Clinical Case Report

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A giant mesenteric fibromatosis involving the muscular layer of the colon wall

A case report

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Abstract

Rationale: Mesenteric fibromatosis (MF) is a rare tumor whose biological behavior is intermediate between benign fibrous neoplasms and fibrosarcomas, and the characteristic of these tumors are local aggressive lesions which is prone to local recurrence but non-metastasizing. The common symptom is abdominal distention or painless mass. We report a case of giant MF in abdominal cavity with abdominal distention as the main symptom.

Patient concerns: A 26-year-old male presented with 2-month history of abdominal distention, lack of appetite, and symptoms grew progressively more debilitating with time.

Diagnoses: This patient underwent a contrast-enhanced computed tomography scan which showed a giant ($37 \times 25 \times 13$ cm), inhomogeneous enhancing, well-defined, and soft tissue density mass in abdominal cavity, possibly arising in mesocolon, which suggested a high possibility of MF. The postoperative pathology showed that the tumor cells to be positive for β-catenin, vimentin, negative for CD34, CD117, DOG-1, S-100, Desmin, which confirmed the diagnosis of MF.

Interventions: Exploratory laparotomy was performed, which revealed a large mass involving the transverse colon wall, the root of mesocolon, and encasing the middle colic vessels and the 1st branch of jejunal arteries. The complete surgical resection was performed and the mass weighted 10 kilograms (kg).

Outcomes: The patient recovered uneventfully and was discharged 9 days after surgery. Three-month, 6-month, 12-month and 18-month on follow-up after surgery, showed no evidence of recurrence.

Lessons: The MF is a very rare tumor, especially a giant tumor (10 kg) involving the muscular layer of colon wall. In addition, treatment of giant MF still remains a challenge. We consider that surgical resection with negative margins is the goal but not at the expense of damaging the function of vital organs. Specific measures should be considered based on the individual patient in order to relieve symptoms and improve quality of life.

Abbreviations: CA-199 = carbohydrate antigen-199, CA-72-4 = carbohydrate antigen-72-4, CT = computed tomography, GIST = gastrointestinal stromal tumor, HIFU = high-intensity focused ultrasound, MF = mesenteric fibromatosis.

Keywords: abdominal tumor, mesenteric fibromatosis, prognostic factor, treatment

1. Introduction

Mesenteric fibromatosis (MF) is a rare tumor that constitutes nearly 0.03% of all tumors.[1] This tumor is characterized by monoclonal proliferations of spindle cells, which include fibroblasts and myofibroblasts, and its biological behavior is intermediate between benign fibrous neoplasms and fibrosarcomas. Another characteristic of these tumors are local aggressive lesions that are prone to local recurrence but non-metastasizing.[2] The MF occurs most commonly in the mesenteric of small bowel or retroperitoneum and is less likely involved in the mesocolon and omentum. This tumor may remain asymptomatic in the early period and usually manifest as a painless mass. Because of the tumor's characteristic of local infiltration of adjacent organs, symptoms caused by compression can occur as the tumor grows. At present, the precise etiology of this tumor is still undefined, and surgical resection remains the appropriate treatment for intra-abdominal fibromatosis in most cases. To our knowledge, a giant MF (weighing 10 kg) infiltrating the muscular layer of the transverse colon wall and root of the mesocolon has been rarely reported in the literature, and we describe such a case herein.

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2. Case presentation

2.1. Patient information

A 26-year-old male presented with a 2-month history of abdominal distention, lack of appetite, and symptoms that grew progressively more debilitating over time. However, there was no obvious abdominal pain, hyperpyrexia, weight loss, fatigue weakness, or edema in this patient. He denied past abdominal surgery, trauma and a family history of Gardner's syndrome. On physical examination, this patient had a slightly swollen abdomen, and with a painless mass in the abdomen. The mass had an irregular shape, and it was rubbery with a hard consistency. In addition, the hematological profile, such as the leukocyte count, platelet count, percent of neutrophils and serum albumin level, was normal. The levels of tumor markers, for instance, alpha-fetoprotein, carcinoembryonic antigen, and carbohydrate antigen (CA)-199, were also normal, but the level of CA-72-4 was slightly elevated (21.31 U/ml; normal range in our hospital, 0–5.3 U/ml).

2.2. Imaging findings

Contrast-enhanced computed tomography (CT) scan showed a giant, inhomogeneous enhancing, well-defined, soft-tissue density mass possibly arising from the mesocolon and measuring $37 \times 25 \times 13$ cm in the abdominal cavity (Fig. 1). The nearby organs and tissues had been compressed and pushed away from their original location. The preoperative differential diagnosis was challenging for us to make; thus, we focused on liposarcoma, lymphoma, fibrosarcoma, and gastrointestinal stromal tumor (GIST) as possibilities.

2.3. Therapeutic interventions

During laparotomy, a giant mesenteric mass was associated with the root of the mesocolon and transverse colon (the mucosal folds disappeared at a local range). This tumor was removed intact with a segment of the jejunal and transverse colon (Figs. 2**A** and 2**B**). We performed 1-stage end-to-side and end-to-end anastomosis for the jejunum and colon, respectively. The irregular mass weighed 10 kg, and the multiple cut surfaces of this mass were incanus or isabelline, with minor hemorrhage and necrosis (Fig. 2**C**).

2.4. Histopathological findings

The microscopic examination revealed sparse spindle cells surrounded by abundant fibrillar collagen, with minimal mitotic activity, few atypia, edema, and minimal hemorrhaging of the

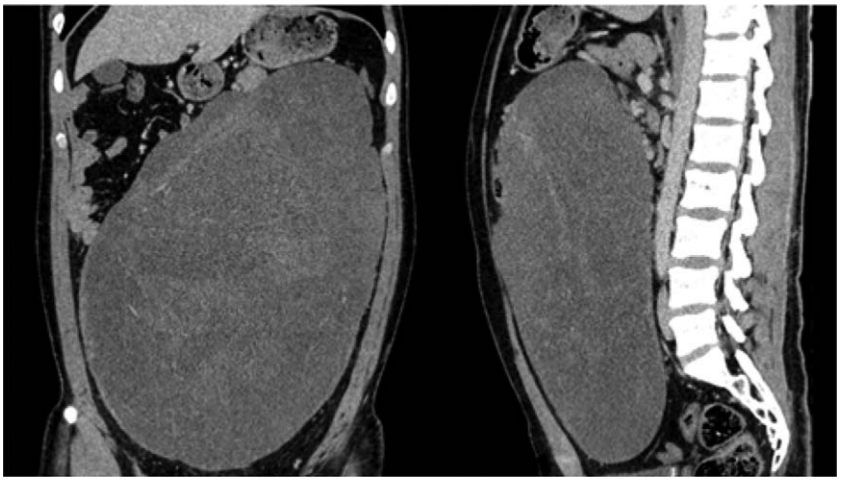

Figure 1. Contrast-enhanced computed tomography scan showing a giant mass in the abdominal cavity.

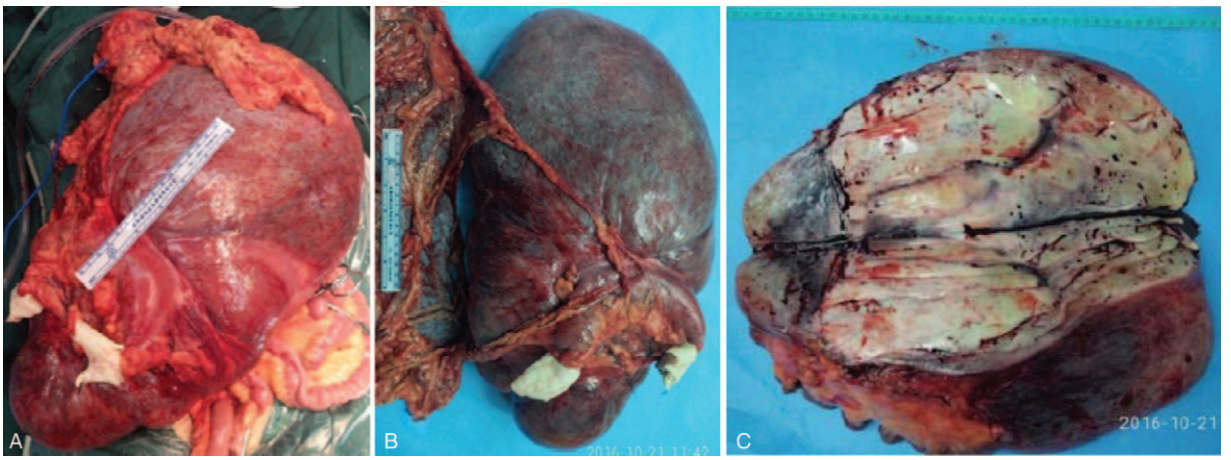

Figure 2. Photographs of the tumor. (A) Surgical exposure of the tumor; (B) removal of tumor mass; and (C) multiple cut surfaces of the tumor are incanous or isabelline, with minor hemorrhage and necrosis.

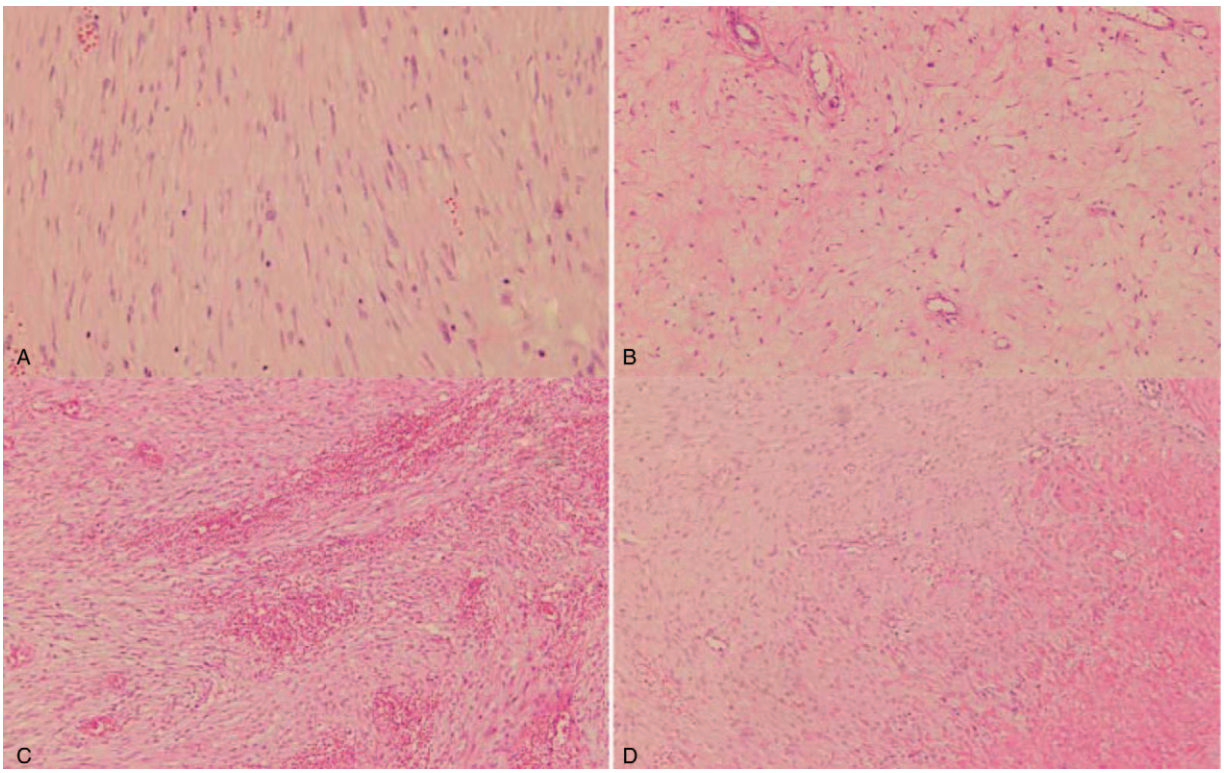

Figure 3. Microscopic examination showing (A) spindle cells surrounded by abundant fibrillar collagen (H&E ×200); (B) edema in a certain part of the tumor tissue; (C) minor hemorrhage in the tumor tissue (H&E ×100); and (D) tumor tissue involving the muscular layer of the transverse colon wall (H&E ×100).

tumor tissue. In addition, this tumor infiltrated the muscular layer of the transverse colon wall (Figs. 3**A**, 3**B**, 3**C**, and 3**D**). Immunohistochemistry revealed that the tumor cells were positive for β-catenin, vimentin and negative for CD34, CD117, DOG-1, S-100, and desmin (Fig. 4**A**, 4**B**, and 4**C**).

2.5. Follow-up and outcomes

The patient recovered uneventfully and was discharged 9 days after surgery. Three-month, 6-month, 12-month, and 18-month on follow-up after surgery, showed no evidence of recurrence.

3. Ethics statement

Case reports are exempt from formal ethical approval in our hospital. Informed written consent was obtained from the patient for publication of this case report.

4. Discussion

The MF is the most common primary tumor of the mesentery, and these tumors may occur in all ages, but they are especially seen in the 3rd and 4th decades of life.[3] The certain etiopathogenesis of these tumors is unknown, however, various factors may contribute to the formation of these tumors, such as trauma, especially operative trauma (e.g. MF may develop after the surgical resection of GIST), the estrogen level, genetic predisposition (Gardner's syndrome and familial adenomatous polyposis), infectious etiology (human herpes virus), and autoimmune diseases (Crohn's diseases).[4,5] Nonetheless, there were no identifiable risk factors in our case.

The MF is associated with a broad range of clinical manifestations, including fever of unknown origin, abdominal pain, nausea, vomiting, tarry stool, stoppage of bowel movements,

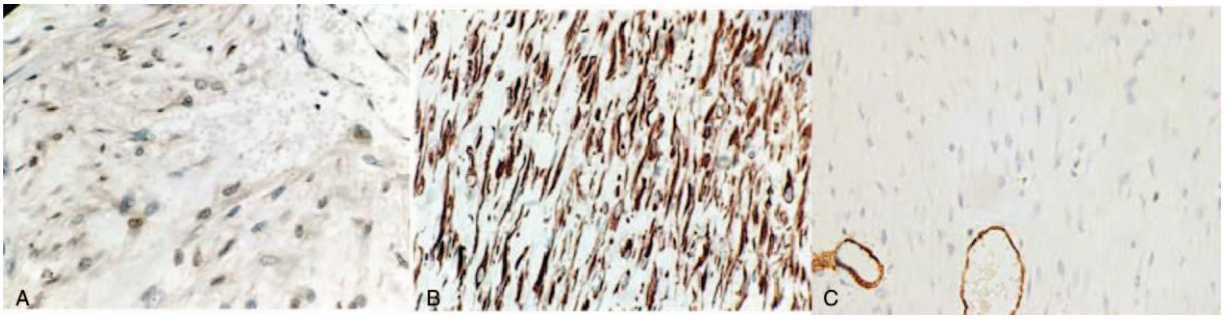

Figure 4. Immunohistochemistry findings. (A) The cell nucleus is positivity for the β-catenin antibody (×400); (B) cell cytoplasm is positivity for the vimentin antibody (×400); and (C) tumor cells are negative for CD34 (×400).

abnormalities of urine, and emaciation.[6–8] In addition, MF may manifest as a surgical emergency requiring immediate surgical intervention.[9] Our patient presented with a 2-month history of abdominal distention and lack of appetite. The CT images indicated the possibility of treating the tumor with complete surgical resection. Therefore, we decided to perform exploratory laparotomy.

The final diagnosis depends on the pathology, which remains the gold standard for diagnosis. In our case, immunohistochemistry revealed the tumor cells to be positive for β-catenin and vimentin, and negative for CD34, CD117, DOG-1, S-100, and desmin, which are characteristic of intro-abdominal MF.[10] However, MF should be differentiated from other diseases in the abdominal cavity or retroperitoneum, such as an inflammatory lesion, liposarcoma, lymphoma, fibrosarcoma, and GIST. Additionally, the level of the inflammatory component, MDM-2 gene, multiple enlarged lymph nodes, CD117 positivity and absence nuclear β-catenin, which may help physicians recognize the aforementioned tumors.

Until now, well-defined and precise guidelines of the optimal treatment for MF have not been formulated in the medical community. Most literature that has been published includes case reports, and the treatment strategies are usually empirical. Mullen et al[11] revealed that the margin status remained an independent risk factor for local recurrence in multivariate analysis of a retrospective review of 177 patients, among whom 22 patients had intra-abdominal desmoid tumors. In a retrospective review of 211 patients, among whom 23% of patients had intra-abdominal desmoid tumors, Peng et al[12] suggested that negative margins may obtain a longer recurrence-free survival, and that the tumor location and patient age may affect the prognosis or recurrence. Further, the mutation status of the β-catenin gene was recently revealed to have a significant effect the outcome after surgical treatment.[13] Wilkinson et al[14] reported the non-familial adenomatous polyposis-associated intra-abdominal fibromatosis may have a lower recurrence rate after radical surgical resection. Hence, radical surgical resection has been the mainstay treatment in most cases.

The MF may not be detected until later, and it has a high local recurrence rate; thus, the treatment strategies are usually difficult to perform and have unsatisfactory results. The MF is rarely small enough or appropriately for complete resection. These tumors usually invade the root of the mesentery and mesenteric vessels, and short gut syndrome may occur if a considerable length of the small bowel is removed to achieve complete surgical resection, which may significantly increase the risk of morbidity and negatively affect the quality of life for some patients. At the same time, some researchers have suggested that the risk of local recurrence still remained, and ranged from 25 to 50% in most studies of complete surgical resections.[15,16] The surgical operation itself is a kind of trauma that may lead to further recurrence. In addition, the unpredictable nature of these tumors and the factors associated with recurrence are inconsistent. Dalén et al[17] reviewed 5 of 8 patients with intro-abdominal desmoid tumors who received no treatment, and they concluded that some types of desmoid tumors with few or no symptoms may spontaneously decrease or disappear, which has led the researchers to assess the role of a wait-and-see policy. Regardless, we still used the radical surgical resection strategy in our case because the preoperative imaging assessment indicated that we would be able to completely remove the tumor. Moreover, the rapid growth of giant mass would cause serious consequences if we were unable to take effective measures.

Radiotherapy, systemic therapy (including tamoxifen, nonsteroidal anti-inflammatory drugs, and chemotherapy), molecular-targeted therapy, ultrasound-guided high-intensity focused ultrasound (HIFU) ablation may be valuable options in treating MF for patients who are unable to undergo surgery, positive histological margins after surgical resection, postoperative recurrence, and surgery may lead to unacceptable damage of the function of vital organs.[18–22] In our case, we performed complete surgical resection of the tumor, and we did not prescribe further adjuvant treatment, except routine follow-up.

Our experience showed an uncommon, huge MF in the abdominal cavity. To our knowledge, this is the first documented case of tumor tissue invading the muscular layer of the transverse colon wall and root of the mesocolon. At present, a giant MF remains a challenge for patients and surgeons. We consider that surgical resection with negative margins is the goal but not at the expense of damaging the function of vital organs. Specific measures, such as radiotherapy, anti-estrogenic therapy, nonsteroidal anti-inflammatory drugs, molecular-targeted therapy, and HIFU, should also be considered based on the individual patient in order to relieve symptoms and improve quality of life.

Author contributions

Haibin Ji, Wentao Zhu, and Baolei Zhao contributed to the literature research and drafting the manuscript. Jian Shi and Qiang Wei contributed to providing the relevant images. Baofang Sun contributed to collecting the clinical data. Qiangpu Chen contributed to revising the manuscript and editing the figures. All authors have approved the final version of the manuscript.
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