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Rare case of small bowel inflammatory myofibroblastic tumor

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Abstract

The inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal neoplasm with variable biological behavior, which predominantly affects the lungs but can also appear in extrapulmonary sites, including the gastrointestinal tract. When situated in the small intestine, IMT presents substantial diagnostic and therapeutic difficulties. This case emphasizes the complications in diagnosing and surgically managing IMT in an unusual location. A patient, aged 67 with a past record of secondary iron-deficiency anemia, underwent a diagnostic examination due to gastrointestinal symptoms. The esophagogastroduodenoscopy showed a minor hiatal hernia, while a 5 mm hyperemic polyp, confirmed as a tubular adenoma, was identified by colonoscopy. Further MR enterography imaging detected a polypoid tumor approximately 28 mm in the right iliac region, which led to surgical intervention. A lower midline laparotomy revealed an intraluminal tumor in the proximal ileum, 60 cm from the ileocecal valve. A 20 cm segment of the impacted bowel was resected and an end-to-end ileo-ileal anastomosis was successfully achieved. The diagnosis of IMT with an R0 resection was confirmed through histopathological and immunohistochemical analysis. This case underlines the significance of considering IMTs in the differential diagnosis of unexplained gastrointestinal symptoms and highlights the role of imaging and surgical resection in reaching definitive management. Boosting awareness of IMTs in medical practice can lead to earlier recognition and improved patient outcomes.

Keywords: inflammatory myofibroblastic tumor, small intestine neoplasm, chronic secondary anemia, rare gastrointestinal neoplasms

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Introduction

Inflammatory myofibroblastic tumor (IMT), also referred to as inflammatory pseudotumor or plasma cell granuloma, is a rare mesenchymal neoplasm. It is distinguished by the proliferation of myofibroblastic spindle cells as well as an inflammatory infiltrate consisting of plasma cells and lymphocytes. Brunn first described it in 1939 as a lung tumor, initially believed to be benign with localized growth. However, later variations with local malignancies and rare metastases have been recorded. The World Health Organization (WHO) now classifies IMT as a mesenchymal neoplasm of intermediate malignancy [1].

IMT can occur at any age, with most patients receiving diagnoses before the age of 40. However, IMTs can also develop later in life, even in the eighth decade. IMT has been observed in nearly all body and visceral soft tissue organs. The most common site for IMT is the lungs, but extrathoracic forms have been detected at various other locations, primarily in soft tissues and visceral organs [2]. The small intestine is a rare site for IMT, and cases in this region frequently pose a significant diagnostic and therapeutic challenge.

The etiology of IMT is unclear, with certain factors suggested to be associated with it, such as viral infections (particularly HHV-8), trauma, surgical interventions, and autoimmune mechanisms. The anaplastic lymphoma kinase (ALK) gene is found in 33-67% of cases and is more commonly present in children and young adults [3,4].

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The clinical manifestations of IMT are nonspecific and vary depending on the location of the tumor. For instance, in the gastrointestinal tract, IMT may result in symptoms like abdominal pain, anemia, obstruction, or intussusception, which are attributed to its intraluminal growth. Furthermore, it can present as either acute or chronic gastrointestinal bleeding and may ultimately lead to iron-deficiency anemia [5, 6].

IMTs are typically benign and are usually treated with radical excision, steroids, radiation, and/or chemotherapy [7]. CO_2 laser treatment represents a novel therapeutic option. In the case of small intestine IMT, the recommended treatment is segmental resection with negative margins, carrying a minimal risk of recurrence if the excision is thorough.

Case presentation

A 67-year-old patient has been diagnosed with, and treated for, secondary iron-deficiency anemia within the past year by a gastroenterologist. Upon evaluation, an esophagogastric duodenoscopy (EGD) revealed merely a minor hiatal hernia, absent of any additional pathological findings. A colonoscopy detected a hyperemic polyp, approximately 5 mm in size, situated in the descending colon; it was subsequently removed via electrosurgery.

Histopathology confirmed a tubular adenoma. Further examination revealed no abnormalities in the entire colon and the terminal 30 cm of the ileum. A CT scan of the chest, abdomen, and pelvis also showed no remarkable findings. However, MR enterography revealed a loop of a small bowel in the right iliac region with indications of mechanical obstruction, as well as a polypoid tumor measuring around 28 mm.

Following the diagnostic evaluation, a need for surgical intervention was established to firm up the diagnosis and excise the tumor. Preoperative preparations entailed routine lab tests, a cardiopulmonary assessment, and optimizing the patient's readiness in light of the heightened risks associated with the perioperative period. Informed consent was gathered for the surgery, which delineated potential complications such as infection, hemorrhage, anastomotic leakage, and the possible necessity for additional intervention.

The surgery was conducted under general endotracheal anesthesia, commencing with a lower midline laparotomy for peritoneal access. The abdominal cavity was extensively explored to identify any additional pathology. During the exploration, an intraluminal tumor was discovered in the proximal ileum, roughly 60 cm from the ileocecal valve. The extent of the tumor was confirmed by palpation, and the segment of the small intestine containing the lesion was mobilized.

The resection involved a meticulous dissection of the mesentery supplying the affected segment in order to ensure proper vascular control. Once the ileum was sufficiently mobilized, a 20 cm resection of the affected bowel was carried out (Figure 1, 2). This step was executed with thorough attention to detail, ensuring clear margins for complete tumor removal. Throughout the process, hemostatic control was consistently maintained.

Following resection, an end-to-end ileo-ileal anastomosis was performed. The anastomosis was created using delayed absorbable sutures in a two-layer technique, including an inner continuous mucosal layer and an outer interrupted seromuscular layer. This ensured a tension-free, well-perfused anastomosis. Additionally, the mesenteric defect was sutured to prevent internal herniation. Before closing the abdominal wall, the surgical area was irrigated to remove any debris or contaminants.

The histopathological examination of the resected specimen revealed an IMT located in the submucosa. The tumor was completely removed, thus achieving an R0 resection. Characteristically, IMT is composed of myofibroblastic and fibroblastic spindle cells, supplemented by an inflammatory infiltrate including lymphocytes, plasma cells, and eosinophils. The tumor demonstrated variable cellularity, possessing areas with sparse cellular activity within the hyalinized stroma and other areas showcasing dense myofibroblastic proliferation.

The immunohistochemical analysis of the tumor confirmed the diagnosis, showing positivity for vimentin, smooth muscle actin (SMA), and muscle-specific actin, which is consistent with myofibroblastic differentiation. Further, the tumor also demonstrated positivity for ALK, a feature frequently seen in IMTs. Other markers like desmin, cytokeratins, and S-100 were negative, thus eliminating other differential diagnoses.

The patient had an uneventful postoperative recovery, with bowel function gradually returning. Oral intake was resumed without issues and there were no signs of complications such as infection or anastomotic leakage. The patient was discharged home in stable condition and instructed to follow up regularly with the attending surgeon for monitoring and additional treatment as needed.

Figure 1: Resected small intestine with umbilication of serosa at the site of the intraluminal inflammatory myofibroblastic tumor.

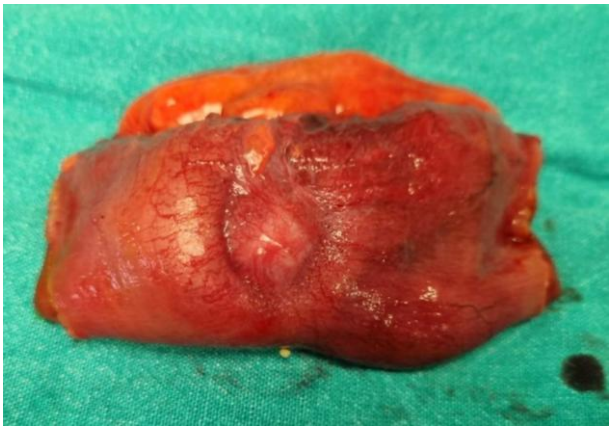
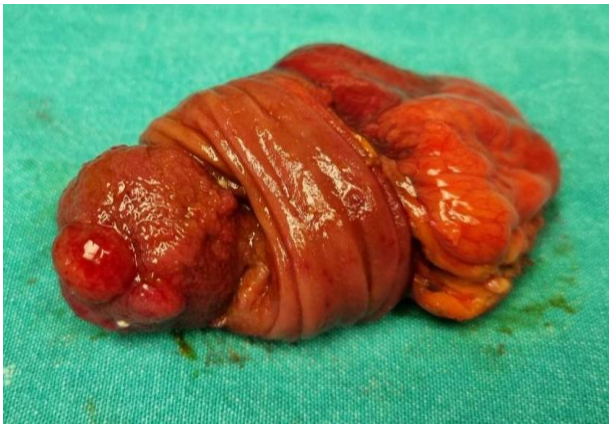


Figure 2: Resected small intestine with intraluminal inflammatory myofibroblastic tumor.



Discussion

IMT represent a rare and heterogeneous group of neoplasms, presenting significant challenges for diagnosis and treatment. Our case report emphasizes key aspects of IMT, including its clinical presentation, surgical approach, and histopathological as well as immunohistochemical diagnosis.

IMTs are typically benign neoplasms that exhibit locally aggressive behavior. These are most commonly found in younger patients, with a peak incidence in children and young adults, but they can occur at any age, including in elderly individuals as demonstrated in our case [1,2]. The clinical presentation of IMT is location-dependent and can include symptoms such as obstruction, a palpable mass, or nonspecific abdominal pain [3]. In the case of this patient, the secondary iron-deficiency anemia is likely attributed to chronic, hidden blood loss induced by the tumor.

The differential diagnosis of small bowel tumors includes gastrointestinal stromal tumors (GISTs), adenocarcinomas, neuroendocrine tumors, and lymphomas. Imaging and histopathological findings are crucial in distinguishing IMTs from these entities. Given the polypoid nature and obstructive symptoms in our patient, considerations included GISTs and adenocarcinomas. These were ruled out through immunohistochemical analysis.

Radiologic assessment plays a key role in detecting IMTs, although the imaging findings are often nonspecific. In this patient, MR enterography revealed a polypoid tumor causing mechanical obstruction. Because of the tumor's location, obstructive symptoms, and the need for a definitive diagnosis, surgical management was prioritized over medical therapy options like corticosteroids or NSAIDs [4,5].

Surgical excision remains the cornerstone of treatment for IMTs, aiming for R0 resection to minimize the risk of recurrence [6]. In our case, an R0 resection was achieved, which is associated with a favorable prognosis. Studies indicate that complete resection leads to excellent survival outcomes, while incomplete excision carries recurrence rates of up to 25–37% [5,7].

Histopathologically, IMTs are characterized by the proliferation of spindle cells amidst an inflammatory background made up of lymphocytes, plasma cells, and eosinophils [3]. The diagnosis is confirmed by the presence of ALK positivity in tumor cells. ALK rearrangements, which occur in roughly 50% of IMTs, are frequently observed in younger patients and act as a crucial diagnostic marker [8].

For unresectable or recurrent IMTs, ALK inhibitors like crizotinib have demonstrated effectiveness in reducing ALK-positive tumors [9,10]. These agents are especially relevant for patients who aren't suitable candidates for surgery. In our case, successful complete resection made additional therapy unnecessary. However, long-term monitoring is critical in identifying potential recurrence.

Conclusion

This case emphasizes the need to consider IMT in the differential diagnosis of unexplained gastrointestinal symptoms, especially in instances of chronic anemia with initial inconclusive findings. Due to the tumor's potential for local aggression and recurrence, prompt imaging and histopathological confirmation are vital for directing definitive management. Surgical resection continues to be the primary treatment, with R0 resection providing the best prognosis. Enhancing clinical awareness of IMT, particularly in atypical locations like the small intestine, can facilitate earlier diagnosis, optimize treatment strategies, and ultimately improve patient outcomes.

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