

Intra-abdominal Desmoid-Type Fibromatosis Mimicking Diverticulitis With Abscess: A Case Report

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Abstract. *Background: Intra-abdominal desmoid-type fibromatosis (DF) rarely necessitates emergency surgery. However, the condition is difficult to diagnose preoperatively and can become life-threatening if left untreated. Case Report: A 46-year-old man complained of fever and right lower quadrant pain. In computed tomography, the mesenteric side of the ascending colon demonstrated air and fluid collections, suggesting diverticulitis with abscess. After 2 weeks of conservative treatment with fasting, the patient started to consume food; nonetheless, fever returned. Colonoscopy and contrast enema detected a fistula extending from the ascending colon to the abscess, with no surrounding lesions. Surgery was then performed because the abscess was refractory. During laparotomy, the scar tissue of the abscess was found to be attached to the lateral wall of the ascending colon. Hence, right colectomy combined with abscess resection was performed. Histopathological findings revealed DF in the mesentery. Conclusion: Although rare, DF should be included in the preoperative differential diagnosis of intra-abdominal abscesses.*

Desmoid-type fibromatosis (DF) is a rare monoclonal fibroblastic proliferation characterized by locally aggressive growth and frequent recurrence but without metastasis (1, 2). The incidence of DF is approximately 5-6 cases per million annually, with a peak age of 30-40 years (1). Most DF cases

occur sporadically but approximately 5-10% arise from familial adenomatous polyposis caused by germline mutation of the adenomatous polyposis coli gene (2). DF can be classified into extra-abdominal (trunk/extremity), abdominal wall, and intra-abdominal (bowel and mesentery) according to its location (3). Most intra-abdominal cases are asymptomatic and can be followed-up without surgery. However, intestinal obstruction, perforation, and abscess formation may occur because of enlarged desmoid tumours, thereby requiring surgical intervention (2, 4). Moreover, approximately one-fifth of patients die owing to complications from intra-abdominal DF. The local aggressive growth of DF can cause infiltration of the surrounding structures and hence be life-threatening (6). Therefore, complete DF resection is recommended (1, 5). Herein, we report a rare case of intra-abdominal DF that appeared to be diverticulitis with abscess. Preoperative diagnosis was difficult, and a precise diagnosis was possible only postoperatively. In this case report, we discuss the importance of considering DF, as well as diverticulitis, whenever pericolic abscess is detected to ensure appropriate decision-making and surveillance. We obtained oral informed consent from the patient for the publication of this case report and accompanying images.

Case Report

A 46-year-old man with no significant medical or family history was referred to our Department with complaints of fever and right lower quadrant abdominal pain of 3 days' duration. During physical examination, abdominal rebound tenderness and abdominal guarding in the right lower quadrant were noted. Laboratory tests revealed a white blood cell count of 13,700/mm² and a C-reactive protein (CRP) level of 30.81 mg/dl. Abdominal computed tomography (CT) showed a 4.5×5.2×5.4-cm protrusion of air and fluid collection in the mesenteric side of the ascending colon, indicating diverticulitis with abscess (Figure 1A and B).

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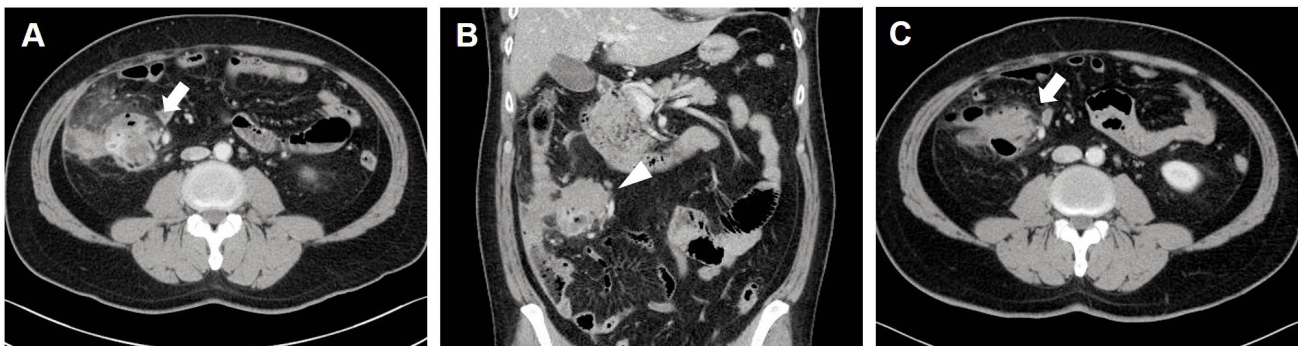


Figure 1. Abdominal computed tomography (CT) findings. Horizontal (A) and coronal (B) views by CT before conservative treatment. Abdominal CT showed air and fluid collection (arrow) in the mesenteric side of the ascending colon (arrowhead). Three weeks later, CT (horizontal view; C) showed a reduction in the abscess size, but the abscess and fistula formation in the ascending colon remained (arrow).

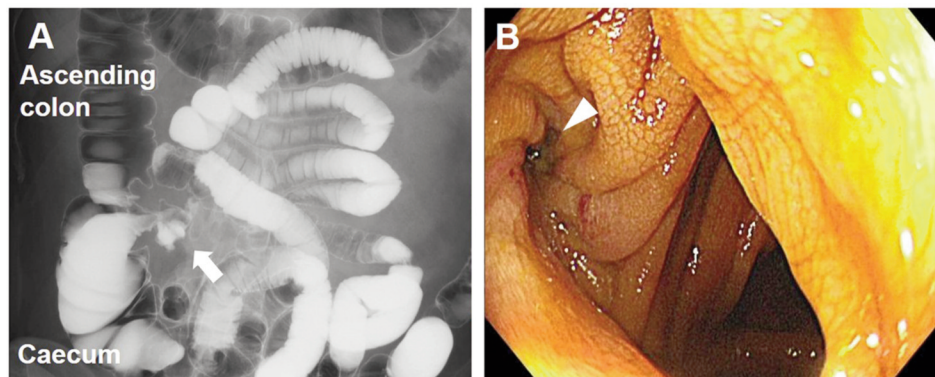


Figure 2. Contrast enema and colonoscopy findings. A: Contrast enema showed the abscess and fistula formation in the ascending colon (arrow). B: Colonoscopy revealed a fistula in the ascending colon (arrowhead).

Peritonitis was localized, the abscess was not large, but safe percutaneous drainage was impossible. Therefore, a conservative treatment consisting of fasting and antibiotics was provided. After 2 weeks, the CRP level decreased to 1.74 mg/dl; however, fever and CRP elevation returned when the patient started consuming food. Fasting was restarted, and after another week, CT revealed a reduction in the abscess (Figure 1C). Additionally, contrast enema detected a fistula extending from the ascending colon to the abscess (Figure 2A). Colonoscopy also revealed a fistula in the ascending colon without surrounding lesions as the possible cause of the disease (Figure 2B). Thus, surgical intervention was decided. During laparotomy, the scar tissue related to the abscess was found to be attached to the lateral wall of the ascending colon, and had spread to the retroperitoneum. Hence, right colectomy with abscess resection was performed. No evident neoplastic lesions were observed macroscopically (Figure 3). Histopathologically, we found proliferation of spindle-shaped cells with collagenous stroma in the mesentery (Figure 4A and B). Immunohistochemical

study showed that the tumour cells were positive for β -catenin (Figure 4C). Hence, the final pathological diagnosis was intra-abdominal DF that developed at the mesentery and diffusely infiltrated the ascending colon. A portion of the DF had infiltrated the *muscularis propria* of the ascending colon and had formed a fistula. However, the boundary between the tumour and the normal area was unclear, and no evident positive margin was observed. No recurrence was observed 6 months postoperatively.

Discussion

DF is extremely rare and almost always asymptomatic. However, it is difficult to diagnose the condition preoperatively, and can be life-threatening. Thus, care should be taken when an intra-abdominal abscess is detected. The patient in our case highlights important clinical implications. Firstly, DF does not always present as a neoplastic lesion; therefore, preoperative diagnosis can be difficult. The most commonly employed imaging modality for DF, especially

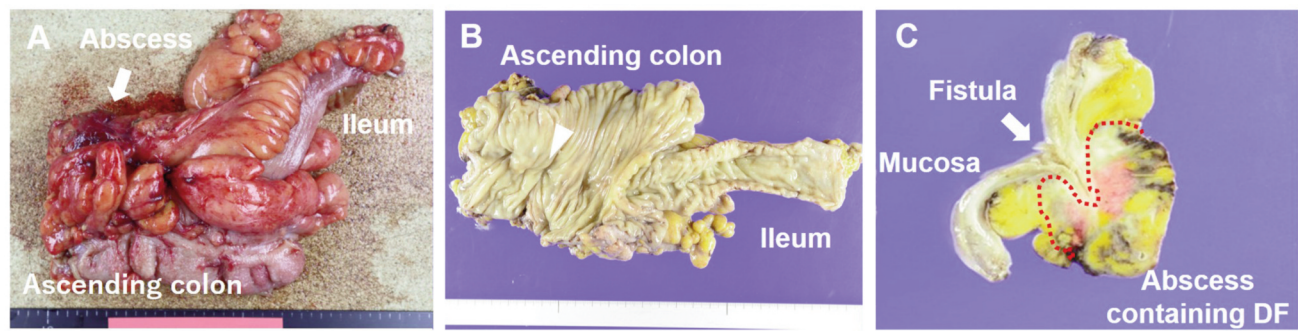


Figure 3. Macroscopic findings in the resected specimen. A: View of the serous side. B: View of the mucosal side. The scarred abscess (arrow) was attached to the lateral wall of the ascending colon in which the fistula was located (arrowhead). No evident diverticulum was observed. C: The boundaries of the tumour in the resected specimens were not clear (the dotted line indicates an example). The fistula between DF and the ascending colon was detected (arrow).

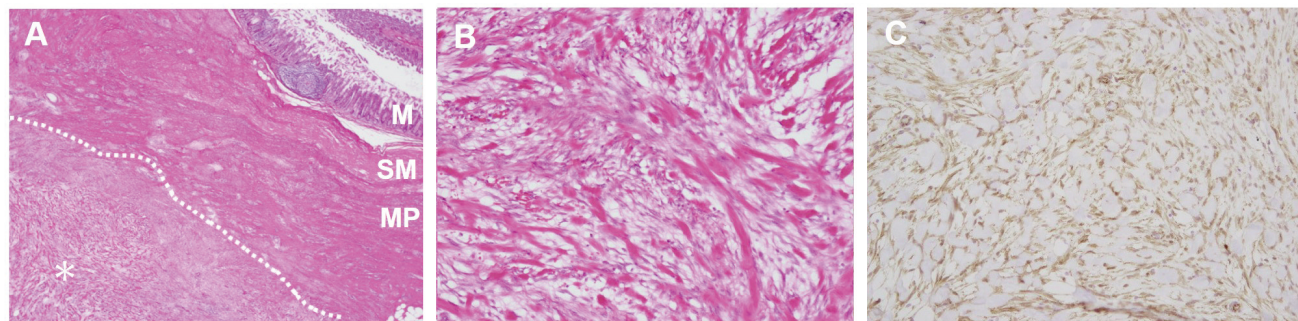


Figure 4. Histological and immunohistological features of the tumour. A: The boundary (dotted line) between the tumour (asterisk) and the ascending colon was unclear ($\times 40$). M: Mucosa; SM: submucosa; MP: muscularis propria. B: Haematoxylin and eosin staining showed the proliferation of spindle-shaped cells in collagenous stroma ($\times 100$). C: Immunohistological staining for β -catenin showed the focal staining of cell nuclei ($\times 100$).

intra-abdominal DF, is CT (7). In cases involving extra-abdominal DF and its recurrence, magnetic resonance imaging provides a better resolution of the soft tissue and is therefore recommended for evaluation (4, 7). However, positron-emission tomography/CT is insufficient for DF diagnosis and management because DF is typically not very metabolically active and often demonstrates standardized uptake values of ≤ 4.8 (4, 7). On CT images, DF is typically seen as a soft-tissue mass of variable attenuation and enhancement, with radiating spicules extending into the adjacent mesenteric fat (4, 7). Differential diagnoses of DF include gastrointestinal stromal tumour, lymphoma, soft-tissue sarcoma, and mesenteric metastases; no radiographic characteristics can reliably distinguish DF from other tumour types (4, 8). In some cases, including ours, it was not possible to diagnose as DF preoperatively because of complications (Table I) (9-11). Furthermore, our case and a previous one involving cyst formation (11) did not reveal any neoplastic lesions intraoperatively and were only diagnosed on the basis of postoperative pathology. In our case, contrast-enhanced CT

detected the abscess with capsular ring enhancement. The centre had a relatively low attenuation, thereby indicating the presence of a necrotic component. Meanwhile, colonoscopy revealed a fistula in the ascending colon, but no lesions were detected at the mucosal surface. Therefore, the patient was diagnosed with abscess formation due to diverticular penetration. However, in contrast-enhanced CT, DF might have been present in low-density areas. Although DF rarely forms an abscess and cyst, it should still be considered as a preoperative differential diagnosis.

Secondly, refractory intra-abdominal abscesses may indicate DF. Most DF cases are asymptomatic and have chronic progression, resulting in a solid abdominal mass which might be associated with abdominal pain. Complications of DF result from its locally aggressive nature, causing compression and invasion of the adjacent structures. Although intra-abdominal DF may cause intestinal obstruction, bleeding, and perforation, it rarely forms an abscess (4). In a previous report, six out of eight DF cases with abscess formation were refractory despite drainage and

Table I. Reported cases of desmoid-type fibromatosis that were difficult to diagnose preoperatively due to complication.

| Reference | Year | Years | Gender | FAP related | Symptoms | Imaging findings | Location | Preoperative diagnosis | Treatment | Operative findings | Tumour size (cm) | Prognosis |
|--------------|------|-------|--------|-------------|---|------------------------------------|---|-------------------------------|--|---|------------------|------------------------------|
| 9 | 2006 | 32 | F | Non | Fever, left upper quadrant pain | Cyst | Between the spleen, stomach, and left large bowel | NA | Left hemicolectomy | A tumor mass in the intestinal wall of the descending colon | NA | 2 Years without recurrence |
| 10 | 2012 | 22 | F | Non | Left upper quadrant pain, vomiting, hemorrhagic shock | Fluid collections | Around the spleen | NA | Splenectomy | A tumor strangulating the splenic artery and branches of the artery to the pancreas | 1.5 | NA |
| 11 | 2015 | 13 | M | Non | Left upper quadrant pain | Cyst (10 cm) | Adjacent to the pancreatic tail | Splenic cyst | Distal splenopancreaticectomy with en bloc resection of the left colonic flexure | A cystic mass invading the pancreatic tail and left colonic flexure, spleen | NA | 19 Months without recurrence |
| Current case | 2020 | 46 | M | Non | Fever and right lower quadrant pain | Fluid collections and air (4.5 cm) | Mesenteric side of the ascending colon | Diverticulitis with abscesses | Right colectomy | A scarred abscess attached to the lateral wall of the ascending colon | NA | 6 Months without recurrence |

NA: Not available; F: female; M: male; FAP: familial adenomatous polyposis.

antibiotic administration and were treated surgically, as in our case (12). Intra-abdominal abscesses that are refractory despite conservative treatment are possibly caused by DF.

Optimal management of DF requires a multidisciplinary approach tailored to the needs of individual patients. Stable, asymptomatic primary or recurrent DF can be followed-up without surgical intervention because conservative management and surgery yield comparable outcomes (1). Additionally, recurrence after surgical resection is common (15-30%) in intra-abdominal DF, and recurrent DF is often more aggressive than the initial DF (2). Regarding a failed surveillance approach for symptomatic desmoids, especially those with mass effect on critical structures, surgery is necessary. Any residue of aggressive DF poses a high risk of regrowth, and negative margins are important (13). Therefore, the possibility of DF must be considered prior to surgery because complete resection is required in such cases.

In conclusion, intra-abdominal DF with a fistula in the ascending colon and abscess formation is difficult to diagnose preoperatively. Although the preoperative diagnosis of DF with complications such as abscess is challenging, this possibility needs to be considered in patients with intra-abdominal abscesses, especially those that are refractory to conservative treatment.

Conflicts of Interest

The Authors have no conflicts of interest to declare.

Authors' Contributions

SO researched the literature, and wrote the article. SI provided direct patient care, researched the literature and edited the article. All Authors read and approved the final article.

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