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Incomplete Intestinal Obstruction Caused by a Rare Epithelioid Inflammatory Myofibroblastic Sarcoma of the Colon

A Case Report

Yanjun Bai, MD, Maofen Jiang, MD, Wenjie Liang, MD, and Feng Chen, MD, PhD

Abstract: We reported on 1 case of epithelioid inflammatory myofibroblastic sarcoma (EIMS) that occurred in the colon and resulted in an incomplete intestinal obstruction.

A 65-year-old male patient presented with abdominal pain without any obvious predisposing cause. He reported a paroxysmal dull pain. Hematochezia occurred occasionally. The symptoms appeared repeatedly and became progressively more aggravated. The patient sought medical advice in our hospital, and his enteroscopy showed colon tumors and an incomplete colonic obstruction. The laboratory examination indicated mild anemia. Plain and enhanced computed tomography (CT) scans showed a large, dumbbell-shaped, soft-tissue mass of 4.1 cm × 9.3 cm in the curved lumen of the descending colon near the spleen. After enhancement, the lesion presented with progressive and uneven enhancement. The boundary between the lesion and parts of the left kidney and spleen was obscured. A small amount of exudation was observed around the lumen, and a slightly enlarged lymph node shadow was observed in the mesangial gap.

After each preoperative examination was completed, the tumors invading the spleen and left kidney were excised. Based on the surgical specimen pathological histology and immunohistochemistry, epithelioid inflammatory myofibroblastic sarcoma was diagnosed. Tumor recurrence occurred a short time after excision.

EIMS in the abdominal cavity could occur on the intestinal wall, occasionally manifesting as large masses that expand to the inside and the outside of the cavity. It needs to be distinguished from other tumors. Tumor recurrence can easily occur after surgery. Anaplastic lymphoma kinase (ALK) inhibitors are a potential alternative treatment option.

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Abbreviations: CT = computed tomography, DWI = diffusion-weighted imaging, EIMS = epithelioid inflammatory myofibroblastic sarcoma, IMT = inflammatory myofibroblastic

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Received: November 18, 2015; revised and accepted: November 26, 2015. From the Department of Radiology, (YB); Department of Pathology, the People's Hospital of Beilun District, Beilun Branch Hospital of the First Affiliated Hospital (MJ); and Department of Radiology, the First Affiliated Hospital, College of Medicine, Zhejiang University, Hangzhou, Zhejiang, China (WL, FC).

Correspondence: Wenjie Liang, Department of Radiology, the First Affiliated Hospital, College of Medicine, Zhejiang University, 79# Qingchun Road, Hangzhou, Zhejiang, China (E-mail: baduen.c@163.com).

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tumor, MRI = magnetic resonance imaging, T1WI = T1-weighted imaging, T2WI = T2-weighted imaging.

INTRODUCTION

The inflammatory myofibroblastic tumor (IMT), previously known as an inflammatory pseudotumor, is a type of mesenchymoma composed of spindle cells and often accompanied by a large number of plasmacytes and lymphocytes. IMT was first described in the lung, followed by reports of cases outside the lung.^{1,2} Approximately two-thirds of IMT cases are extrapulmonary, usually occurring in the mesenteries, omentums, organs within the abdominal cavity, urinary system, and upper respiratory tracts.^{3,4} In the past, IMT was considered to appear mostly in children and teenagers and to have a relatively benign clinical course. However, in a group of extrapulmonary IMT cases, 13/84 cases recurred at least once, although there were no metastatic tumors.⁴ In another group of IMT cases in the mesenteries and omentums, a more significant recurrence rate of 10/38 occurred, with 3 of those being metastases.⁵ Based on this evidence, IMT was listed as an intermediate-grade tumor in the WHO classification.⁶ Recently, research has shown that IMT cases within the abdominal cavity have an epithelioid shape and a positive anaplastic lymphoma kinase (ALK) of the karyotheca or perinuclear areas.⁷ All 11 cases recurred quickly, and 2 of those were metastases with stronger invasiveness.⁷ Therefore, this group of cases was named “epithelioid inflammatory myofibroblastic sarcoma.”⁷ In this paper, we report on 1 case of incomplete intestinal obstruction caused by a rare epithelioid inflammatory myofibroblastic sarcoma (EIMS) of the colon.

CONSENT

The patient signed relevant documents to allow clinical information and images to be used for teaching and publishing.

CASE REPORT

A 65-year-old male patient presented with abdominal pain without any obvious predisposing cause that began 1 month before. It was a tolerable paroxysmal dull pain that was more severe below the umbilicus. The pain could be relieved after flatus and defecation. Stools mixed with hematochezia occurred occasionally and were of a kermesinus color. There were no symptoms of illness, such as chills, fever, nausea, vomiting, headache, or dizziness. There was no obvious change in bowel habits and no special handling was used. Thereafter, the symptoms noted above appeared repeatedly with a progressively aggravated trend. He sought medical advice in our hospital, and his enteroscopy showed “colon tumors and incomplete colonic obstruction.” For the purpose of further treatment, he came to

our hospital again. The outpatient department admitted him to the hospital for “colon tumors.” Despite the attack, the patient had no obvious change in body weight and was generally healthy. The patient had a 30-year history of smoking and alcoholism. In a local hospital, a plain computed tomography (CT) scan of the upper abdomen showed a large, dumbbell-shaped mass opacity in the left descending colon, which was considered to be an inflammatory mass, a tumor, or a tumor with infection. The CT scan of his lung in the local hospital showed chronic inflammation with local mild bronchiectasis. The patient’s vital statistics were T: 37.1 °C, P: 111/min, R: 19/min, and BP: 113/69 mm Hg. The abdomen was flat. There was mild tenderness around the umbilicus without rebound tenderness. No obvious mass was felt. The liver and the spleen were not felt under the ribs. The gall bladder was tender when touched. The Murphy sign was negative, and there was no percussion pain in the renal region. Borborygmi were 4/min with negative shifting dullness. No abnormal neoplasm was found during an anal examination, and there was no blood on the dactylothea. The routine blood examination results were RBC: $3.78 \times 10^{12}/L$ (reference range 4.00–5.50 $\times 10^{12}/L$); HGB: 115 g/L (reference range 120–160 g/L); PLT: $329 \times 10^9/L$ (reference range 100–300 $\times 10^9/L$); PCT: 0.35 (reference range 0.11–0.28); NEU: 72.5% (reference range 50.0–70.0%); LYM: 16.2% (reference range 20.0–40.0%); TP: 67.2 g/L (reference range 60.0–83.0 g/L); GLO: 35.2 g/L (reference range 20.0–35.0 g/L); A/G ratio: 0.9 (reference range 1.3–2.5); and γ -GGT: 79 U/L (reference range 11–50 U/L). Other biochemical indexes were roughly normal. The ferritin (FER), a tumor marker, was 1097.55 ng/mL (reference range 21.80–274.66 ng/mL). Other tumor markers, such as the carcinoembryonic antigen, the carbohydrate antigen 125, the total prostate-specific antigen (TPSA), the carbohydrate antigen 19-9, and the alpha-fetoprotein, were normal.

The colonoscopy showed a large mass with a hard touch in the colonic lumen near the spleen. A deep biopsy was difficult to conduct because yellow mucoid matter and stools were on its surface, and lilac vascular tissue could be observed. The colonic lumen in the mass area was obviously narrow, and it was difficult to pass an endoscope through it. The mass was considered to be a colon tumor with incomplete colonic obstruction. Plain and enhanced CT scans of the abdomen showed that there was a large, dumbbell-shaped, soft-tissue mass of 4.1×9.3 cm in the curved lumen of the descending colon near the spleen (Fig. 1). The enteric cavity was obviously narrow, and the proximal intestinal canal was obviously expanded. After enhancement, the lesion presented with progressive uneven

enhancement. The boundary between the lesion and parts of the left kidney and spleen was obscured. A small amount of exudation was observed around the lumen, and a slightly enlarged lymph node shadow was observed in the mesangial gap.

After admission, each preoperative examination was completed to exclude contraindications of the surgical procedure. A radical operation of expanded colon tumors, a splenectomy, and excision of the left kidney were carried out under general anesthesia. During the operation, a dumbbell-shaped mass was observed on the left colon region that varied widely from the splenic flexure of colon, where the mass was immobile with a relatively hard texture, to the lower descending colon, where the mass was movable with a slightly tough texture. The proximal transverse colon was aerated and expanded significantly, and no swollen lymph nodes were felt on the mesentery. There was obvious adhesion between the mass and the spleen bottom and left kidney, which were inseparable. The tumor had invaded the spleen and the left kidney. The intraoperative frozen section examination indicated that a malignant tumor of the colon and mesenchymoma should be considered first. Symptomatic and supportive treatment was given after the operation, and postoperative recovery was good. Section specimens showed that the tumor was a dumbbell-shaped mass with a hard texture and a complete mucosa near the splenic flexure of the colon and had invaded the spleen and the left kidney. The mass in the descending colon was movable with a slightly tough texture and a complete mucosa and had a lot of necrotic tissue and dull-red blood clots in it. The immunohistochemistry results were calponin (+); desmin (+); smooth muscle actin (SMA) (dispersive +); vimentin (+); CKpan (focal +); CD117 (–); CD34(±); Ki67 (the positive rate was low); and S-100 (–). During the postoperative pathology, an epithelioid inflammatory myofibroblastic sarcoma was diagnosed (Fig. 2). Approximately 3 months after the operation, a CT scan of the abdomen showed adrenal metastases (Fig. 3). The patient had traditional Chinese medicine treatment. The tumor shrank in a short time but later enlarged. One and one-half years after the operation, a plain magnetic resonance imaging (MRI) scan indicated obvious metastases in the left middle abdomen and the left adrenal gland (Fig. 3).

DISCUSSION

Epithelioid inflammatory myofibroblastic sarcoma was first identified by Marino-Enriquez et al in 2001.⁷ The name reflects its distinct biobehavior characteristics of malignancy.⁷ Only a few studies in English have been reported in the

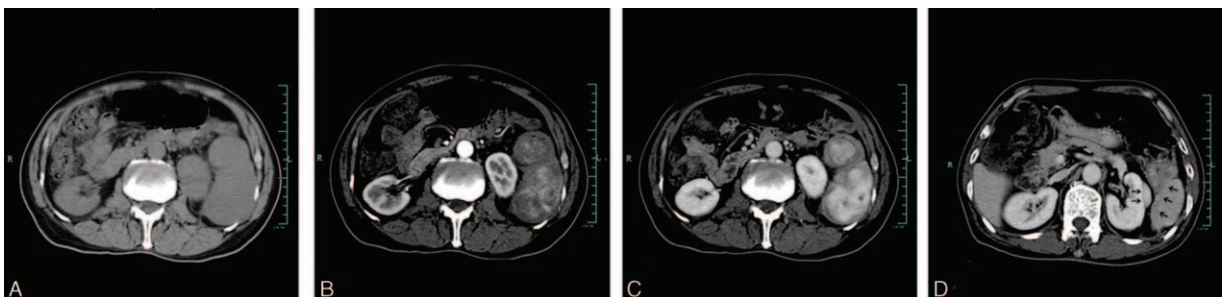


FIGURE 1. Abdominal CT shows a large stripe-like mass in the lumen of the descending colon with obvious lumen stenosis. The plain scan shows homogeneous density of tumor, and the CT value is ~ 28.6 HU (A). Enhancement scanning shows heterogeneous enhancement in the arterial phase and apparent enhancement in the later phase, with CT values of 75.8 HU (B) and 148.6HU, respectively (C). The boundary between the lesion and the left kidney and spleen was obscured (arrow, D). CT = computed tomography.

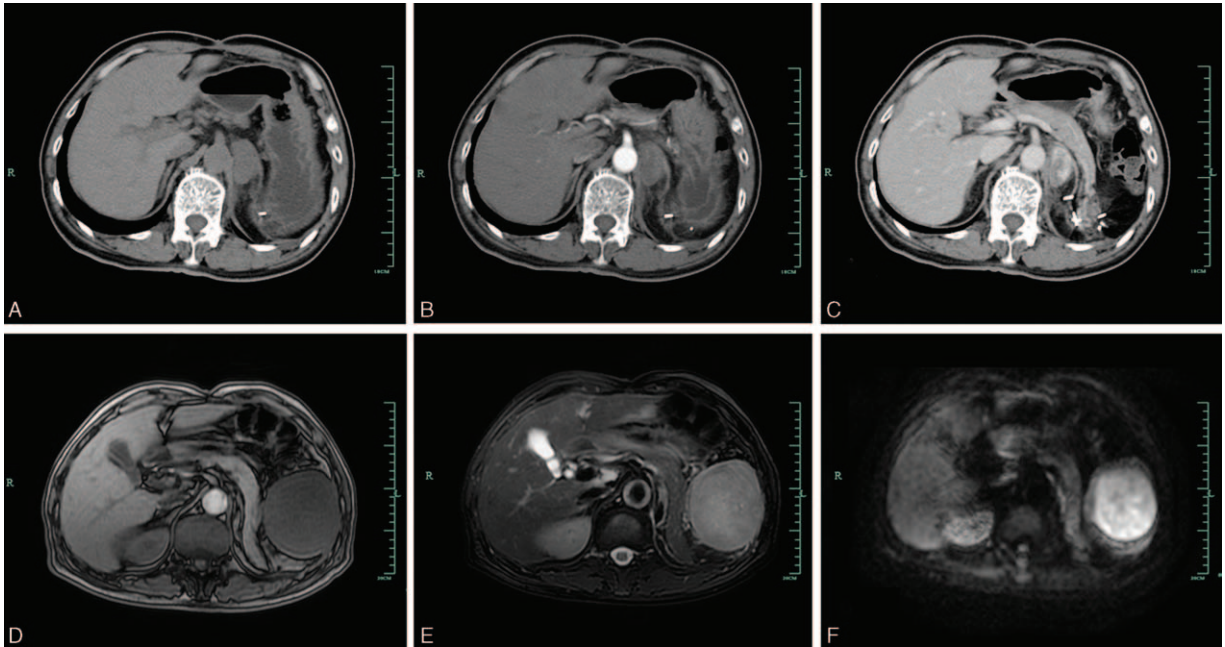


FIGURE 2. Histopathology shows abundant tumor cells, some of which are epithelioid; obvious nucleoli; and multiple inflammatory cells that can be observed in the tumor background (A). (hematoxylin eosin staining_400) Positive SMA staining of tumor cells (B) and positive Calponin staining of some tumor cells (C).

literature to date.⁷⁻¹² The age of onset of extrapulmonary IMT ranges from 3 months to 46 years, with a median age of 9 years and a male–female ratio of 3:4.⁴ Unlike extrapulmonary IMT, the age of onset of EIMS ranges from 7 months to 63 years, with a median age of 32 years, and the majority of cases occur in men.⁷⁻¹² Among the different occurrence sites of extrapulmonary IMT, EIMS mostly occurs in the abdominal cavity, with a majority of cases found in the mesentery or omentum. There have been 2 reported cases in the chest, with one in the lung and the other in the chest wall.⁷⁻¹² The clinical presentation and the laboratory examination of EIMS lack specificity. The patient in our case was an older man. Though EIMS mostly occurs in the abdominal cavity, this is the first time that colon EIMS has been reported to our knowledge.

EIMS is a type of variable IMT, and its pathologic features include⁷ the tumor being composed of round-to-epithelioid cells with large vesicular nuclei and amphophilic-to-eosinophilic

cytoplasm; abundant myxoid stroma; a high mitotic rate; and immunohistochemistry showing positive ALK, desmin, CD30 and focal smooth muscle actin, but negative EMA, caldesmon, MYF4, S-100, and keratins. Tumors pathologically identified as EIMS include anaplastic large cell lymphomas, high-grade epithelioid leiomyosarcoma, rhabdomyosarcoma, and undifferentiated sarcoma.⁷ IMT was once regarded as a type of relatively benign tumor-like lesion; however, the malignant biological behavior of some cases was noticed and reported in the literature.^{13,14} The clearly atypical polygonal, round, or spindle cells, as well as the vesicular and large nuclei, are believed to be connected with the vicious transformation of IMT.¹⁴ It is not difficult to discover that there is consistency between the cytology of the high-invasive IMT cases described previously and that of EIMS. ALK expression, which is found in 36% to 60% IMT cases, is attributed to ALK gene rearrangement at 2p23.¹⁵ ALK fusion oncogenes, including TPM3,

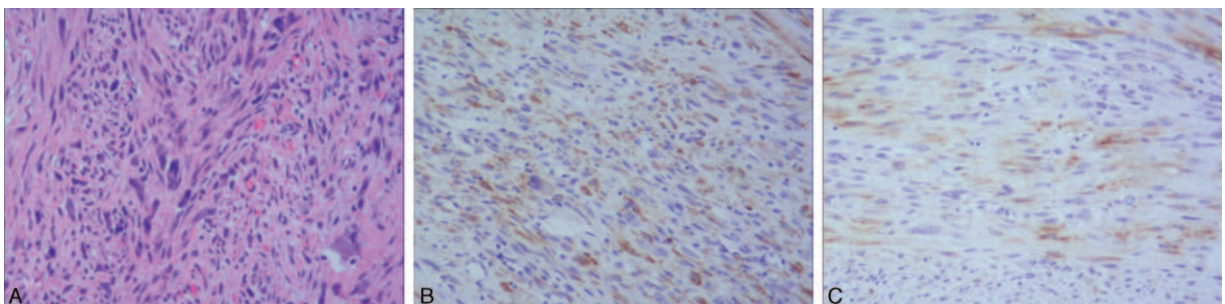


FIGURE 3. Abdominal CT shows an oval soft-tissue mass with a clear boundary and distinct heterogeneous enhancement in the left adrenal area (A–C). A circular abnormal signal is observed in the spleen, with a low T1 W1 signal (D), a slight increase in the T2 W1 signal (E), and heterogeneous signal intensity and an obvious high diffusion-weighted imaging signal (F). CT = computed tomography.

CLTC, TPM4, RANBP2, SEC31 L1, CARS, and ATIC, have been verified in some IMT cases.¹⁶ Coffin et al discovered when researching a set of 59 IMT cases that tumors were larger in the abdominal and pelvic cavities and were likely to recur.¹⁷ They also thought that ALK reactivity was related to recurrent IMT but had nothing to do with metastases.¹⁷ However, in a set of 11 EIMS cases, ALK was positive, and the cases were characteristic of early recurrence, with 2 of them developing distant metastases.⁷ In the reports of another 2 individual EIMS cases, one developed liver and lung metastases from the mesentery, and the other developed bone metastasis from the lung.^{11,12} Therefore, we think that ALK expression cannot exclude the possibility of distant metastases in IMT, especially for EIMS. Li et al focused on IMTs with RANBP2-ALK gene rearrangement.¹⁸ The occurrence sites and histological and immunohistochemical features of these cases were similar to those of EIMS. There are some overlapping cases that present with apparent invasiveness.¹⁸ Therefore, it is necessary to perfect the diagnostic criteria of EIMS to differentiate it from conventional IMT or other types of IMT.

The imaging features of IMT occurring in different sites are summarized, although they lack specificity.¹⁹ The imaging features of lung IMT include a single, peripheral, well-defined mass that mostly locates in both lower lungs.^{19,20} Lung IMT shows heterogeneous enhancement and may be accompanied by calcification.^{19,20} Similarly, 1 case of lung EIMS occurred in the lower lung with a lobulated and heterogeneously enhanced mass.¹² However, the lesion in that case was larger, and multiple tumor blood vessels could be observed in the lesion. One case of EIMS in the chest wall showed rapid enlargement of the chest wall mass, which was irregular in shape and accompanied by apparent heterogeneous enhancement.⁹ Therefore, EIMS that occurs in the chest is usually larger and has heterogeneous enhancement. Imaging is likely to confirm the diagnosis of malignancy, but characteristic imaging findings are often absent. The imaging findings of IMT in the mesentery or omentum have been reported and mostly occur in children or teenager.^{21–23} A large well-defined or ill-defined mass can be observed; the lesion infiltrates peripherally and involves the adjacent intestinal tube.^{21–23} Calcification can be observed in some cases with necrosis in the center.^{21–23} Enhancement scanning shows varied enhancement methods of IMT in the mesentery or omentum, such as low enhancement, heterogeneous enhancement, or edge enhancement.^{21–22} Only individual cases of the imaging findings of EIMS in the abdominal cavity have been reported.^{8,11} The lesion is larger, likely involves adjacent intestinal tubes, and is mass-like, well-defined, and heterogeneously enhanced.^{8,11} EIMS in our case occurred in the descending colon. The lesion was larger and showed heterogeneous enhancement, which is similar to the reports above. The difference is that the lesion in this case was mostly located in the lumen, causing lumen stenosis. Meanwhile, the extra-luminal growth of the tumor invaded the peripheral organs and showed distinct invasiveness. In addition, the colon EIMS in this case had a dumbbell shape and needed to be identified with neurofibroma. Neurofibroma in the intestinal walls is a part of neurofibromatosis and may present as a tubular mass with a smooth border, homogeneous density, and homogeneous or heterogeneous enhancement.²⁴ However, the enhancement degree of neurofibroma is lower, which is different from the colon EIMS in our case, which showed distinct enhancement in the later phase of enhancement scanning. The imaging of intestinal EIMS should be identified with other lesions, such as stromal tumor, lymphoma, and soft tissue

sarcoma. Moreover, preoperative and postoperative imaging evaluations of EIMS should note that multiple lesions could be present.⁷

The treatment of EIMS is generally surgical resection of the mass supplemented by chemotherapy.⁷ The tumor is likely to recur soon after surgery and can be treated by a second resection.⁷ Some cases may develop distant metastases, including liver, lung, bone, and lymph nodes.^{7,11,12} Short-term death occurs in some cases.^{7,10–12} Biologically targeted ALK inhibitors have been used in some EIMS cases reported in the literature, among which 2 benefitted and 1 obtained no definite benefit and death occurred a short time later.^{7–9,12} Therefore, ALK inhibitors may be a new therapeutic choice for EIMS, especially for recurrent cases or those cannot be treated with surgical resection. However, their indications and curative effects need to be further explored.

CONCLUSION

We reported on a rare case of colon EIMS in which the endo-luminal growth of the lesion resulted in colonic obstruction whereas the extro-luminal growth invaded the adjacent organs. The lesion showed heterogeneous enhancement. When endo/extro-luminal growth of intestinal tumors, though rare, is encountered, EIMS should be included in the differential diagnosis. Surgical resection of EIMS is the preferred treatment, whereas ALK inhibitors are a potential alternative option.

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