JOURNAL OF THE
ANUS, RECTUM AND COLON
dx.doi.org/10.23922/jarc.2022-043
http://journal-arc.jp

Case Report

# Inflammatory Myofibroblastic Tumor of the Anus: A Case Report

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**Abstract**
Inflammatory myofibroblastic tumors (IMTs) are neoplastic lesions characterized by the proliferation of spindle cells with myofibroblastic features and lymphocyte infiltration. Primary lesions can develop in several locations but rarely arise in the colon as described herein. The present case was that of a 69-year-old woman who visited our hospital with complaints of bloody bowel discharge and a prolapsed mass from the anus. A 20-mm tumor was identified on visual and digital examination. Lower gastrointestinal endoscopy revealed a pedunculated, elevated lesion above the dentate line, which showed contrast enhancement on abdominal computed tomography. The patient was preoperatively diagnosed with an anal polyp, which was resected transanally. During the procedure, a mobile tumor coated by anal epithelium was observed at the 11 o'clock position above the dentate line. Deeper parts of the tumor were contiguous with the internal anal sphincter (IAS) muscle. Suspecting a neoplastic lesion, we resected the mass *en bloc* with part of the IAS. Tumor histopathology after surgery led to a final diagnosis of an IMT of the anus. IMT is difficult to diagnose preoperatively. No adjuvant therapy has been formally established; thus, an adequate surgical margin and close monitoring are essential.

**Keywords**
inflammatory myofibroblastic tumor, transanal tumor resection, anaplastic lymphoma kinase, case report
J Anus Rectum Colon 2024; 8(1): 39-42

## Introduction

Inflammatory myofibroblastic tumors (IMTs) are neoplastic lesions characterized by the proliferation of spindle-shaped cells with myofibroblastic features, as well as lymphocyte infiltration. Formerly called "inflammatory pseudotumors" or "plasma cell granulomas"[1], the disease has been classified as an intermediate malignancy with the potential for local recurrence and distant metastasis since the 2013 revision of the *WHO Classification of Tumors of Soft Tissue and Bone*[2]. IMT is diagnosed based on pathological findings. Specimens generally test positive for α-smooth muscle actin, muscle-specific actin, and calponin in immunohistochemical assays; additionally, roughly half of the cases test positive for anaplastic lymphoma kinase (ALK). Their propensity to develop in the lungs notwithstanding, IMTs have been documented in a broad range of other sites, including intra-abdominal organs such as the bladder, mesentery, liver, and gastrointestinal tract, as well as the retroperitoneum and craniocervical region[3]. Nonetheless, IMTs seldom arise in the colon and are especially rare in the lower rectum. We could find no prior reports of primary anal IMT; thus, to our knowledge, the present study comprises the first such report.

## Case Report

The present case was that of a 69-year-old woman who

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Received: June 19, 2023, Accepted: November 8, 2023

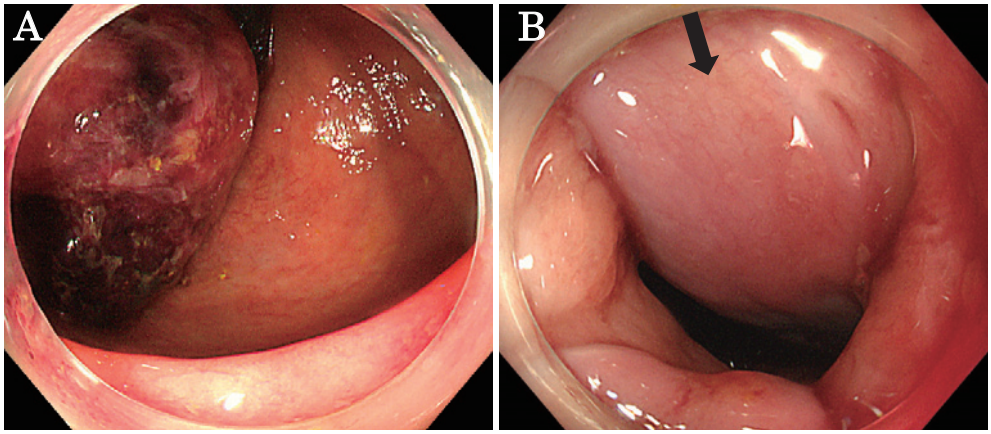


**Figure 1.** Lower gastrointestinal endoscopy.
(A) A semi-pedunculated, 20-mm, dark reddish mass with regular surface was observed.
(B) The anal canal epithelium at the 11 o'clock direction is elevated due to the presence of a tumor (arrow).

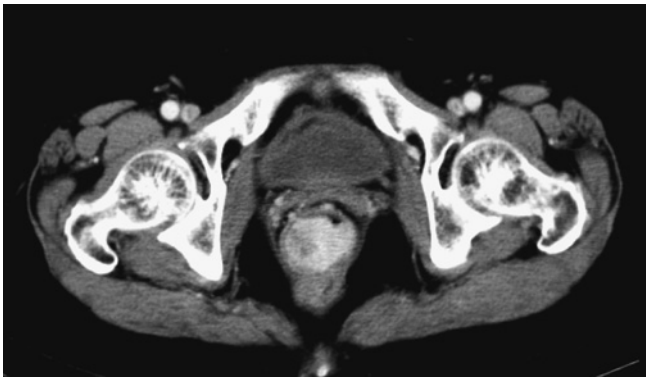

**Figure 2.** Abdominal contrast computed tomography scan. A 20-mm tumor with contrast enhancement is visible in the rectal canal.

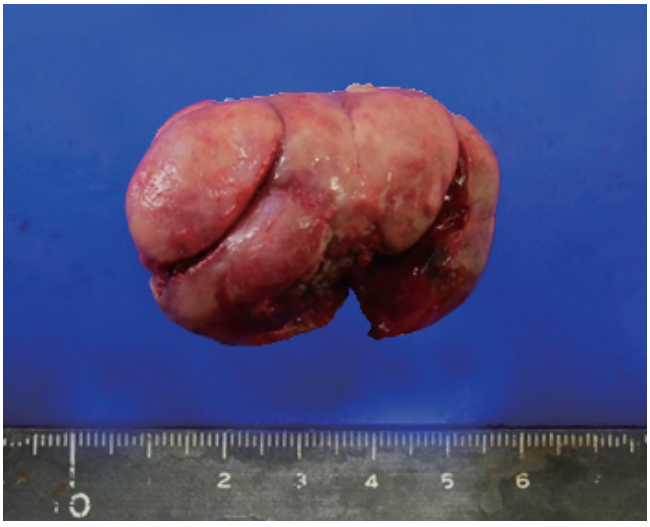


**Figure 3.** The excised lesion was elastic and soft and displayed a smooth surface.

visited our hospital with complaints of bloody bowel discharge and a prolapsed mass from the anus. The prolapse was manually reducible, but it would repeatedly prolapse and require reduction every time she defecated. The patient's previous medical history was unremarkable. An elastic, soft, 20-mm mass was identified on digital rectal examination. The patient's abdomen was flat, soft, and firm. A blood panel showed no signs of inflammation (white blood cell count: 6600/μL, C-reactive protein 0.10 mg/dL) nor anemia (red blood cell count 4,590,000/μL, hemoglobin: 13.7 g/dL). Colonoscopy showed a semi-pedunculated, well-defined, dark reddish mass in the anal canal measuring 20 mm in diameter and displaying a regular surface (Figure 1). The only salient biopsy finding was an observation of inflammatory granulation tissue. Abdominal computed tomography confirmed the presence of a 20-mm intrarectal tumor showing contrast enhancement (Figure 2). No other tumors were observed in the liver or other intraperitoneal organs. The patient was preoperatively diagnosed with an anal polyp, despite its atypical presentation, and scheduled for transanal excision. During the procedure, a mobile tumor coated by anal epithelium was observed at the 11 o'clock position above the dentate line. Its oral side extended to the rectum. Deeper parts of the tumor were contiguous with the internal anal sphincter (IAS) muscle. Suspecting a neoplastic lesion of some kind, we resected the mass *en bloc* with part of the IAS (Figure 3).

Next, the resection specimen was examined by histopathology. Spindle cell proliferation and lymphocyte/neutrophil infiltration were observed using hematoxylin/eosin staining, and ALK overexpression was observed using immunostaining. The specimen was also weakly positive for smooth muscle actin (Figure 4). Conversely, it was negative for

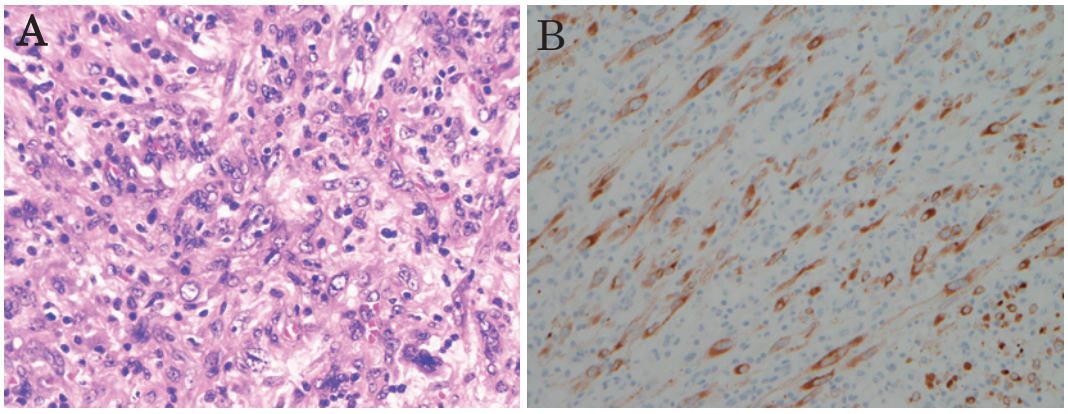


**Figure 4.** Histopathological findings.
(A) Spindle cell proliferation and lymphocyte/neutrophil infiltration are visible (hematoxylin and eosin stain: ×200 magnification).
(B) Spindle cells overexpressing anaplastic lymphoma kinase.

CD34, S-100 protein, myogenin, and cytokeratin. These findings led to a diagnosis of primary anal IMT. Upon performing immunostaining again on preoperative biopsy specimens, a mixture of ALK-positive cells was observed.

The patient's recovery was uncomplicated, and she was discharged three days after the surgery. There were no signs of recurrence at the 12-month follow-up.

## Discussion

IMTs are mesenchymal neoplasms considered to have intermediate malignant potential. Their prevalence is highest in young people, as 70% of patients are 20 years of age or younger; however, there is no difference in the prevalence between men and women[4]. Besides the lung, IMTs can develop in a broad range of locations such as the bladder, mesentery, and extremities. Primary IMTs of the colon, however, only accounted for one of 84 cases of non-lung IMTs compiled by Coffin et al.[5]. In a comprehensive review of 60 cases of colonic IMTs[6], Karaisli and colleagues found that the ascending colon was the most commonly affected site, followed in descending order by the transverse colon, cecum, and right colon, whereas IMTs developed in the rectum in just five cases (8%).

We searched the PubMed database for relevant cases reported in English between 2000 and 2022 using the keywords "inflammatory myofibroblastic tumor," "rectum/rectal," and "anus/anal." Although we found 10 reports of rectal IMT, we could not locate any reports of primary anal IMT. More than half of cases were in patients under 20 years of age at the time of diagnosis, substantiating past reports of IMT's greater prevalence in young people. IMT is a mesenchymal tumor characterized by the proliferation of myofibroblastic cells, and it often presents as a submucosal lesion. Because of this, histological diagnosis through endoscopic biopsy is challenging. In our case, the preoperative biopsy only revealed inflammatory granulation tissue, which did not lead to a diagnosis of IMT. Additionally, immunostaining performed on the preoperative biopsy samples showed ALK-positive cells, but these were mostly found within the granulation tissue. This is thought to be due to the exposure of a small amount of tumor tissue, likely due to the loss of epithelium. In the aforementioned 10 reported cases, the majority lacked a preoperative diagnosis. However, in the two most recent cases, a preoperative diagnosis was achieved using Endoscopic Ultrasound-Guided Fine-Needle Aspiration (EUS-FNA)[7,8]. In cases where preoperative biopsy samples, like in our patient, lack epithelial elements such as granulation tissue, ALK immunohistochemistry may contribute to the preoperative diagnosis. However, when normal anal epithelium or rectal mucosa is obtained in the biopsy, preoperative diagnosis through immunohistochemistry is considered challenging. Therefore, for lesions demonstrating the morphology of submucosal tumors, techniques such as depth assessment using endoscopic ultrasound and EUS-FNA may prove to be valuable diagnostic tools.

All prior cases employed surgical resection for diagnosis and treatment. The surgical procedure comprised full-thickness resection, including intestinal resection in all but two cases—the present case and one other—which were treated by local resection with partial resection of the muscular layer. One concern when an IMT develops in the lower rectum is the feasibility of preserving the anus. Shimodaira et al. performed a Miles operation to remove an IMT in the lower rectum, which was an ～8-cm mass adjoining the dentate line with infiltrative and ulcerative features[9]. We speculate that the complete resection indicated in this prior report was an additional factor that made preservation of the anus difficult in that case. Our patient's tumor aligned with the dentate line. Based on intraoperative

findings, we suspected it arose from the IAS muscle. We excised the tumor *en bloc* with part of the muscular layer and confirmed margin negativity after the surgery based on histopathological features.

No standard treatment has been established for IMTs; however, the efficacies of adrenocortical steroids, nonsteroidal anti-inflammatory drugs, and tyrosine kinase inhibitors have been reported[10]. Complete resection with a suitable surgical margin is considered effective in principle. The recurrence rate of IMTs is certainly not low. Estimates range from 18% to 40%, albeit with considerable variation, and recurrence is higher in primary non-lung than lung cases[5,11]. Furthermore, distant metastasis is observed in roughly 10% of cases[4]. Coffin and colleagues described several risk factors thought to impact IMT recurrence and distant metastasis, e.g., a larger tumor diameter was associated with greater risks of both outcomes. Recurrence and metastasis were also influenced by patient age but in opposing directions, i.e., the former was more common in older people, whereas the latter was more common among young people. Their report also links these outcomes with the presence of ALK overexpression, i.e., 54% of recurrent cases were ALK-positive, whereas all metastatic cases were ALK-negative[4]. Recurrence affected only two of the cases of rectal IMT: one case of ALK-positive local recurrence[12] and one case of ALK-negative metastatic recurrence in the liver[9]. ALK seems to influence IMT outcomes in some manner, but the detailed mechanism remains unclear.

Clinicians have few opportunities to encounter IMT, and the disease is difficult to diagnose preoperatively. Because no adjuvant therapy has been formally established, ensuring a sufficiently large surgical margin and closely monitoring patients during follow-up appear to be crucial for positive outcomes.

Acknowledgements

We thank Keisuke Ishizawa and Mieko Doi for additional immunostaining.

Conflicts of Interest

There are no conflicts of interest.

Author Contributions

Hiroshi Asano determined the treatments. Tetsuyoshi Takayama, Ayako Nakame and Masaomi Suzuki and Hiroshi Asano performed the surgeries and participated in the treatment of the patient. Ling Jin provided pathological data. All authors read and approved the final version of this manuscript.

Approval by Institutional Review Board (IRB)

Not applicable.

Informed Consent

Written informed consent was obtained from the patient and her family.

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