

Giant ileocecal inflammatory myofibroblastic tumor associated with paraneoplastic inflammatory syndrome in an 11-year-old girl: a case report

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Case Report

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Abstract

Background

Inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal neoplasm that infrequently arises in the pediatric colorectum. Ileocecal IMT accompanied by severe paraneoplastic inflammatory syndrome (PIS) is particularly uncommon and may mimic other childhood abdominal malignancies.

Case presentation

An 11-year-old girl presented with generalized abdominal pain, fever of 39°C, and a palpable right lower abdominal mass. Laboratory evaluation showed severe anemia (hemoglobin, 53 g/L), leukocytosis, marked thrombocytosis ($1,063 \times 10^9/L$), elevated C-reactive protein (197.89 mg/L), and hypoalbuminemia (22.6 g/L). Contrast-enhanced computed tomography demonstrated a large hypervascular ileocecal mass. Exploratory laparotomy identified a $17 \times 17 \times 8$ cm tumor intimately adherent to the ileocecal region. Ileocecal resection with ileocolic anastomosis achieved complete resection. Histopathological examination confirmed IMT; the tumor cells expressed vimentin, smooth muscle actin, and desmin and showed focal ALK positivity with both D5F3 and 5A4 antibody clones. By postoperative day 8, fever, leukocytosis, and C-reactive protein had normalized, while the anemia, thrombocytosis, and hypoalbuminemia progressively improved without additional treatment. At 2 months after surgery, the patient remained clinically well without recurrent symptoms.

Conclusions

Ileocecal IMT should be considered in children with a large hypervascular abdominal mass and systemic inflammatory abnormalities. The rapid resolution of inflammatory and hematologic abnormalities after complete resection supported tumor-associated PIS. Complete resection remains the preferred treatment for localized disease, followed by long-term surveillance.

Background

Inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal neoplasm of intermediate biological potential, characterized by myofibroblastic spindle cells and a mixed inflammatory infiltrate [1]. IMT accounts for approximately 0.04%–0.7% of pediatric tumors, with the lung being the most common primary site, followed by the abdomen and retroperitoneum [2]. The World Health Organization classifies IMT as an intermediate neoplasm that rarely metastasizes [3].

Pediatric colorectal IMT is exceptionally rare. A recent systematic review identified only 53 pediatric colorectal IMTs reported worldwide through January 2025. Among cases with a specified location, the rectum and ascending colon were the most frequently involved sites, whereas only four cecal tumors

were reported [4]. Ileocecal IMT may present with abdominal pain, fever, anemia, or a palpable abdominal mass and may clinically and radiologically mimic Burkitt lymphoma and other pediatric abdominal tumors, posing a substantial diagnostic challenge [4, 5].

We report a giant 17-cm ileocecal IMT in an 11-year-old girl with severe systemic inflammatory manifestations consistent with paraneoplastic inflammatory syndrome (PIS), including fever, severe anemia, marked thrombocytosis, elevated C-reactive protein (CRP), and hypoalbuminemia. The combination of an exceptionally large tumor, a rare ileocecal location, and the rapid postoperative resolution of the systemic inflammatory abnormalities makes this case clinically noteworthy.

Case presentation

Patient information and clinical findings

An 11-year-old girl was admitted in June 2026 with a 1-day history of persistent, dull, generalized abdominal pain without vomiting, diarrhea, or hematochezia (Table 1). Her medical, family, and psychosocial histories were unremarkable. On admission, her body temperature was 39°C. Physical examination revealed pallor of the lips and palpebral conjunctivae. A firm, mobile mass measuring approximately 15 × 10 cm was palpable in the right lower abdomen, with mild tenderness and poorly defined margins.

Table 1
Hospital Course Timeline.

Time Point	Event
1 day before admission	Onset of abdominal pain
Hospital day 1 (Admission)	hemoglobin: 53 g/L; Platelets: $1063 \times 10^9/L$; C-reactive protein: 197.89 mg/L; albumin: 22.6 g/L
Hospital day 2	Transfusion of 2 units of leukocyte-reduced packed red blood cells; Contrast-enhanced CT of the whole abdomen; complete gastrointestinal barium fluoroscopy; Ascitic fluid aspiration with cytology for malignant cells
Hospital day 4	Transfusion of 2 units of leukocyte-reduced packed red blood cells, Intravenous infusion of human albumin injection
Hospital day 6	Onset of fever (T 38°C / 100.4°F)
Hospital day 7	Fever (T 39°C / 102.2°F), Blood culture
Hospital day 10	Multidisciplinary consultation (MDT): The primary consideration is IMT; Burkitt lymphoma is not ruled out.
Postoperative day 0	Exploratory laparotomy with ileocecal resection
Postoperative day 1	Pathological diagnosis: IMT
Postoperative day 8	Body temperature: 36.7°C / 98.06°F; Hemoglobin: 76 g/L; White blood cells: $7.03 \times 10^9/L$; Platelets: $715 \times 10^9/L$; Albumin: 30.4 g/L; C-reactive protein < 10 mg/L
Postoperative day 13	Discharge
Postoperative day 20	Hemoglobin: 104 g/L; White blood cells: $5.92 \times 10^9/L$; Platelets: $382 \times 10^9/L$

Laboratory and imaging findings

Laboratory evaluation revealed severe anemia (hemoglobin, 53 g/L), leukocytosis (white blood cell count, $14.99 \times 10^9/L$), marked thrombocytosis (platelet count, $1,063 \times 10^9/L$), markedly elevated CRP (197.89 mg/L), hypoalbuminemia (albumin, 22.6 g/L), and a lactate dehydrogenase level of 94 U/L. Serum alpha-fetoprotein, carcinoembryonic antigen, and β -human chorionic gonadotropin levels were within the reference ranges. Cytological examination of ascitic fluid revealed no malignant cells, and blood cultures were negative.

Contrast-enhanced CT: The mass was located in the right lower abdomen at the ileocecal region. Dynamic contrast-enhanced scanning demonstrated significant heterogeneous progressive

enhancement. The demarcation between the lesion and adjacent bowel loops was poorly defined. Prominent arteriovenous vessels were visualized around the lesion (Fig. 1).

Diagnostic assessment and differential diagnosis

The differential diagnosis prioritized ovarian malignant germ cell tumor and abdominal Burkitt lymphoma, while also considering gastrointestinal stromal tumor and inflammatory myofibroblastic tumor.

Treatment

The patient received 4 units of leukocyte-depleted packed red blood cells before surgery. At exploratory laparotomy, a giant mass covered by the omentum was identified, with prominent hypertrophied vessels on its surface. The mass had an intact capsule and was intimately adherent to the ileocecal region. Ileocecal resection followed by ileocolic anastomosis was performed (Fig. 2).

Pathological Reports

Ileocecal mass: A lobulated, nodular mass was identified on the serosal surface of the intestinal wall, measuring $17 \times 17 \times 8$ cm. The tumor was composed of spindle-shaped fibroblasts and myofibroblasts arranged in fascicular and interlacing patterns, with abundant lymphocytic and plasma cell infiltration in the stroma. The tumor cells showed no significant atypia. Mitotic figures were rare (3 per 4 mm²). Pericolic lymph nodes (0/11) showed no evidence of tumor metastasis. Tumor cells were positive for vimentin, SMA, and desmin, with focal positivity for ALK using both the D5F3 and 5A4 antibody clones. Partial expression of AE1/AE3 and P16 was noted. CD117, DOG-1, S100, STAT6, and CD30 were negative, supporting IMT and excluding major histological mimics. The Ki-67 index was 12% in hotspots (Fig. 3).

Outcome and follow-up

By postoperative day 8, the patient's body temperature, white blood cell count, and C-reactive protein level had normalized (98.1°F, 7.03×10^9 /L, and < 10 mg/L, respectively). Her hemoglobin level had increased to 76 g/L, albumin level to 30.4 g/L, and platelet count had decreased to 715×10^9 /L. By postoperative day 20, her hemoglobin level had further increased to 104 g/L, while her platelet count had decreased to 382×10^9 /L, indicating continued improvement in the hematologic abnormalities without additional treatment. Her body temperature remained normal after surgery. Given the complete surgical resection with negative margins (R0) and the absence of lymph node metastasis (0/11), no adjuvant therapy was administered. At the most recent follow-up, 2 months after surgery, the patient remained clinically well, without recurrent fever, abdominal pain, or other tumor-related symptoms.

Discussion

The most notable features of this case are the exceptionally large tumor size (17 cm), its ileocecal location, and the presence of severe PIS. In the classic series of 84 extrapulmonary IMT cases reported by Coffin, the mean diameter was 6.4 cm, with abdominal tumors tending to be the largest, though most commonly arising from the mesentery and omentum (36/84) [6]. Of particular note, the patient exhibited pronounced systemic inflammatory abnormalities, including fever, severe anemia, leukocytosis, marked thrombocytosis, markedly elevated CRP, and hypoalbuminemia, a constellation of findings characteristic of severe PIS in patients with solid tumors [7].

In the present case, the attribution of the systemic inflammatory abnormalities to PIS was supported by their close temporal relationship with the tumor and their rapid improvement following tumor resection. Before surgery, the patient presented with a fever of up to 39°C (102.2°F), severe anemia (hemoglobin, 53 g/L), leukocytosis, marked thrombocytosis ($1,063 \times 10^9/L$), a markedly elevated CRP level (197.89 mg/L), and hypoalbuminemia (22.6 g/L), collectively indicating a pronounced systemic inflammatory response. Following complete tumor resection, her body temperature, white blood cell count, and CRP level normalized by postoperative day 8, while the albumin level increased and the platelet count progressively decreased without additional treatment. These findings support a tumor-associated inflammatory process. Although infection, tumor-related blood loss, and impaired nutritional status could also have contributed to individual abnormalities, their concurrent improvement after tumor removal favored PIS as the unifying explanation. Mechanistically, IL-6 has been proposed as a major mediator of PIS by promoting hepcidin production, stimulating thrombopoietin-mediated megakaryocyte proliferation, increasing positive acute-phase proteins such as CRP, and reducing negative acute-phase proteins such as albumin [8–10]. However, serum IL-6 was not measured in this patient; therefore, its involvement remains mechanistically plausible but unconfirmed. It is worth noting that tocilizumab has shown potential as a symptomatic treatment for severe PIS. In a retrospective study of 35 patients with solid tumors, it was associated with resolution of paraneoplastic fever and improvement in several inflammatory abnormalities; performance status improved in 31% of patients [7]. In selected patients whose inflammatory condition initially precluded surgery, temporary control of PIS with tocilizumab facilitated subsequent surgical treatment.

In this case, LDH was not elevated, no tumor cells were identified in the ascitic fluid, and CT demonstrated heterogeneous progressive enhancement with prominent feeding vessels, which also differs from the homogeneous mild-to-moderate enhancement pattern typical of lymphoma. These findings therefore do not support this diagnosis [11]. Meanwhile, normal AFP and β -hCG levels made several secreting germ cell tumor subtypes less likely [12].

Complete surgical resection (R0) is the gold standard for the treatment of IMT. The recurrence rate in patients with positive margins was significantly higher than in those with negative margins (40% vs. 0%, $p = 0.04$) [4]. Although pediatric IMT has an overall recurrence rate near 20%, colorectal tumors appear to recur less often, at approximately 11% [4].

Approximately 50–60% of IMTs harbor ALK gene rearrangements, which represent the most common molecular alterations in IMT [13]. The EORTC 90101 CREATE study indicated that crizotinib achieved an objective response rate of 50% in ALK-positive IMT patients [14]. ROS1 and NTRK3 are among the targetable driver genes detectable in ALK-negative IMT [3]. A recent institutional molecular profiling study identified a rare VCAN–IL23R fusion in one IMT case [15].

In this case, ALK status was assessed solely by immunohistochemistry, without further confirmation of ALK gene rearrangement or identification of the specific fusion partner by FISH or RNA sequencing [16].

Conclusion

Ileocecal IMT should be considered in children with a large hypervascular abdominal mass accompanied by systemic inflammatory abnormalities. Rapid resolution of the inflammatory and hematologic abnormalities after complete resection can support the diagnosis of tumor-associated PIS. Complete margin-negative resection remains the preferred treatment for localized disease, and long-term surveillance is warranted.

Declarations

Ethics approval and consent to participate

Not applicable

Consent for publication

Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images. The written consent form has been uploaded as a supplementary document (Supplementary File 1).

Availability of data and materials

All data generated or analyzed during this report are included in this published article. Additional de-identified information is available from the corresponding author on reasonable request, subject to protection of patient confidentiality.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

J.W. conceived the case report, collected and interpreted the clinical data, and drafted the manuscript. J.Z. reviewed and interpreted the radiological findings and prepared Figure 1. G.L. performed the pathological evaluation, interpreted the histopathological and immunohistochemical findings, and prepared Figure 3. C.R. and Y.Z. contributed to the clinical management, data collection, and follow-up of the patient. E.L. supervised the clinical management and critically revised the manuscript. All authors read and approved the final manuscript.

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Patient Perspective

As the parents of the patient, we were very worried about our daughter's abdominal mass after she was admitted to the hospital. We were afraid that it might be diagnosed as a malignant tumor. When she developed a fever, we felt even more overwhelmed and anxious. After the medical team's thorough evaluation and treatment, we agreed to proceed with surgery. The surgery was very successful, and our daughter recovered well without any complications. We would like to express our heartfelt gratitude to the medical team. We also consent to sharing our daughter's medical records and treatment course with other physicians.

Clinical trial number

not applicable

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Figures

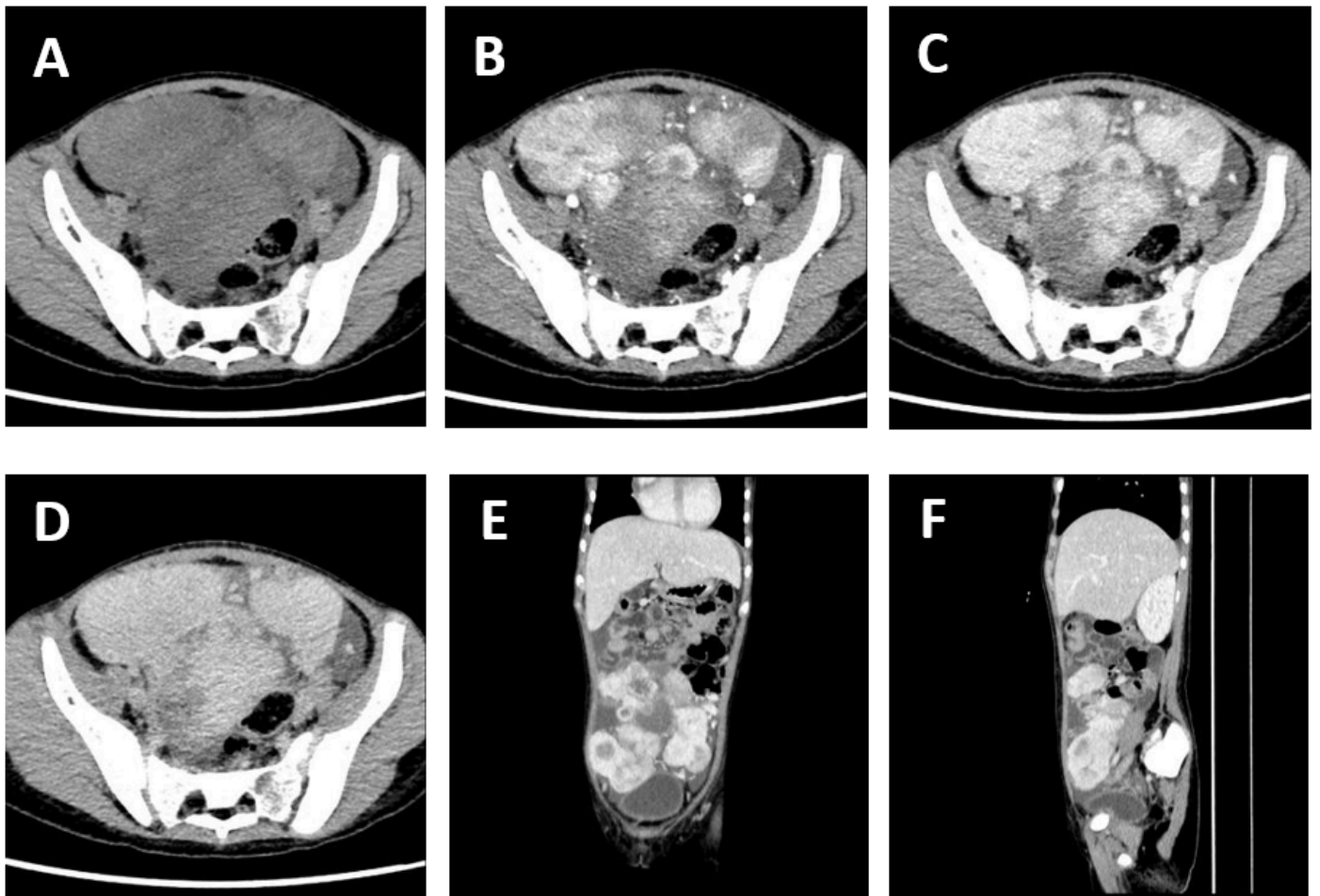


Figure 1

Multiphasic Contrast-Enhanced CT Findings of Inflammatory Myofibroblastic Tumor. A. Non-contrast phase; B. Arterial phase; C. Portal venous phase; D. Equilibrium phase; E. Coronal portal venous phase; F. Sagittal portal venous phase.

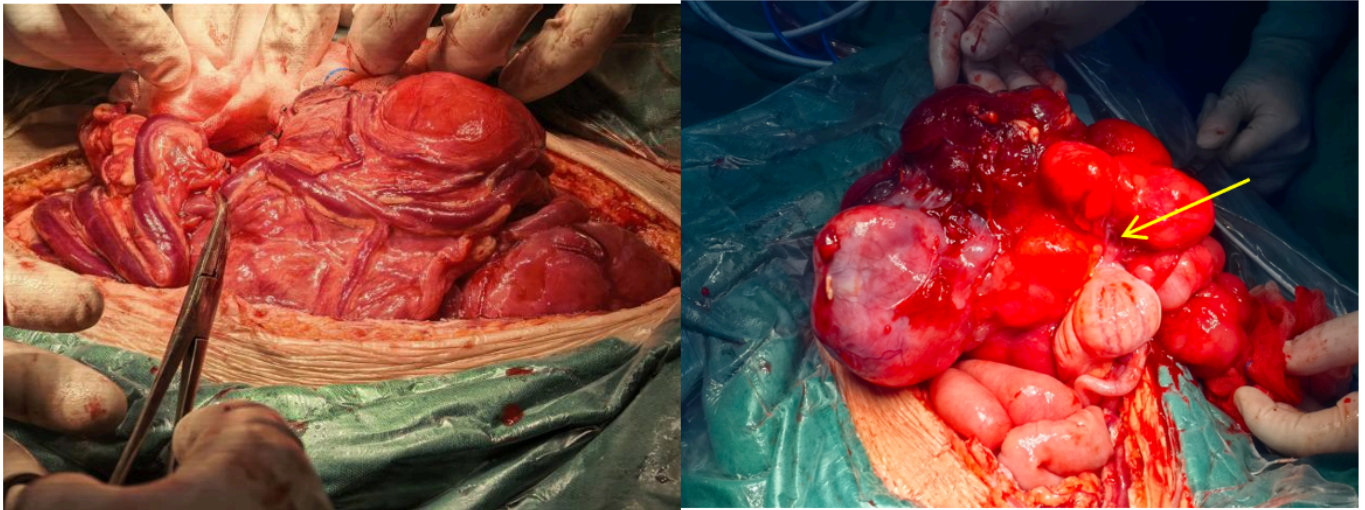


Figure 2

Intraoperative findings of intestinal **inflammatory myofibroblastic tumor**. A. Hypertrophic and enlarged feeding vessels supplying the inflammatory myofibroblastic tumor; B. Inflammatory myofibroblastic tumor adherent to the ileocecal region (Yellow arrow indicates the site of adhesion).

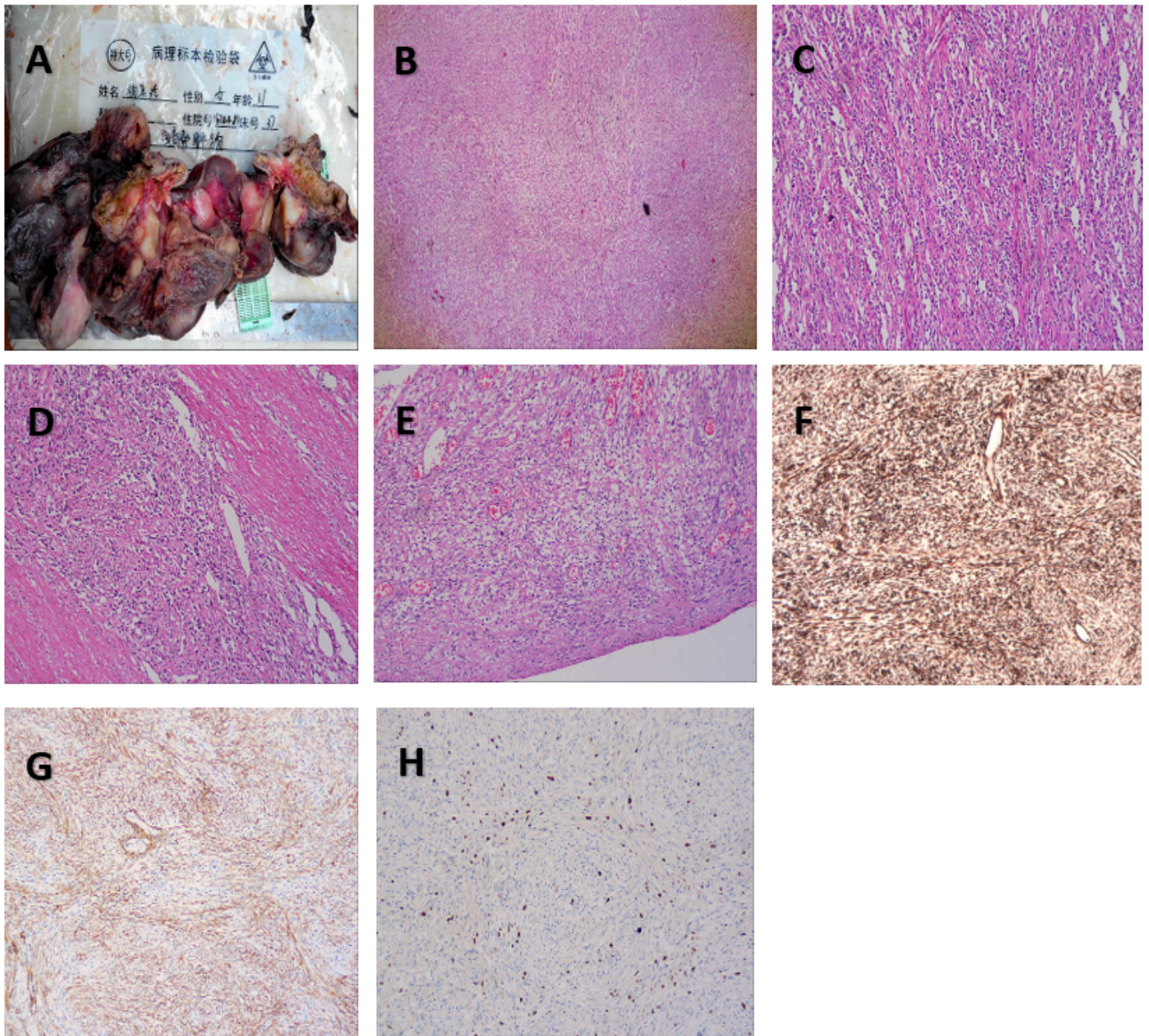


Figure 3

Clinicopathological Features and Immunohistochemical Analysis of Inflammatory Myofibroblastic Tumor. A. Gross specimen showing a lobulated, well-circumscribed nodular mass on the serosal surface of the intestinal wall with a solid cut surface; B. Spindle-shaped tumor cells arranged in fascicular and interlacing patterns (H&E); C. Prominent lymphoplasmacytic infiltration within the tumor stroma (H&E); D. Tumor infiltrating the muscularis propria of the intestinal wall (H&E); E. Well-defined tumor border at the serosal surface (H&E); F. Immunohistochemistry showing cytoplasmic and membranous positivity for ALK (D5F3 clone); G. Immunohistochemistry showing positive expression of Desmin in tumor cells; H. Ki-67 proliferation index of 12% in hotspot areas.