Clinical Case Report

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Rare acute abdominal condition caused by mesenteric fibromatosis perforation

A case report

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Abstract

Rationale: Mesenteric fibromatosis is a rare benign neoplasm with a tendency to spread and recur locally, without metastasis. It may present with a wide spectrum of clinical features; however, onset as a perforation is extremely rare.

Patient concerns: The present patient was an 18-year-old female with a 10-hour history of increasing abdominal pain that arose suddenly with nausea and vomiting. She had experienced an appendectomy 2 years before this admission.

Diagnoses: A gastrointestinal perforation was initially suspected on the basis of complaints and physical examination. The patient was thoroughly investigated for further diagnosis. Computed tomography showed a large well-defined intra-abdominal mass measuring $7.1 \times 6.7 \times 5.9$ cm in the right lower quadrant, with adjacent small intestine compression and free intraperitoneal air. Then, the patient underwent a laparotomy. Finally, postoperative pathology and immunohistochemistry confirmed mesenteric fibromatosis, with a consecutive perforation from ileum to the bottom of tumor.

Interventions: The patient has been treated by a resection of the mass with the adhesive small intestine, without chemotherapy or radiotherapy postoperatively.

Outcomes: The patient had an uneventful postoperative recovery. Three months after surgery, the patient reviewed the colonoscopy, no intestinal polyps were noted. The present case has been followed up for 17 months without tumor recurrence.

Lessons: Our case illustrates another possible cause of acute abdominal pain. Although rare, treating physicians should maintain a high suspicion index while managing a patient with an abdominal mass and pain. Close follow-up is essential because of the high incidence of local tumor recurrence.

Abbreviations: CD = cluster of differentiation, FAP = familial adenomatous polyposis, GIST = gastrointestinal stromal tumor, MM = mesenteric fibromatosis, NSAIDs = non-steroidal anti-inflammatories drugs.

Keywords: acute abdomen, mesenteric fibromatosis, perforation

1. Introduction

Mesenteric fibromatosis (MF) is a rare benign proliferation of the mesentery fibrous that tends to spread and recur locally, without metastasis. The exact etiopathogenesis of MF is not known. Primary or spontaneous MF is rare, and secondary MF most commonly occurs after or accompany trauma, previous abdominal surgery, pregnancy,[1] Garden Syndrome, and Crohns disease.[2] With a wide range of differential diagnose, including gastrointestinal stromal tumors (GISTs), lymphoma, carcinoid tumor, fibrosarcoma, inflammatory fibroid polyp, this condition poses a diagnostic and therapeutic challenge to clinicians. MF may present with a wide spectrum of clinical features, however, its onset as a perforation is extremely rare.

The present report outlines the case of a patient with MF who presented with an ileal perforation. Our case illustrates another possible cause of acute abdominal pain. We also conducted a comprehensive review of international literature to summarize the clinical characteristics of MF.

2. Case report

We present an 18-year-old woman who was admitted to our institution with a 10-hour history of increasing abdominal pain that arose suddenly with nausea and vomiting. She had experienced an appendectomy 2 years before this admission and denied melena or chronic nonspecific abdominal pain. She had no history of pregnancy and intestinal polyposis. On admission, her baseline observations were normal. An abdominal examination revealed tenderness, rigidity, and rebound tenderness. Her liver dullness decreased, and bowel sounds disappeared, her systemic examination was normal.

The patient was thoroughly investigated. A complete blood count revealed leukocytosis (18.10×10^9/L) with neutrophilia (16.05×10^9/L). Her routine biochemistry results and human

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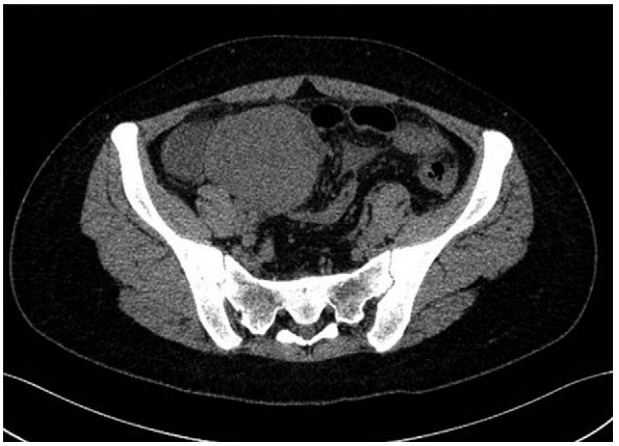

Figure 1. CT picture showing a large well-defined intra-abdominal mass in the right lower quadrant with adjacent small intestine compressed. CT = computed tomography.

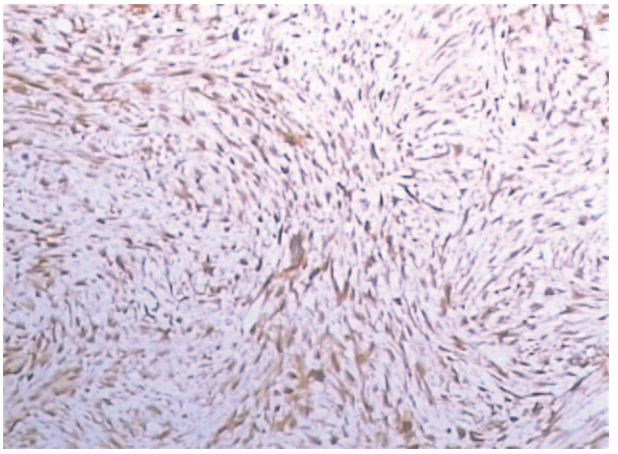

Figure 3. The tumor was immunoreactive to vimentin and β-catenin.

chorionic gonadotropin levels were normal, and the ultrasound revealed a right adnexal mass (teratoma). Computed tomography was performed, and a large well-defined intra-abdominal mass measuring $7.1 \times 6.7 \times 5.9$ cm in the right lower quadrant, with adjacent small intestine compression and free intraperitoneal air, was detected (shown in Fig. 1). A mild amount of ascites was presented in the pelvic cavity. There was no dilatation of the intestine.

A laparotomy was performed, which revealed a massive growth occupying the terminal ileum mesentery. The adjacent terminal ileum was compressed and tightly adhered to the tumor. A rupture was found at the bottom of the tumor, and about 300 mL pus was detected in the abdominal cavity. The mass and the adhesive small intestine were dissected, and anastomosis of the ileum and ascending colon was performed to reconstruct the digestive duct. A consecutive perforation, from ileum to the bottom of tumor, was discovered in the specimen (shown in Fig. 2). The histopathology of the specimen revealed a tumor composed of spindle cells arranged in a storiform pattern. The tumor was immunoreactive to vimentin and β-catenin and CD-117-negative (shown in Fig. 3). A final histological diagnosis of MF was established, with no infiltration to the adjacent small intestine. The patient had an uneventful postoperative recovery and she was discharged on the tenth postoperative day. Three months after surgery, the patient reviewed the colonoscopy, no intestinal polyps were noted. The present case has been followed up for 17 months without tumor recurrence.

The patient provided informed consent for the publication of her clinical data. Medical ethical committee approval of our report was not necessary because this is a case report.

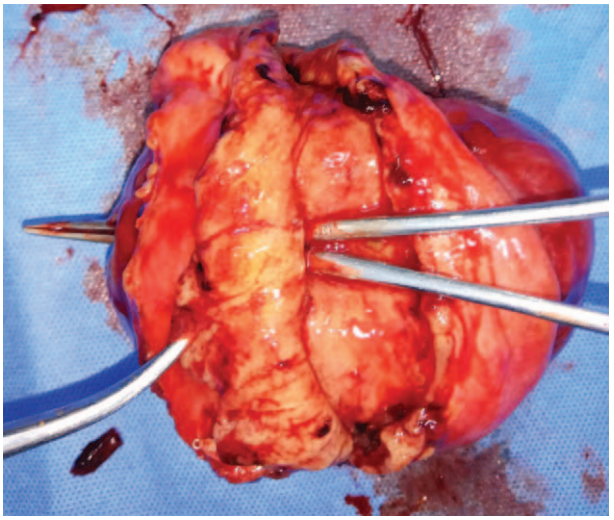

Figure 2. A consecutive perforation from ileum to the bottom of tumor.

3. Discussion

Fibromatosis are rare tumors and are also known as desmoid tumors, derived from the Greek word "desmos" meaning band or tendon.[3] They arise from musculoaponeurotic elements due to fibroblast cell mutations, and account for about 3.5% of all fibrous tissue neoplasms and 0.03% of all neoplasms, with the incidence of 2 to 4 cases per million people.[4,5] Fibromatose can occur in the superficial and deep parts of the body. Superficial fibromatose affect the face and neck (fibromatosis coli), palms (Duputyren contracture), feet (Ledderhose disease), penis (Peyronie disease), shoulder, thigh, buttock, and trunk. Deep fibromatose are classified into abdominal, extra-abdominal (desmoids outside the abdominal wall), and intra-abdominal types.[6]

MF is a type of intra-abdominal fibromatosis that accounts for about only 8% of all desmoid neoplasms. Its biological behavior follows benign fibroproliferative processes, however, it can be locally aggressive and have the capacity to infiltrate or recur without metastasis.[7] Based on the clinical course, MF is classified into 5 categories: spontaneous regression, stable, variable growth, progressive growth, and aggressive growth.[8]

Most commonly, MF lesions occur in the mesostenium, then the omentum, and mesocolon.[9] The fourth decade of life is the typical age of disease onset, but MF has been reported in patients in the age range of 14 to 75 years of age, without any racial preference.[7]

The exact etiopathogenesis of MF is unknown, but various factors are correlated with this disease, including trisomy of chromosomes 8 or 20,[10] trauma, previous abdominal surgery,

hormonal stimulation,[1] Garden Syndrome, and Crohns disease.[2] MF occurs more frequently in females, especially during pregnancy and premenopausal periods, since estrogen that predisposes one to MF and plays a role in MF formation.[11] A strong association has also been reported between MF and familial adenomatous polyposis (FAP), especially Gardner syndrome.[12] Approximately 13% of the patients with MF have FAP, specifically the Gardner syndrome variant of FAP.[7] Therefore, patients with FAP and a family history of MF have a greater than 25% chance of developing this tumor.[12] Additionally, 83% of the patients with FAP and MF have a history of abdominal surgery, most commonly a total colectomy.[7] In this case, the patient was a young woman who underwent an appendectomy 2 years before this admission, but she had no history of FAP, and no polyps were visible via colonoscopy after the operation.

Most patients with MF are asymptomatic, while a few present with an abdominal mass, abdominal pain, fatigue, and vomiting. Severe complications, like intestinal obstruction, ischemia and perforation, hydronephrosis, ureteric fistula, and even aortic rupture, may result from MF infiltration into the adjacent organs.[13,14] In this case, a consecutive perforation from the ileum to the bottom of tumor was noted during the laparotomy. The ileum was perforated due to ischemia, which resulted from compression by the tumor, but not due to the direct infiltration by the tumor since no tumor cells were found in the ileal wall. The body of the mass was eroded and perforated by intestinal fluid, which leads to acute peritonitis. To our knowledge, this is an extremely rare clinical condition, and this is first case report of MF with perforation in the English literature.

Differential diagnoses of MF include GISTs, lymphoma, carcinoid tumor, fibrosarcoma, or inflammatory fibroid polyp. The 2 most commonly misdiagnosed conditions are MF that involve the bowel wall and GISTs, due to their different biological behavior and rarity.[12] However, the differentiation between these 2 tumor types is obligatory for patient prognosis and treatment.[15] It is difficult to make a definite diagnosis preoperatively when using only radiological tools, even with magnetic resonance imaging. Therefore, exploratory laparotomy is always the final treatment option. Intra-operative MF findings include grayish and grossly homogenous tissue with no metastases, whereas GISTs appear soft, fatty, and sometimes present hemorrhage, necrosis, and even direct invasion or metastasis. Thus, a large firm homogenous tumor without hemorrhagic and necrotic regions is an important indication of MF. However, the essential method for proper MF diagnosis is histological examination, especially immunohistochemical analysis. MF is characterized by a spatially homogenous proliferation of wavy spindle cells without atypia, that is associated with collagen in dilated vessels. The mitotic cell count is relatively low, with no evidence of necrosis and nuclear dedifferention.[16] If c-kit immunohistochemistry is positive or negative but the tumor is positive for *DOG1*, *PDGFRA*, or *CD34* genes, we can diagnose it as a GIST. However, if the tumor is negative for c-kit, DOG1, CD34, PDGFRA, and S100 but positive for β-catenin, we can confirm that the tumor is a MF.[17] CD117 antigen is also present in 75% of the patients with MF, which may result in misdiagnosis.[18]

The management of MF involves multiple modalities, including surgical resection, hormonal therapy, interferon therapy, and administration of non-steroidal anti-inflammatory drugs (NSAIDs) with chemotherapy. Radiotherapy plays a small role in MF treatment. Complete excision is the first-line treatment and the only indication for non-invading tumors, and R0 and R1 resection have shown the same outcomes.[19] The local recurrence rate may be to 40% to 70% following resection alone.[20] Due to this high local recurrence rate, others modalities play a central role in MF treatment, especially for patients who are unfit for surgery due to multiple comorbidities or when the tumor is unresectable.[21] Administration of indomethacin or sulindac (an NSAID) is the first logical treatment for unresectable tumors. If the tumor continues growing, tamoxifen with vinblastine and methotrexate is the best alternative. If there is no response to the usual chemotherapy drugs, we can use imatinib, a tyrosine-kinase inhibitor, that has been used in the treatment of multiple cancers and has shown good success.[22]

4. Conclusion

MF may manifest as a wide spectrum of clinical features, and the onset of MF as a perforation is extremely rare. Our case illustrates another possible cause of acute abdominal pain. Although rare, treating physicians should maintain a high suspicion index while managing patients with abdominal masses and pain. Once the diagnosis is established, surgical resection, hormonal therapy, interferon treatment, and NSAIDs with chemotherapy can be implemented separately or in combination. Close follow-up is essential because of the tumor's high incidence of local recurrence.

Author contributions

Jian Li was directly involved in the full implementation of this report and was a major contributor in the writing of this manuscript. Xu-Run and Hu-Deng Min were jointly involved in the surgical intervention literature analysis. All authors read and approved the final manuscript. Conceptualization: Jian Li. Methodology: Jian Li. Writing – original draft: Jian Li, Run Xu, Deng-Min Hu. Writing – review and editing: Jian Li, Run Xu, Deng-Min Hu.

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References

[1] Giuliani A, Demoro M, Ciardi A, et al. Mesenteric fibromatosis. Case report. J Exp Clin Cancer Res 2007;26:425–8.
[2] Bn A, Cd JK, Ps S, et al. Giant aggressive mesenteric fibromatosis – a case report. J Clin Diagn Res 2015;9:PD07–08.
[3] Polat C, Aktepe F, Turel S, et al. A giant mesenteric fibromatosis case presenting with mechanical intestinal obstruction and successfully resected with partial duodeno-jejunectomy and right hemicolectomy. Clinics 2010;65:110–3.
[4] Sinukumar S, Gomes RM, Kumar RK, et al. Sporadic giant mesenteric fibromatosis. Indian J Surg Oncol 2014;5:242–5.
[5] Guglielmi G, Cifaratti A, Scalzo G, et al. Imaging of superficial and deep fibromatosis. Radiol Med 2009;114:1292–307.
[6] Nicolas G, Kfoury T, Shimlati R, et al. Incidental finding and management of mesenteric fibromatosis. Am J Case Rep 2016;17:389–94.
[7] Mahajan A, Singh M, Varma A, et al. Mesenteric fibromatosis presenting as a diagnostic dilemma: a rare differential diagnosis of right iliac fossa mass in an eleven year old – a rare case report. Case Rep Surg 2013;2013:569578.
[8] Koh PK, Loi C, Cao X, et al. Mesenteric desmoid tumours in Singapore familial adenomatous polyposis patients: clinical course and genetic profile in a predominantly Chinese population. Dis Colon Rectum 2007;50:75–82.

[9] Burke AP, Sobin LH, Shekitka KM, et al. Intra-abdominal fibromatosis: a pathologic analysis of 130 tumors with comparison of clinical subgroups. Am J Surg Pathol 1990;14:335–41.
[10] Daryani NE, Momani AR, Azizi MR, et al. Mesenteric fibromatosis (desmoid tumour) presenting as a recurrent abdominal abscess: report of a rare case. Govaresh 2008;13:128–32.
[11] Yang CH, Sheen-Chen SM, Lu CC, et al. Computed tomographic presentation of mesenteric fibromatosis. Dig Dis Sci 2005;50:348–50.
[12] Stoidis CN, Spyropoulos BG, Misiakos EP, et al. Surgical treatment of giant mesenteric fibromatosis presenting as a gastrointestinal stromal tumor: a case report. J Med Case Rep 2010;4:314.
[13] Holubar S, Dwivedi AJ, O'C onnor J. Giant mesenteric fibromatosis presenting as small bowel obstruction. Am Surg 2006;72:427–9.
[14] Collins D, Myers E, Kavanagh D, et al. Mesenteric desmoid tumour causing ureteric obstruction. Int J Urol 2008;15:261–2.
[15] Azizi L, Balu M, Belkacem A, et al. MRI features of mesenteric desmoid tumors in familial adenomatous polyposis. Am J Roentgenol 2005;184: 1128–35.
[16] Rodriguez JA, Guarda LA, Rosai J. Mesenteric fibromatosiswith involvement of the gastrointestinal tract. A GIST simulator: a study of 25 cases. Am J Clin Pathol 2004;121:93–8.
[17] Yantiss RK, Spiro IJ, Compton CC, et al. Gastrointestinal stromal tumor versus intra-abdominal fibromatosis of the bowel wall: a clinically important differential diagnosis. Am J Surg Pathol 2000;24:947–57.
[18] Colombo P, Rahal D, Grizzi F, et al. Localized intraabdominal fibromatosis of the small bowel mimicking a gastrointestinal stromal tumor: a case report. World J Gastroenterol 2005;11:5226–8.
[19] Smith AJ, Lewis JJ, Merchant NB, et al. Surgical management of intra-abdominal desmoid tumours. Br J Surg 2000;87:608–13.
[20] Khorsand J, Karakousis CP. Desmoid tumors and their management. Am J Surg 1985;149:215–8.
[21] Janinis J, Patriki M, Vini L, et al. The pharmacological treatment of aggressive fibromatosis: a systematic review. Ann Oncol 2003;14:181–90.
[22] Bertagnolli MM, Morgan JA, Fletcher CD, et al. Multimodality treatment of mesenteric desmoid tumours. Eur J Cancer 2008;44:2404–10.