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Case Report

Colonic aggressive fibromatosis with perforation: a case report

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Abstract: Aggressive fibromatosis (AF) is a rare soft tissue tumor derived from mesenchymal tissue characterized by invasion into the surrounding muscle and soft tissue growth without distant metastasis. Although AF originates from the mesentery, omentum, and pelvis and has been described, primary AF originating from the colon has rarely been reported. In this case, an 18-year-old male patient with sudden left lower abdominal pain for 10 hours with nausea and vomiting was admitted to the hospital for an emergency medical examination. Imaging revealed a mass located near his sigmoid colon that was considered interstitial or malignant. The patient underwent urgent exploratory surgery, and perforation of the tumor was found. Histopathological examination showed that the tumor was colonic AF. Considering the patient's medical history of acute abdomen pain, we assume this to be a rare case of colonic AF with perforation.

Keywords: Aggressive fibromatosis, colon, perforation, therapy

Introduction

Aggressive fibromatosis (AF) is a rare soft-tissue tumor characterized by the proliferation of collagen fibers (including fibroblasts and myofibroblasts), with local progressive invasion of the surrounding soft tissue, without recurrence or transfer ability [1, 2]. Its biological features lie between fibroblastoma and fibrosarcoma. The malignant manifestations are often invasive and show postoperative recurrence, which accounts for 0.03% of all malignant tumors [3]. AF in the peritoneal cavity often occurs in the mesentery, posterior peritoneum, and pelvis, with little involvement of the colon. The etiology is still unclear and may be related to abnormal regulation of endocrine and connective tissue growth. The disease can be free of any symptoms in the early stage; with tumor growth, symptoms may occur due to compression; local infiltration, growth, and invasion of adjacent organs may lead to acute abdominal symptoms, for instance, perforation. In most cases, extended resection is the most effective treatment for invasive fibromatosis in the abdominal cavity. Adjuvant therapy, such as postoperative radiotherapy, can reduce the local recurrence rate. The diagnosis of the disease depends on histopathology and immunohistochemistry. Here, we describe an example of AF that originated from the colon, in which tumor invasive growth broke through the colon and acute abdominal symptoms of colonic perforation occurred. This case report provides us with a new perspective to identify acute abdominal symptoms and diagnose a colonic mass.

Case report

An 18-year-old male patient was admitted to our hospital due to "sustained left lower quadrant pain" for 10 hours, with nausea and vomiting. The patient had had a "right inguinal hernia" 10 years prior, and he denied any family history of tumors. Physical examination: body

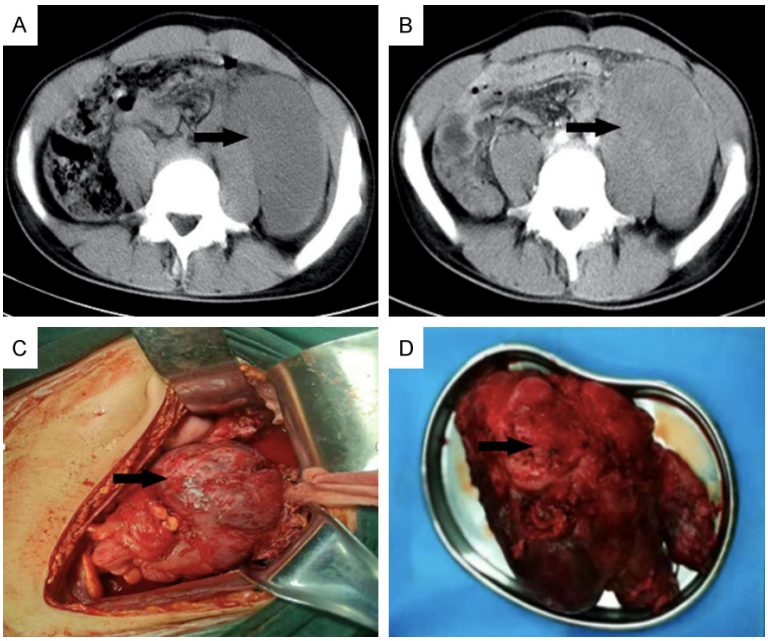

Figure 1. Computed tomography: (A) and (B) Show an oval decreased-density lesion that was detected in the left descending colon; the boundary was clear, and the lesion was progressively enhanced. Surgery photos: (C) There was a mass (approximately 12*8 cm) in the left lower abdomen, with a 1*1 cm ulcer and adherence to the colon; (D) Removal of the tumor mass.

temperature 38.7°C, pulse 92 beats/min, breathing 30 beats/min, blood pressure 126/64 mmHg, flat abdomen, no intestinal or peristaltic waves, liver and spleen palpated under the ribs, total abdominal tenderness with positive rebound tenderness, abdominal muscle tension, plate-shaped abdomen, negative referred pain, negative renal angle pain, weak bowel sounds, approximately 2 times/min. Laboratory examination: routine blood work: WBC: 20.81*10^9/l, NEUT%: 96.20%; biochemical: D-BIL: 10.50 μmol/l, Abl: 32.50 g/l; coagulation: mild abnormality. Abdominal B-ultrasound: An 8*12 cm hypoechoic sound was detected in the lower left abdomen, and the boundary was clearly identifiable, diagnosed as a left lower abdomen space-occupying lesion (with characteristics to be determined). An abdominal CT plain scan revealed an oval decreased-density lesion in the left descending colon, with a size of approximately 5.5*10 cm. The boundary was clear: the lesion was progressively enhanced, and a fluid density could be seen in the rectovesical pouch, which was diagnosed as a large space-occupying lesion in the descending colon of the left lower abdomen, considered to be a lymphoma (**Figure 1A**, **1B**). The patient was currently in an emergency condition. We explained the test results to the patient and his family, and they agreed to actively prepare for surgery. We performed a laparotomy in the emergency department. During the intraoperative exploration, there was a large amount of purulent fluid (approximately 300 mL) in the abdominal cavity; the entire colon, small intestine and intestinal wall had pus attached, and there was a mass (approximately 12*8 cm) in the left lower abdomen, with 1*1 cm ulcer and adhered to the colon. The surrounding colon was edematous (**Figure 1C**), which led to enlargement of the colonic mass. The intraoperative diagnosis was colonic mass ulceration with diffuse peritonitis. Postoperative pathology showed a section of intestinal tissue (approximately 17 cm) that could be seen by the naked eye, with circumferences of both ends at 4 cm and 3.5 cm, respectively (**Figure 1D**). No tumor signs was observed at the incision edge of intestine, however, the cancer tissue (approximately 12*6 cm) was discovered at 4.5 cm from the incisal margin. The tumor occupied the whole layer of the intestinal wall with grayish white matter on the cut surface; five lymph nodes were visible. Immunohistochemistry showed that the cells were positive for Vimentin and β-catenin and negative for CD34, S-100, CD117, desmin and Dog-1 (**Figure 2D-F**). These findings combined with pathology (**Figure 2A-C**) and immunohistochemistry results revealed that the patient had colonic fibromatosis, which aggressively invaded the intestinal muscle layer; the mucosal tissue at both ends showed chronic inflammation; lymph node tissue showed chronic reactive proliferative lymphadenitis. One week after the operation, the relevant biochemical tests and imaging revealed no obvious abnormalities. The patient was discharged from the hospital. One week after discharge, the patient received radiotherapy in our hospital. After the end of the radiotherapy, abdominal CT and laboratory examina-

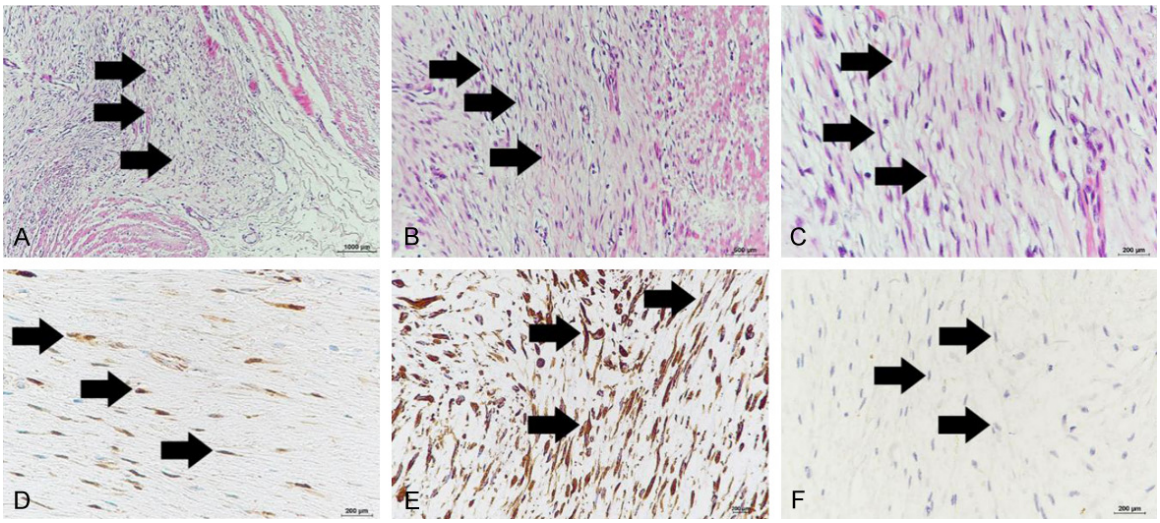

Figure 2. Microscopic examination showing: (A) Tumor tissue involving the muscular layer of the colon wall (H&E×100); (B) The junction of the tumor tissue with the muscular layer of the colon wall (H&E×200); (C) Spindle fibroblast cells arranged in parallel bundles and surrounded by abundant fibrillary collagen (H&E×400). Immunohistochemistry findings. (D) Cell positivity for the β-catenin antibody (×400); (E) tumor cell positivity for the vimentin antibody (×400); (F) Cell negativity for CD34 (×400).

tions were reviewed at 1 month and 3 months. No recurrence was found. At present, the patient's quality of life is good, with no abdominal pain, abdominal distension, or other symptoms.

Discussion

In 1838, Muller designated aggressive fibromatosis (AF) as a desmoid tumor, which is a hard fibroma with the tendency to invade surrounding muscle and soft tissue and to recur after surgery. This tumor is prone to local recurrence but exhibits no distant metastasis [1]. Its biological behavior lies between that of fibroblastoma and fibrosarcoma. AF can occur at any age, with a high incidence in children and youth and no significant gender differences. AF mostly occurs as a single tumor. At present, the lesion location of AF can be divided into extra-abdominal (50%-60%), on the abdominal wall (approximately 25%) and intra-abdominal (approximately 15%). The intra-abdominal type often occurs in the mesentery, posterior peritoneum and pelvic cavity, with a round-like mass that can surround the internal organs, e.g., intestines. It is often associated with FAP and Gardner syndrome. Primary invasive fibromatosis of the colon is even more rare, and there has not been any relevant report in other publications. The clinical symptoms are mainly related to the size and location of the tumor, the duration of the disease and the tumor growth rate. The maximum diameter can be up to more than 10 cm. This patient had experienced problems with defecation for several years, with pain in the left lower abdomen and other discomfort. After ingesting cathartics and adjusting his diet, his symptoms were improved, and he consequently did not pay attention to them.

The patient presented with acute left lower abdomen pain, and his whole body was tender, rebound tenderness was positive, his abdominal muscles were tense, and his bowel sounds were weak. Laboratory tests indicated infection and an inflammatory response. Based on this finding, we considered perforation, intestinal torsion leading to intestinal necrosis, and incarceration of a left inguinal hernia. However, the whole abdominal CT showed a large lesion in the lower left descending colon. The mass surrounded the colon, but not the intestinal lumen, and the surrounding boundary was clear (**Figure 1A**, **1B**). Excluding the above diseases, combined with imaging results, we assumed that the case could involve a colon tumor or GIST (Gastrointestinal Stromal Tumors). In this case, CT showed that the mass was located in the descending colon, next to the sigmoid colon and the surrounding small intestine. In addition, the mass was large and grew rapidly, indi-

cating the possibility of malignancy and GIST. Colon cancer is the most common malignant tumor of the colon, which occurs at the junction of the rectum and the sigmoid colon. Most patients are over 40 years old, while young people rarely have this condition. The CT manifestations are mainly intestinal wall thickening, an intraluminal mass, intestinal stenosis and abnormal intestinal wall enhancement. In this case, the patient was 18 years old without family cancer history. His clinical symptoms were only difficulty in defecation, abdominal distension and no blood in the stool. Although the clinical symptoms were similar, the imaging examination results did not correspond to colon cancer. In addition, there was no abnormality in the postoperative tumor index, which may also rule out colonic malignant tumors. GIST is also the most common mesenchymal tumor in the gastrointestinal tract. It can occur in any part of the gastrointestinal tract and lacks specificity in clinical symptoms. Imaging results often show a mass with a round border. However, there are some differences between AF and GIST. The main pathological manifestations of AF are spindle-shaped fibroblasts and myofibroblasts arranged in parallel bundles and invading into surrounding muscle or soft tissue (**Figure 2A**, **2C**). Immunohistochemical examination usually shows positive for β-catenin and vimentin. GIST is positive for CD117 and CD34 and consists of epithelial-like cells and spindle cells. Pathological examinations are usually able to identify the type of tumor. Due to the emergent condition of the patient at the time of admission, other tests and imaging examinations could not be performed immediately, and therefore, the diagnosis was based on the intraoperative examination and postoperative pathological analysis. Intraoperative exploration revealed a colonic mass originating in the colon, and an ulcer was seen above the mass. The surrounding area occupied the entire colon, and the edges were smooth and clear. This also explained why the patient had difficulty in defecation. Combined with the pathology and immunohistochemistry results, we assumed colon AF with perforation. At present, gastrointestinal fibromatosis is rare, and colonic fibromatosis is extremely rare. There is no clear diagnosis or treatment guidelines. The preferred imaging examination for AF is MRI+DWI. The scope of invasion can be preoperatively assessed (to see whether it has aggressed peripheral blood vessels and other organs) to make a precise surgery plan. Preoperative ultrasound or CT-guided fine-needle aspiration cytology (FNAC) or immunohistochemical staining (e.g., β-catenin) can be used as a preoperative diagnosis [4]. Because the patient was admitted to the hospital with acute abdominal symptoms, MRI was not performed. However, postoperative immunohistochemical results showed that the cells were positive for Vimentin and β-catenin (**Figure 2D-F**); the other indexes were negative, which was consistent with the features of AF. The most advocated treatment for AF is surgical resection. Because of its non-enveloped and invasive growth characteristics, it is difficult to determine the tumor boundary. Most scholars recommend cutting at 2-3 cm from the tumor and removing the surrounding tissue. The intraoperative macroscopic view of the incision should have no hard tissue, and pathology should double-check the negative margin (R0) [5]. Regarding this case, colon mass enlargement resection was performed, and the tumor was approximately 4 cm away from the margin. No tumor tissue was observed by the naked eye or under the microscope. Mullen et al. [6] performed a retrospective study of 177 patients and showed that marginal status was an independent risk factor for AF recurrence. A meta-analysis summed up the relationship between the surgical resection margin and postoperative recurrence in 698 AF patients in 13 clinical centers. Even after R0 resection, the local recurrence rate was 12%-27%, and patients with gross resection margins would be highly likely to present recurrence after surgery, with a local recurrence rate of approximately 100%. The recurrence rate of positive R1 under the microscope was 42%-68%, and the overall recurrence rate was 40%. Most of these tumors are recurrent in situ [7]. In this case, both a macroscopic and a postoperative pathological examination detected no tumor tissue in the surgical margin, and the radical resection criteria were achieved. Moreover, postoperative radiotherapy can significantly reduce the local recurrence rate, but preoperative radiotherapy does not improve the rate of negative surgical margins [7, 8]. Studies have shown that postoperative radiotherapy of R0 patients yields a 17% improvement in the local control rate compared with surgery alone, which was 40% for R1 and 78% for R2. The total local control rate of radiother-

apy was 78%-93% [7]. To reduce postoperative recurrence, the patient underwent radiation therapy 2 weeks after surgery. Although the current expansion of the resection may theoretically be cured, there is still a high rate of postoperative recurrence. Therefore, adjuvant chemotherapy such as chemoradiotherapy, nonsteroidal anti-inflammatory drugs (NSAIDs), selective estrogen receptor modulators (SERMs), and tyrosine kinase inhibitors (TKIs) can be used to reduce the postoperative recurrence rate.

Conclusion

In conclusion, because AF disease is rare, colonic perforation is extremely rare, and the current research is mostly composed of retrospective and pilot studies. There is a lack of perspective and multicenter studies. Surgical treatment is still the main treatment for AF; meanwhile, R0 resection should be performed as precisely as possible. To reduce the recurrence rate after surgery, adjuvant radiotherapy is the first choice. If necessary, this treatment should be combined with NSAIDs, antihormone drugs, chemotherapy, and other personalized treatments. We hope this case report provides a new perspective to identify acute abdominal symptoms and diagnose colonic masses.

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Disclosure of conflict of interest

None.

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