Case Reports in Oncology

Case Rep Oncol 2025;18:436–442
DOI: 10.1159/000544814
Received: November 26, 2024
Accepted: February 11, 2025
Published online: February 26, 2025

Case Report

Hemorrhagic Shock Caused by an Aggressive Fibromatosis in the Gastropancreatic Region: A Case Report

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Keywords
Aggressive fibromatosis · Intra-abdominal aggressive fibromatosis · Gastrointestinal oncology · Pathology · Surgery

Abstract
Introduction: Aggressive fibromatosis (AF), also known as desmoid tumor, is a rare condition characterized by the proliferation of monoclonal fibroblasts, primarily originating from connective tissue. Despite being histologically benign, AF exhibits malignant-like behavior, including local invasion and a high recurrence rate. AF can be classified based on its location into extra-abdominal, abdominal wall, and intra-abdominal types. Intra-abdominal AF (IAF), though least common, has the worst prognosis and highest mortality rate. Traditionally, complete surgical resection (R0) was the preferred treatment, but recent strategies favor conservative management, especially for asymptomatic patients. Emergency surgery is reserved for complications like bleeding, perforation, or obstruction. ***Case Presentation:*** This report details a rare case of IAF in the retroperitoneum of a 39-year-old woman presenting with hemorrhagic shock. Emergency surgery, including partial distal pancreatectomy and partial gastrectomy, was performed. The tumor was β-catenin positive, confirming the diagnosis of IAF. Postoperatively, the patient recovered well and showed no recurrence after 2 years without additional therapy. ***Conclusion:*** In summary, IAF presents significant diagnostic and therapeutic challenges. Effective management relies on a multidisciplinary approach, combining various diagnostic tools to improve early detection and patient outcomes. Continued research is essential to understand the pathogenesis of AF and to develop less invasive treatment options.

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DOI: 10.1159/000544814

Introduction

Aggressive fibromatosis (AF), also known as desmoid tumor or fibromatosis, is formed by the proliferation of monoclonal fibroblasts, primarily originating from the connective tissue of the fascia or aponeurosis. Although histologically benign, its biological behavior exhibits characteristics of malignant tumors, such as invading surrounding tissues, local infiltration, and a high recurrence rate. AF can be classified based on its location into extra-abdominal, abdominal wall, and intra-abdominal types. Among these, intra-abdominal AF (IAF) has the lowest incidence but the worst prognosis and highest mortality rate. Due to the low incidence of IAF, clinical experience is limited, making preoperative diagnosis challenging and often confused with other soft tissue tumors.

Traditionally, R0 surgical resection was considered the preferred treatment. However, in recent years, treatment strategies have shifted toward more conservative management. Most experts recommend close observation for asymptomatic patients and prioritize nonsurgical treatment strategies when positive margins are expected.

Here, we report a rare case of IAF arising from the retroperitoneum in the gastropancreatic region. The patient presented with hemorrhagic shock due to tumor bleeding and underwent emergency surgery, including partial distal pancreatectomy and partial gastrectomy. We share this clinical experience to improve strategies for handling similar cases in emergency situations.

Case Report

A 39-year-old woman presented to our urology clinic with upper abdominal pain. She had no significant medical history. An abdominal plain CT scan showed (1) a small stone in the right kidney; (2) a high-density area in the mid-to-distal segment of the splenic artery, measuring 17 mm, suggesting a splenic artery aneurysm (Fig. 1). The clinic initially attributed the pain to the kidney stone and prescribed medication. Later that afternoon, the patient's pain worsened, and an enhanced abdominal CT scan revealed significant fluid accumulation around the stomach, liver, spleen, and pelvis, indicating a ruptured splenic artery aneurysm (Fig. 2). She was transferred to the emergency room, where her hemoglobin was 96 g/L, and abdominal paracentesis extracted non-coagulating blood. Emergency surgery was performed under general anesthesia. During the operation, approximately 1,000 mL of blood was found in the abdominal cavity. A mass was palpable in the region of the pancreas behind the stomach, with surface rupture and significant blood clots. Clearing the clots revealed arterial bleeding. The tumor had unclear boundaries with the pancreas and was closely attached to the stomach and transverse colon. A distal pancreatectomy, splenectomy, and partial gastrectomy were performed.

On gross section, the tumor was irregularly shaped, measuring 30 × 20 × 25 mm, with a white and dark red cut surface. Histological examination showed abnormal spindle cell proliferation with slightly thickened fibrous structures and no significant atypia. Microscopically, the tumor was attached to the pancreas, splenic artery, and splenic vein, without invading the spleen or stomach. Immunohistochemistry showed β-catenin positive, SMA positive, S-100 negative, and Ki-67 positive (about 1%), diagnosing it as an intra-abdominal aggressive fibromatosis (IAF) originating from the retroperitoneum (Fig. 3).

The patient recovered well postoperatively and was discharged on the 9th day. She did not undergo chemotherapy or targeted therapy and was followed up with observation. Two years post-surgery, there has been no tumor recurrence (Fig. 4).

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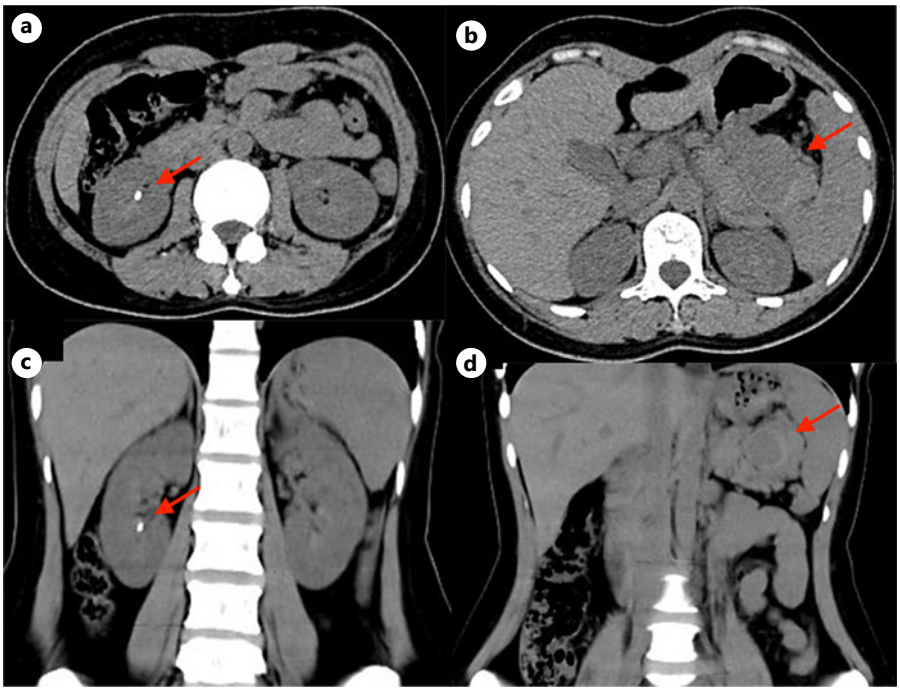

Fig. 1. Abdominal plain CT findings: abdominal axial (**a**) and coronal (**c**) CT revealed a small stone in the right kidney (red arrow); abdominal axial (**b**) and coronal (**d**) CT revealed well-defined round mass in the gastropancreatic region measuring 17 mm in diameter, suggesting a splenic artery aneurysm (red arrow).

Discussion

AF, also known as desmoid tumors, is a rare benign tumor that arises from the connective tissue cells in the body (incidence 5–6 cases per 1 million of the population per annum) [1]. These tumors can occur anywhere in the body but are most commonly found in the abdominal wall, shoulder, and thigh. Although histologically benign, its biological behavior exhibits characteristics of malignant tumors, such as invading surrounding tissues, local infiltration, and a high recurrence rate. AF can be classified based on its location into extra-abdominal, abdominal wall, and intra-abdominal types. Among these, IAF has the lowest incidence but the worst prognosis and highest mortality rate with a higher recurrence rate after treatment [2].

AF are mainly classified into sporadic and hereditary types. The exact cause of AF is not fully understood. Over 90% of these cases are sporadic and are associated with mutations in the β-catenin gene (CTNNB1). A smaller number of cases occur in patients with familial adenomatous polyposis, which is linked to germline mutations in the APC gene. Mutations in the β-catenin oncogene (CTNNB1) or germline mutations in the APC gene in familial adenomatous polyposis patients lead to activation of the Wnt signaling pathway [3]. Imaging tests such as CT scans, MRI scans, and ultrasound can be used to visualize the tumor and determine its location and size [4]. Typical features are a round or fusiform shape, unclear boundaries, uniform signals, uneven enhancement, “tree root” or “claw” infiltration, and invasion of the neurovascular bundles. Although imaging assessment could help characterization, a definitive diagnosis requires histopathological confirmation. Pathology is the gold standard for the diagnosis of IAF. IAF typically presents as a hard mass, and the cut surface appears white and glossy. Microscopically, the lesions are poorly defined and often infiltrate surrounding tissues. The tumor is mainly composed of spindle cells and collagen fiber bundles arranged in an interwoven pattern. In terms of immunophenotype, most IAF cases have CTNNB1 gene

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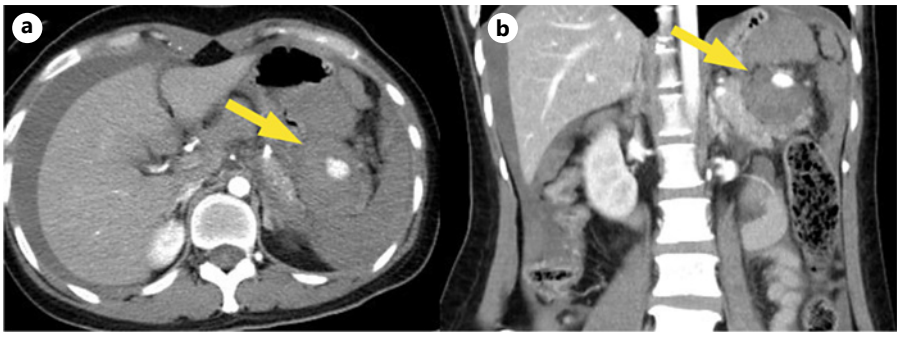

Fig. 2. a, **b** Contrast-enhanced abdominal CT scan revealed significant fluid accumulation around the stomach, liver, spleen, and pelvis, indicating a ruptured splenic artery aneurysm (yellow arrow).

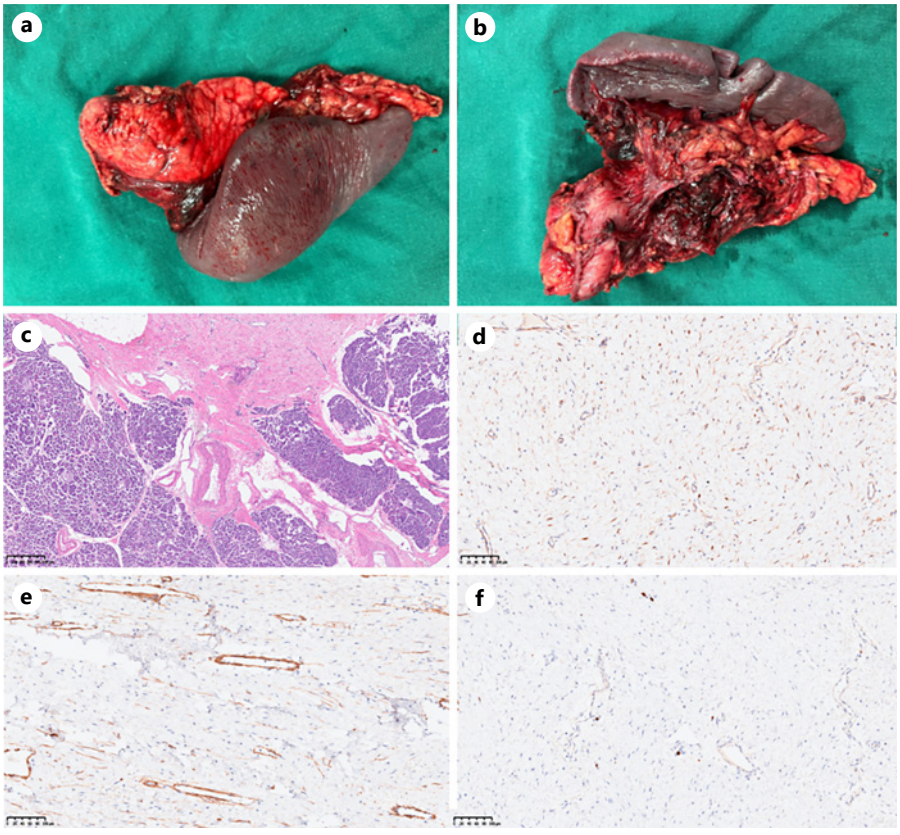

Fig. 3. Contrast-enhanced abdominal CT scan revealed the tumor appeared irregular in shape, measuring 30 × 20 × 25 mm. **a**, **b** The cut surface showed a mixture of white and dark red, with surface rupture and blood clots. **c** Histological examination showed abnormal spindle cell proliferation with slightly thickened fibrous structures. **d** Immunohistochemical staining with nuclear positivity for β-catenin (×200). **e** Partial positive expression of SMA (×200). **f** Ki-67 expression was 1%+ (×200).

mutations, leading to abnormal nuclear expression of β-catenin protein. Therefore, nuclear expression of β-catenin is an important diagnostic marker for IAF, with high sensitivity [5].

In terms of drug therapy over the past decade, tyrosine kinase inhibitors have been a major focus of research, aiming to shrink tumors, reduce recurrence, and control tumor progression. The three most studied targeted drugs for AF are imatinib, sorafenib, and

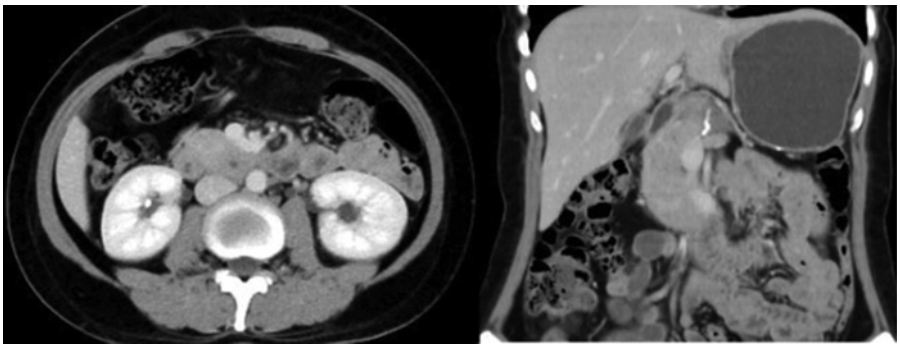

Fig. 4. Contrast-enhanced CT findings 2 years after surgery. There has been no tumor recurrence.

pazopanib. Various studies have shown these drugs to have good therapeutic effects on tumors [6–8]. These new therapies may offer more effective treatment options for IAF patients. More clinical trials are needed in the future to evaluate the efficacy and safety of these therapies, with the goal of improving long-term outcomes for patients.

In the past, complete surgical resection (R0) was considered crucial for treatment, and postoperative adjuvant therapies like radiotherapy or medication were thought to help reduce recurrence risk. Recently, treatment strategies have shifted toward more conservative management. One of the largest studies comparing initial surgery to initial observation, reported by Penel et al. [9], showed no difference in event-free survival (53% versus 58%; p = 0.415) and long-term disease control between patients undergoing surgery and those managed conservatively. Currently, most experts recommend close observation for asymptomatic patients and prefer nonsurgical treatment strategies in cases with expected positive margins, especially when extensive resection would lead to significant loss of organ function or appearance. Positive margins are acceptable in such scenarios.

For surgery, if the tumor causes complications like bleeding, perforation, or obstruction, emergency surgery is required. The goal is to resolve these complications. Even if intraoperative or postoperative pathology indicates positive margins, postoperative radiotherapy or further extensive resection is not recommended. Conservative observation is advised, with treatment only if recurrence is confirmed and there is continuous growth.

In this case, the patient underwent emergency surgery due to massive bleeding caused by the tumor. During surgery, the tumor was indistinguishable from the transverse colon. To avoid a colostomy, the tumor was separated from the transverse colon. The patient was closely monitored postoperatively, and 2 years later, a follow-up CT scan showed no signs of tumor recurrence. From this case, we have learned that IAF has a low incidence but can present with severe complications such as bleeding, obstruction, or perforation, requiring urgent surgical intervention. Early diagnosis is challenging due to the tumor's vague clinical presentation and the difficulty in distinguishing IAF from other soft tissue tumors. Imaging studies and histopathological examination are essential for diagnosis. Conservative management with close observation is recommended for asymptomatic IAF patients, while surgery should be reserved for cases with complications or symptomatic tumors. Positive surgical margins may be acceptable in certain cases, with follow-up monitoring.

Conclusions

IAF is a rare and complex disease that can present significant challenges in diagnosis and treatment. Due to a lack of specific symptoms, diagnosis typically relies on a detailed medical history, imaging studies, and immunohistochemical analysis when necessary. For suspected

cases, a combination of diagnostic methods should be used to improve the accuracy and timeliness of early diagnosis, thereby enhancing patient outcomes. Further research is needed to better understand the underlying causes of desmoid-type fibromatosis and to develop more effective and less invasive treatments for this challenging disease.

Statement of Ethics

Ethical approval is not required for this case report in accordance with national guidelines. Written informed consent was obtained from patient for publication of the details of their medical case and any accompanying images (available upon request). The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000544814).

Conflict of Interest Statement

Yang Xiufang and Xu Ziwei have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements) or nonfinancial interest (such as personal or professional relationships, affiliations, knowledge, or beliefs) in the subject matter or materials discussed in this manuscript.

Funding Sources

This study was not supported by any sponsor or funder. The authors received no financial support for the research, authorship, and/or publication of this article.

Author Contributions

Conceptualization, resources, visualization, supervision, and writing – review and editing: Dr. Xu Ziwei. Methodology and writing – original draft: Dr. Yang Xiufang. All authors had access to the data and participated in the writing of the manuscript.

Data Availability Statement

All data underlying the results are available as part of the article, and no additional source data are required. Further inquiries can be directed to the corresponding author.

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