucosa of the adjacent small intestine was observed, indicating its origin in the mesentery. Histopathological findings revealed that the tumor was located in the mesentery of the small intestine, and spindle-shaped cells resembling fibroblasts or myofibroblasts proliferated sparsely with intervening collagen fibers. Invasive proliferation into the surrounding fat tissue was observed in the marginal area of the tumor (Fig. 4). The immunohistochemistry results were negative for desmin, α-smooth muscle actin, CD34, and c-kit. Moreover, positivity for the nuclear accumulation of β-catenin was observed (Fig. 4). In accordance with our findings, DF of the mesentery was diagnosed.

The postoperative course was uneventful, and the patient was discharged on postoperative day 13. At the time of writing of this report, the patient was free from recurrence with no adjuvant treatment and was being carefully followed up with CT.

Discussion

DF, also called desmoid tumor, deep fibromatosis, or aggressive fibromatosis, is a slow-growing soft tissue tumor that is clinically characterized as locally invasive but without the potential to metastasize [1]. Most cases of DF occur sporadically; however, 5% are observed in patients with familial adenomatous polyposis (FAP), an autosomal dominant inherited cancer predisposition syndrome caused by germline mutation of the adenomatous polyposis coli (APC) gene [2–4].

DF may arise at any anatomical site; however, about 10% of sporadic DF affects intraabdominal organs [5, 6]. It usually occurs in the mesentery, retroperitoneum, or omentum; however, internal organs, such as the pancreas [7] and the stomach [8], are also reported to be involved. Intraabdominal DF shows a worse prognosis owing to the proximity to vital organs, resulting in grave consequences in cases of invasions, such as aneurysm formation [9] and bowel obstruction [10].

The complete etiology of DF remains unclear; however, the activating mutation of *CTNNB1* (which encodes β-catenin) is observed in about 85–90% of DF patients, leading to nuclear accumulation of β-catenin [11]. Both loss of APC in FAP patients and activation of *CTNNB1* mutation lead to the continuous stimulation of the Wnt/β-catenin pathway [12]; this contributes to aberrant cell proliferation. It is noteworthy that the mutation of β-catenin and APC is mutually exclusive; hence, non-FAP DF patients may be recommended for FAP screening [13].

Furthermore, the upregulation of estrogen aggravates the disease, which explains the sex preference for females, especially during pregnancy and before menopause, and the direct effectiveness of tamoxifen, an inhibitor of estrogen, in DF [14].

It is noteworthy that a history of trauma before the surgery is a validated risk factor [15]. Carothers et al. [16] found that DF is caused by acquisition of a proliferative benefit for mesenchymal stem cells in a wound healing setting by continuous stimulation of the Wnt signaling pathway. Moreover, the histology is almost indistinguishable from that of hypertrophic scars or keloids of the skin. The analogy is also applicable, such as high rates of recurrence after resection, predilection for younger females, and continuous activation of the Wnt signaling

pathway [17, 18]. Therefore, further research on the analogy between these conditions is warranted.

The treatment for DF is shifting toward a nonsurgical approach (the "wait-and-watch" approach) for nonprogressing and asymptomatic patients, because of its high recurrence rate after radical resection [19, 20]. However, close follow-up is required for DF owing to the variable and unpredictable natural history; on DF progression, surgical resection is considered.

For surgical resection, local recurrence is the center of interest for its nonnegligibly high rate. Since the tumor has no capsule and due to the infiltrative invasion at the boundaries, the intraoperative determination of R0 resection remains challenging. A study on the challenge faced by operating surgeons that included 426 patients showed that R0 and R1 resection had no significant impact on progression-free survival. Further, recurrent DF is in itself a risk factor for recurrence [21]. However, in a study of 189 patients who underwent surgery for DF, R0 resection showed a significantly lower recurrence rate than R1 resection (27 vs. 54%; p = 0.03) [22]. Thus, when the surgery is planned, surgeons should make their best effort to secure a clear margin from the tumor.

DF after gastrectomy for gastric cancer has been reported by a few authors [23–27]. This condition is very challenging for clinicians because the treatment strategy differs greatly for DF and cancer recurrence. Most patients were presumptively diagnosed with gastric cancer recurrence. This indicates a high prevalence of false diagnosis of DF, given that no effort to obtain the specimen is made when recurrence is diagnosed on imaging studies. The lesson learnt from the literature and the present case is that when there is a novel lesion suspected to indicate lymph node recurrence with no apparent metastasis elsewhere during the post-gastrectomy period, resection is recommended for both diagnosis and treatment, because DF is more manageable if there is no invasion into the vital organs and completely lacks metastatic capability.

Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Disclosure Statement

The authors have no conflicts of interest to declare.

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Author Contributions

R.K. is the primary investigator and contributed to conceptualization, data collection, and drafting of the manuscript. All authors have read and approved this manuscript for publication.

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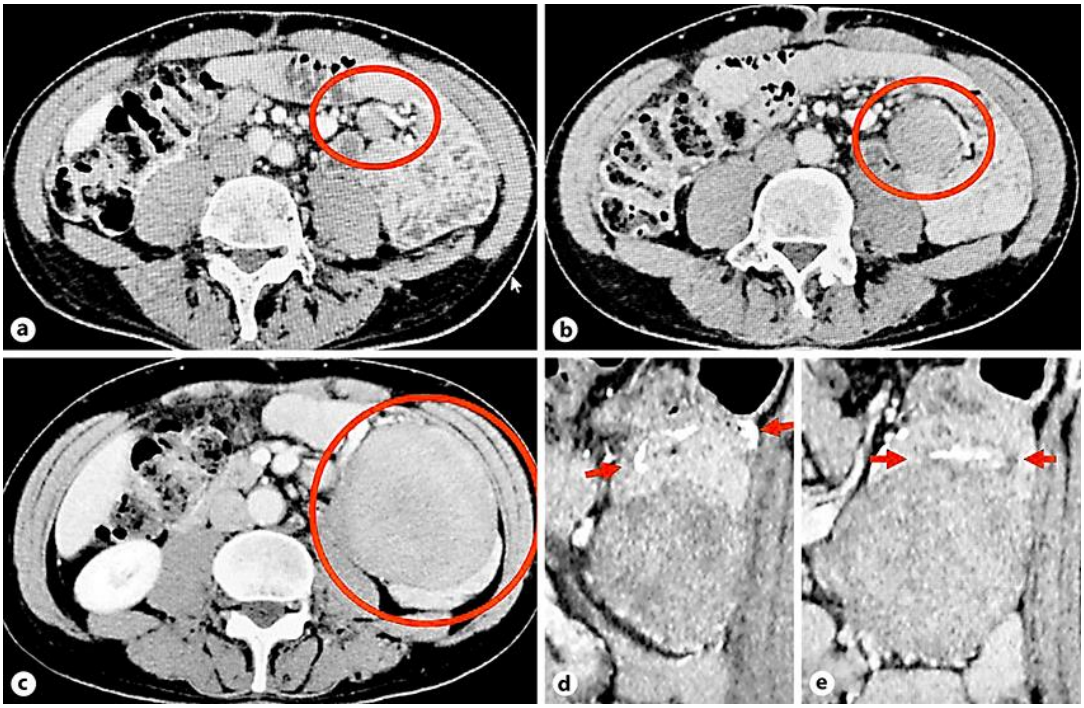

Fig. 1. Computed tomography (CT) images. **a** A new, 14-mm mass appeared adjacent to the Roux-en-Y limb without invasion into the small bowel (22 months ago; circle: tumor). **b** The tumor increased in size to 33 mm 6 months ago (circle: tumor). **c–e** Preoperative CT showed a heterogeneously enhanced tumor with a diameter of 82 × 75 mm by axial (**c**) and coronal imaging (**d**, **e**) (arrows: anastomotic site; circle: tumor).

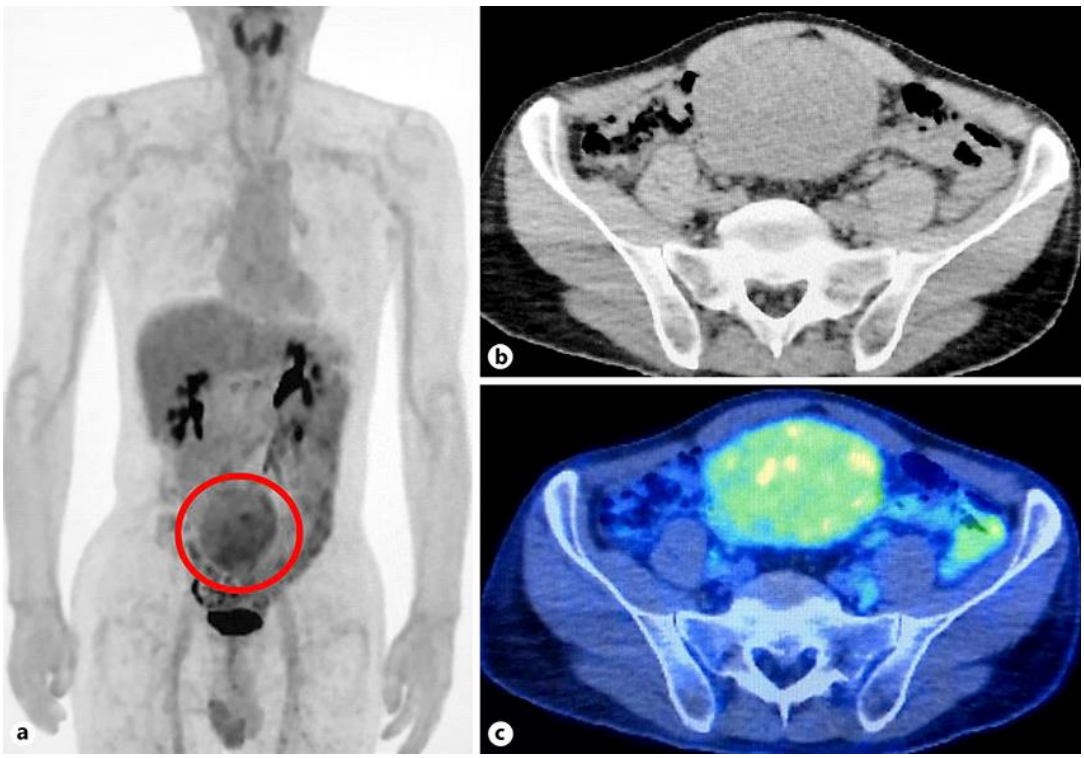

Fig. 2. Positron emission tomography (PET) images. The tumor (circle) showed a heterogeneously enhanced uptake on the lesion (SUV_{max} = 4.40) in the pelvis. No significant findings indicating metastasis or any other lesions were observed. **a** Whole body scan. **b**, **c** Axial images.

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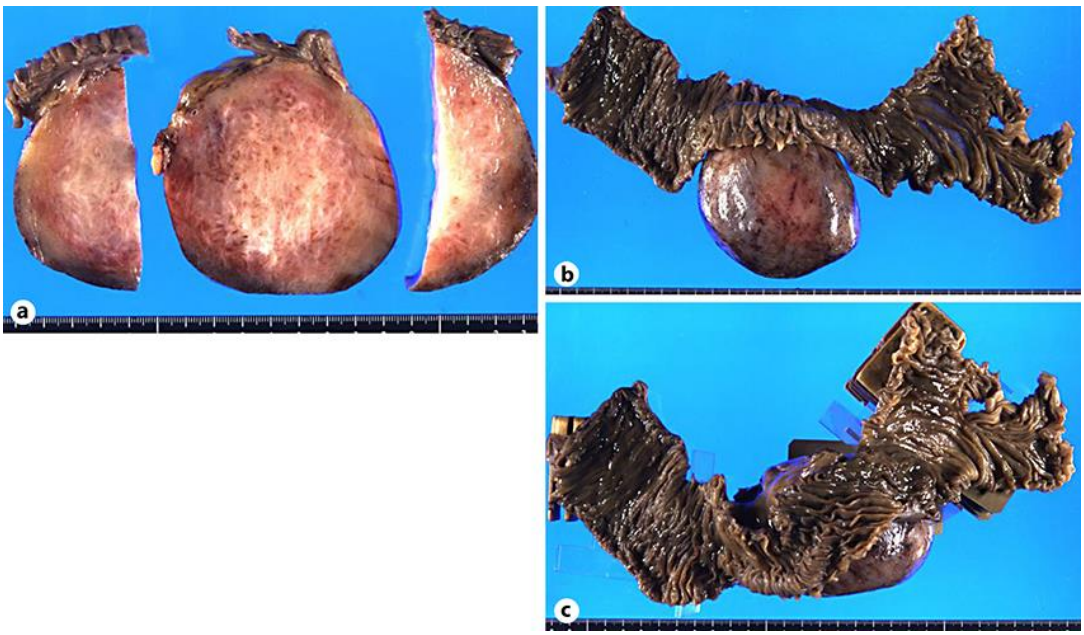

Fig. 3. Macroscopic findings in the resected specimen. **a** The resected specimen was a solid lesion with no necrosis or bleeding, measuring 10 × 9 × 8 cm. **b**, **c** No exposure to the mucosa of the adjacent small intestine was observed, indicating its origin in the mesentery of the small intestine.

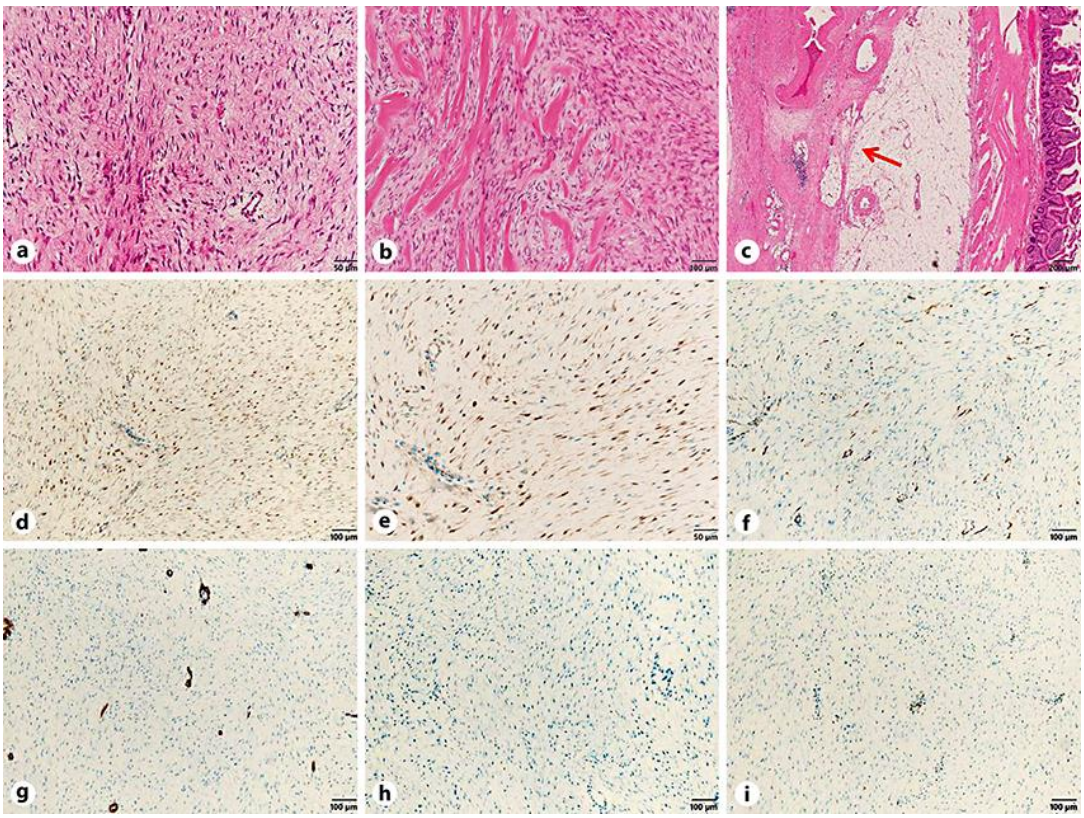

Fig. 4. Histopathology of the resected specimen. **a**, **b** Spindle-shaped cells resembling fibroblasts or myofibroblasts proliferated sparsely with intervening collagen fibers. **c** Invasive proliferation into the surrounding fat tissue was observed in the marginal area of the tumor (arrow). There was no continuity between the tumor (on the left side) and the wall of the small intestine (on the right side). **d–i** The immunohistochemistry results showed positivity for nuclear accumulation of β-catenin (**d**, **e**), and negativity for α-smooth muscle actin (**f**), CD34 (**g**), desmin (**h**), and c-kit (**i**).