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Inflammatory Myofibroblastic Tumors in a Case with Hypogastric Discomfort




Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 68
Final Diagnosis: Inflammatory myofibroblastic tumor
Symptoms: 2–3 kg weight loss • *Helicobacter pylori* positive which were eradicated after treatment course • hypogastric region discomfort • no association between the pain, defecation and eating • no history of HIV, diabetes, smoking and alcohol consumption was recorded • no symptoms of nausea, vomiting, fever and shaking chills • normal vital signs • tenderness in the right lower quadrant of the abdomen with no rebound state was noted
Medication: —
Clinical Procedure: Lab examination • imaging • colonoscopy • surgery
Specialty: Gastroenterology and Hepatology
Objective: Rare disease
Background: Inflammatory myofibroblastic tumors (IMTs) are scarce tumors with discrete immunohistochemical and molecular attributes which are not related to a particular location. There are different reports about the intrinsic nature of these tumors as benign to possibly malignant.
Case Report: Here we report the case of a 68-year-old man referred to the Internal Medicine Department of Razi Hospital in Rasht (a city in the north of Iran) due to right lower quadrant (RLQ) discomfort with no specific symptoms. Colonoscopy revealed a mass-like lesion. Polymorphonuclear cells (PMNCs) admixed by some eosinophils were demonstrated histopathologically. Immunohistochemical evaluation was positive for vimentin, CD34, smooth muscle actin, and ALK, and negative for CD117 and desmin. The tumor was successfully removed by surgery with no chemotherapy. No recurrence was reported.
Conclusions: We have performed surgical excision of the mass with no chemotherapy and no recurrence. Although recurrence is reported to be low, we recommend long-term follow-up after surgery.
MeSH Keywords: Biological Markers • Colonoscopy • Gastrointestinal Tract
Full-text PDF: <http://www.amjcaserep.com/abstract/index/idArt/901813>

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Background

Inflammatory myofibroblastic tumors (IMTs) are scarce mesenchymal solid tumors [1] with discrete immunohistochemistry and molecular attributes [2], which are not related to a particular location [3], but mostly involve the pulmonary tract [4]. There are different reports about the essence of these tumors as benign to possibly malignant [4,5]. IMT may impact any age and sex [1,2]. It may develop in organs such as the liver [6], stomach, small intestine, large intestine, mediastinum, retroperitoneum, and bladder [5,7–9]. One of the rare complications of this tumor is its presence in the colon and intussusceptions [4]. Due to the determination of IMT by the site of origin and mass effects, its diagnosis can be difficult based on non-specific clinical symptoms [1,2]. For example, gastrointestinal (GI) tract derived IMT is not distinguishable from other GI tract tumors, as it has the same clinical indications such as anemia, GI obstruction, fecal occult blood positivity, or intussusceptions [2].

Case Report

A 68-year-old man referred to the Internal Medicine Department of Razi Hospital in Rasht (a city in the north of Iran) with a hypogastric region discomfort, especially in the right lower quadrant for one month. The pain was a colicky form which had a few episodes each day, each episode lasting for 4–5 minutes. The pain radiated to the back and was alleviated by resting to one side. No association between the pain, defecation, and eating were reported. Furthermore, the patient complained of a 2–3 kg weight loss over the last one month followed by anorexia. However, no symptoms of nausea, vomiting, fever, or shaking chills were demonstrated. He was first admitted to another center for a week, then was referred to our hospital for further evaluation. The patient had undergone a surgery for prostatectomy seven years before his presentation to our center. Also, he had a history of endoscopy five years earlier due to dyspepsia, which was found to be *Helicobacter pylori*

positive at that time and which was eradicated after a treatment course. No history of HIV, diabetes, smoking, or alcohol consumption was recorded. His vital signs were normal at the time of admission. On physical examination, the abdomen was soft, there was no distention, and bowel sounds were normoactive. However, tenderness in the right lower quadrant of the abdomen with no rebound state was noted. The peripheral blood analysis is shown in Table 1. Stool examination, urine analysis, and evaluation of electrolytes were all normal. An abdominal CT scan showed a well-demarcated and homogeneous solitary mass in the cecum with no distention in the ileum. The ileum wall was thickened (Figure 1). A colonoscopy revealed a large mass like lesion in the cecum (Figure 2); during the procedure a biopsy was taken from the cecum. The lamina propria was infiltrated by a number of PMNCs admixed by some eosinophils (Figure 3). Immunohistochemical evaluation was positive for vimentin and CD68. C-Kit (CD117) was negative while CD34, smooth muscle actin (SMA), and ALK were focally positive. There were no reports of cyclin D1, desmin, or pancytokeratin (Figure 4). When the colonoscopy was performed, the evidences of invasive obstruction lead us to the suspicion of a malignant tumor. After pathological confirmation of IMT, the patient was referred for surgery in order to remove the mass. On surgery, approximately 40 mL ascites were found in the abdomen. A mass was seen in the cecum with ileocolic intussusception. Afterwards, the patient underwent right hemicolectomy with an end-to-end anastomosis of ileocolic. No enlarge lymph nodes were observed. The patient was discharged seven days after surgery and had no complications during follow-up.

Discussion

In spite of different synonyms for IMTs used in the past, and the difficulty in obtaining data about this rare condition [1], the World Health Organization has now classified IMTs as tumors with transitional nature as indicated by its penchant to local recurrence and low risk of distant metastases [2]. IMT was initially

Table 1. Result of lab test.

WBC	8.7×10 ³ /μl	CRP	45 mg/l
HB	12.1 g/dl	ESR	68 mm/hr
HCT	38.3%	BS	86 mg/dl
Platelet	332×10 ³ /μl	LFT	Normal
Neutrophil	60%	Ferritin	70 ng/ml
Lymphocyte	30%	Fe	59 mg/dl
Monocyte	4%	TIBC	400 mg/dl
Eosinophil	6%		

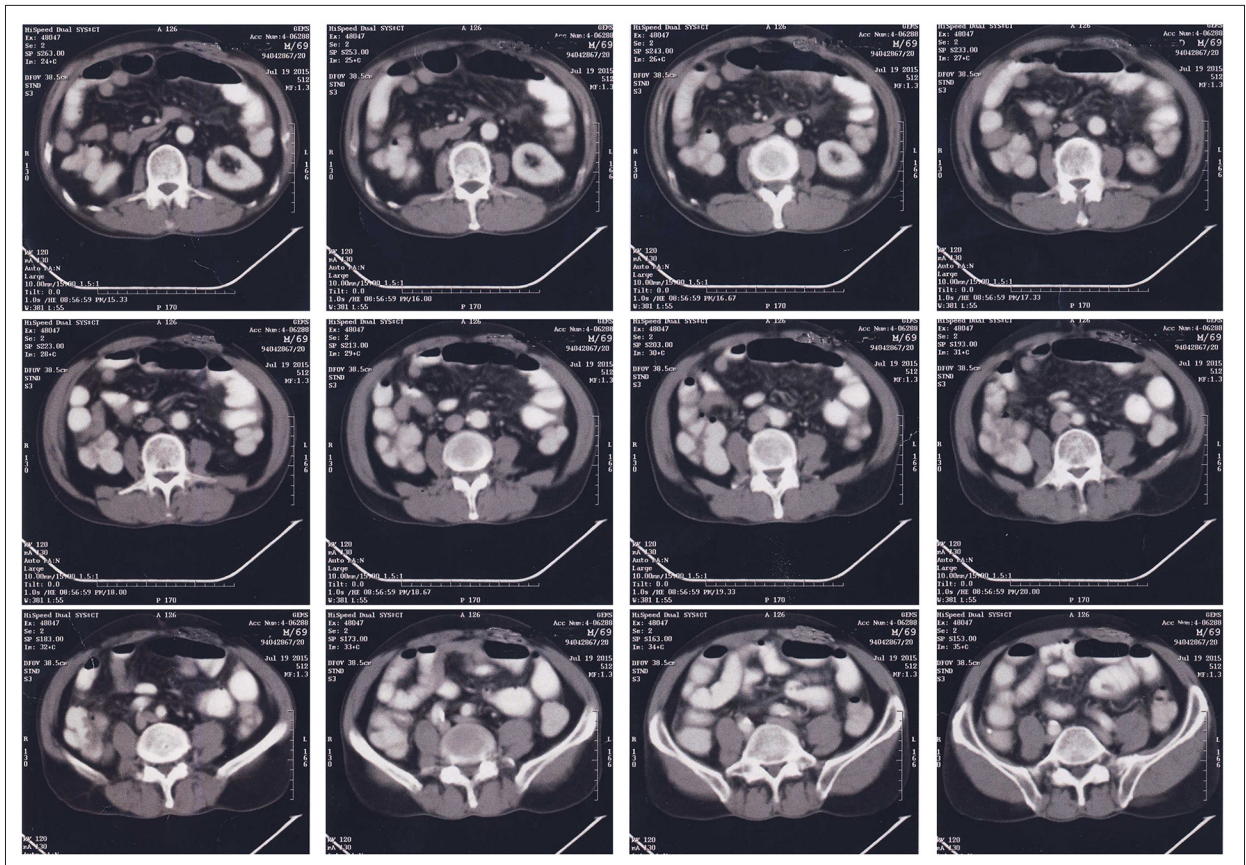


Figure 1. Computer tomography image shows homogenous solitary mass in the cecum.

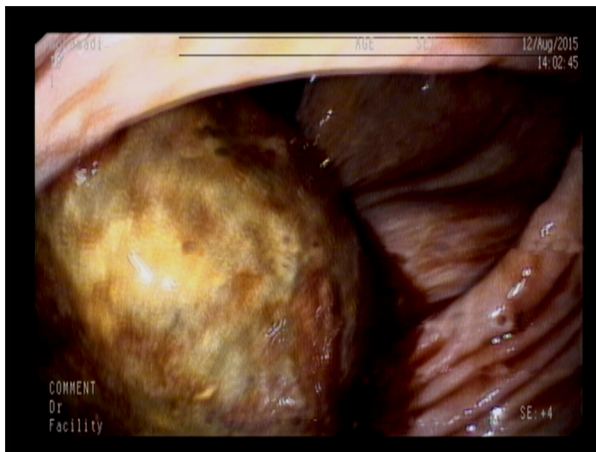


Figure 2. Colonoscopy, mass like lesion in the cecum.

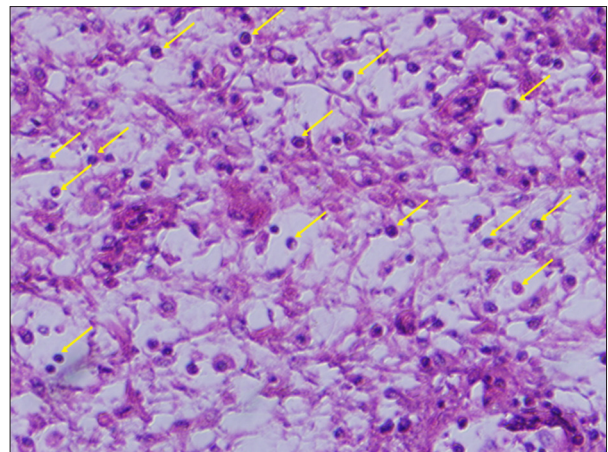


Figure 3. Part of the tumor (IMT) (H & E ×400), the arrows indicate the PMNs.

reported as occurring in the lungs, but it can occur in the upper respiratory tract, mesentery/omentum (the most common extra-pulmonary site at 43%), mediastinum, retroperitoneum, genitourinary tract, gastrointestinal tract, pelvis, trunk, extremities, brain, neck, head, spleen, liver, and pancreas [3]. The etiological elements are not clear; albeit a few instances of IMT are considered an inflammatory reaction to contamination, injury, or surgery [4]. Both genders have similar incidence rates, and

no geographic or ethnic inclination has been identified [10]. The clinical presentation is dictated by the site of origin and the impact of the developing mass [11]. Although the etiology of IMT is still obscure, reports suggest that the advancement of IMT may happen after an injury, surgery, or disease; for example, Epstein-Barr virus infection and human herpes infection have been associated with responsive cytokine creation [6,7]. A recent study

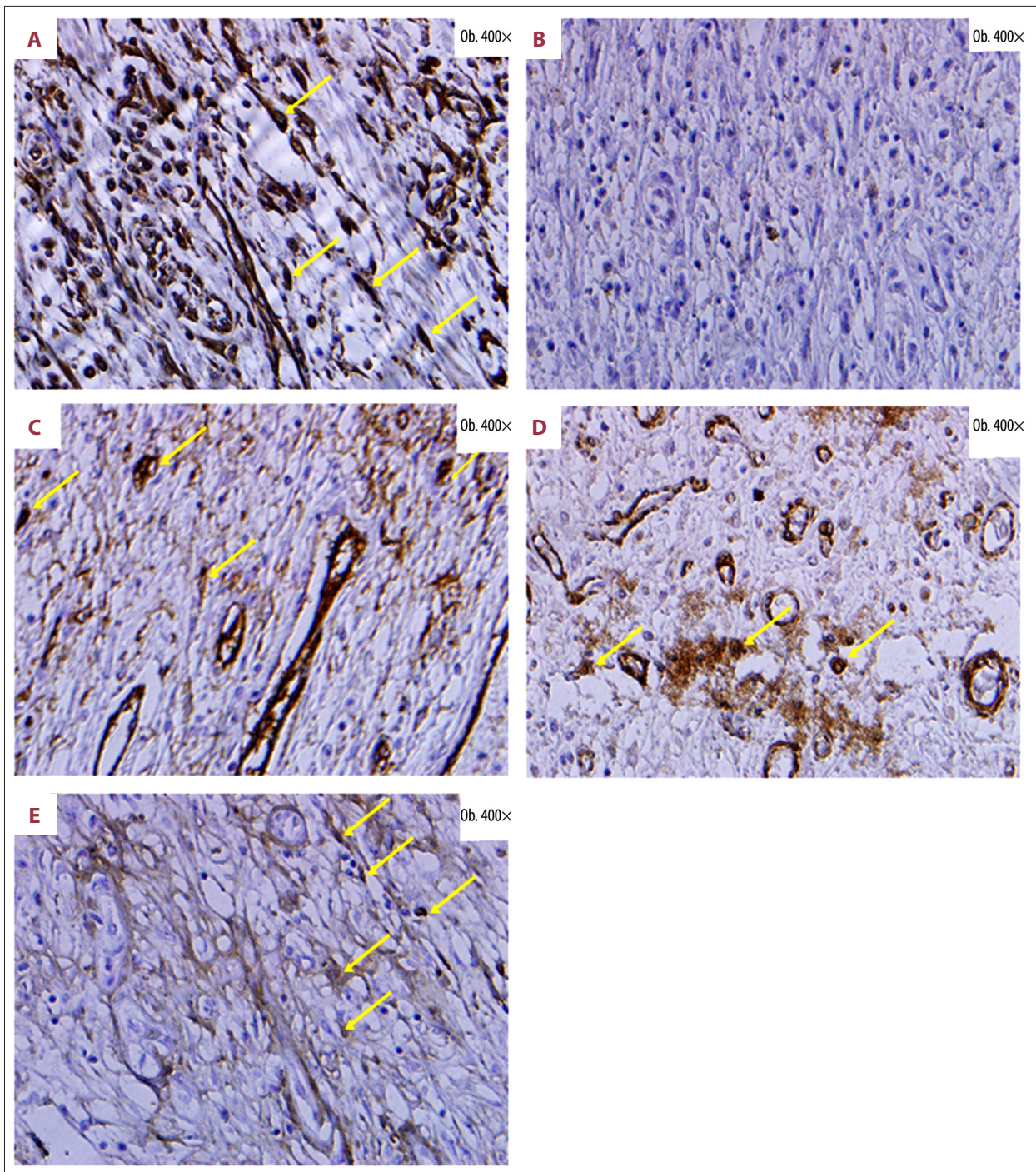


Figure 4. Immunohistochemical evaluation was positive for vimentin (A) and CD68. C-Kit (CD117) (B) was negative while CD34 (C), smooth muscle actin (SMA) (D) and ALK (E) were focally positive.

reported that local aggressive behavior and monoclonality may be the result of chromosomal aberrations induced by lesions. All things considered, IMT may be better viewed as a neoplastic condition. Recent discoveries have demonstrated that chromosomal variations from the norm might be suggestive of clonal inception, not simply a receptive procedure. The origin site and

mass effects determine the clinical presentation as well as possible anemia, abdominal pain, loss of appetite and weight, fecal occult blood positivity, and intussusceptions, which are listed as nonspecific usual symptoms in patients with GI tract IMTs [8,10,11]. Colorectal carcinoma has similar clinicopathological features as IMTs in the colon and rectum, such as altered

bowel habits, diarrhea, or constipation [8]. In order to choose the best treatment, distinguishing IMT from other colorectal tumors is critical, however, endoscopic and computed tomography differential diagnosis would not be able to distinguish these conditions from each other [12]. Ill-defined, none capsulated, protruded masses or infiltrative tumors are the most similar gross features of reported colorectal IMTs that transition to cancers [8]. In our case, the tumor was an intraluminal mass with a focal depression secured by normal mucosa similar to a classic GI stromal tumor (GIST) or lymphoma. It has been reported that the diagnosis of IMT from other malignant, infectious, and congenital lesions depends on the histologic examination of the tissue [13]. Spindle cells along with a variable infiltration of dense polymorphic mononuclear inflammatory cells made out of lymphocytes, plasma cells, histiocytes, and occasional eosinophils are histological characteristics of IMTs, which arrange in short fascicles with focal storiform architecture, as a result of the spindle cells [11,14]. For a conclusive finding of an IMT, immunohistochemistry is essential [8]. In our case, tumor cells were characteristically positive for vimentin. They were focally positive for CD34, SMA, and CD68 expression except for CD117. In general, IMTs seem to be well vascularized and contained numerous thick-walled vessels. As indicated an earlier study, the presence of SMA and CD34 likely demonstrates late angiogenesis [15]. Although roughly half of IMTs are positive for immunohistochemical cytoplasmic ALK utilizing an assortment of monoclonal antibodies, this positivity is not particular for IMT [12]. In our case, ALK was positive. Abdominopelvic site, larger size, and older age are generally associated with local recurrences. Furthermore, larger size, and both abdominopelvic and pulmonary sites among younger age patients are related to distant metastasis. While

the predilection for lung, brain, liver, and bone has been estimated to be less than 5% [10], the treatment decision is generally complete surgical resection. Several studies have reported disappointment with glucocorticoids, radiotherapy, and chemotherapy [16–18]. In patients with morbid, impossible resection or an inadequate resection, chemotherapy has been restrained. In furtherance of reducing the surrounding inflammation, steroids may be added if required, especially in patients with nervous system involvement. In limited circumstances, because of invading vital structures of tumors, nonsteroidal anti-inflammatory drugs (NSAIDs) have been used successful. A powerful remedial alternative for patients with unresectable IMT might be chemotherapy combined with oral NSAIDs [3].

Conclusions

We have performed surgical excision of the mass with no chemotherapy and no recurrence. Although recurrence is reported to be low, we recommend long-term follow-up after surgery.

Acknowledgment

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Conflict of interest

There is no conflict of interest.

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