



# A Rare Etiology of Small Intestinal Obstruction - Solitary Myelogenic Sarcoma of the Small Intestine: CT Findings and a Review of the Literature

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**Abstract:** Background: Myelocytic sarcoma is a rare extramedullary tumor consisting of immature myeloid cells-granulocytes, mononuclear cells, or both. It usually occurs in patients with acute or chronic myeloid leukemia and is often found in the skin, bone, and lymphatic tissues. Isolated myeloid sarcomas are more uncommon in patients without leukemia in the small intestine. Case presentation: In the present study, a hospitalized case is discussed with intestinal obstruction as the primary symptom. During the CT examination, the local intestinal wall of the jejunum was found to possess ring thickening, intestinal lumen stenosis and proximal intestinal obstruction. The tumor presented with uniform and moderate progressive enhancement. The patient underwent emergency laparoscopic surgery in order to remove the tumor. Immunohistochemical staining on postoperative paraffin sections revealed myelosarcoma. The patient also underwent a bone marrow biopsy to exclude acute and chronic myeloid leukemia and the bone marrow smear indicated normal range. Conclusions: Myeloid sarcoma must be included in the differential diagnosis of small intestinal tumors, although the patient may not have history of leukemia. Radiologists should improve their imaging perception of myelosarcoma.

**Keywords:** Myeloid Sarcoma, Intestinal Obstruction, Tomography, X-ray Computer

## 1. Introduction

Myelocytic sarcoma (MS) is a rare extramedullary malignancy consisting of progenitor granulocytes or immature myeloid cells with an incidence of approximately 2.5-2.9% worldwide [1-3]. The onset age between children and elderly patients varies [4]. Typically, this disease occurs during the development of acute myeloid leukemia, or chronic myeloid leukemia or during the progression of myelodysplastic syndrome [1-3]. A few cases can also present with peripheral blood or bone marrow disease [1-3]. Extramedullary myeloid cell sarcoma usually occurs in the skin, bone and lymphoid tissues [5-8] and rarely in the small intestine [4]. We present a case of isolated MS in the small intestine with no history of malignancy.

## 2. Consent

The patient agreed to allow the use of her data for writing, teaching and publication purposes.

## 3. Case Report

A 41 year old female patient was admitted to the hospital for more than 2 weeks following repeated defecation. The patient did not have previous history of "blood disease". The physical examination indicated upper left abdomen tenderness and no rebound pain. Specific tumor markers were measured and they exhibited the following values: cell keratin fragment 19: 4.58 ng/ml (normal range 0-3.3), carbohydrate antigen 125 (CA125): 136.2 U/ml (normal

range 0-35). No abnormality was found in the other laboratory biomarkers tested.

The orthostatic plain abdominal film indicated signs of intestinal obstruction. The abdominal CT tip center of the right ileum indicated partial annular thickening of the bowel wall and lumen stenosis, whereas the thickness and length were approximately 13 mm and 35 mm, respectively. The progressive relatively homogeneous enhancement scan CT value was 51, 76 and 90 HU (hounsfield unit) for the arterial venous phase, the bowel wall and the mesangial side, respectively. The latter two exhibited thickening and increased fat density. In the mesenteric area, multiple enlarged lymph nodes were noted along with proximal ileal apparent effusion, expansion and obstruction of the plane appearing as "waste" sign (Figure 1).

The laparoscopic surgical exploration revealed an intestinal mass 3 meter from the troxton ligament, forming a narrow ring, whereas extensive proximal intestinal dilatation,

intestinal wall edema and distal intestinal and colon tumors were also present. "Small intestine partial resection and peritoneal adhesion release (including peritoneum and omentum)" were performed.

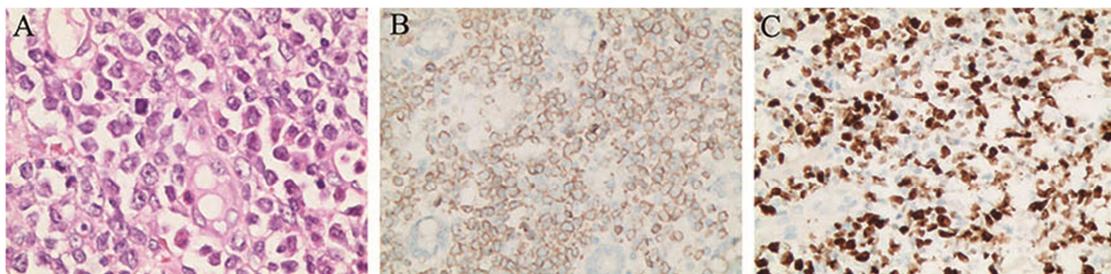
The pathological analysis was performed on the excised specimens. Microscopically, a large number of round and ovoid tumor cells were noted that infiltrated the entire intestinal wall. Immunohistochemical staining indicated that 80% of Ki67 cells were expressed by tumor cells, whereas positive expression was noted for Bcl2, Vimentin, CD38, myeloperoxidase (MPO) and CD117 (figure 2). The final pathological diagnosis was MS.

Based on this diagnosis, bone marrow biopsy was also performed to exclude generalized acute and chronic myeloid leukemia. The bone marrow smear indicated normal range.

Following operation, the patient was treated with chemotherapy and followed up. The patient was in good condition and had no secondary malignant blood disease.



**Figure 1.** A: Local ileum wall thickening and intestinal cavity narrowing. Contrast homogeneous enhancement (white arrow) also demonstrates obstruction, plane proximal small intestine effusion, dilation and fecal signs (red circle); B: Local ileum wall thickening, intestinal cavity narrowing and contrast homogeneous enhancement (white arrow); C: Local ileum thickening, homogeneous enhancement (white arrow).



**Figure 2.** A: Microscopically, a large number of round and ovoid tumor cells can be seen infiltrating the entire wall of the intestine (H&E staining 40X); B: Immunohistochemical staining: positive for myeloperoxidase (MPO) (Immunohistochemistry 20X); C: Approximately 80% of Ki67 positive tumor cells were noted (Immunohistochemistry for Ki67 20X).

## 4. Discussion

Although the small intestine accounts for 90% of the gastrointestinal tract, small intestinal tumors are uncommon, accounting for less than 5% of all gastrointestinal tumors [9]. Primary small intestinal malignancies account for approximately 2% of all gastrointestinal malignancies [10]. The proportion of isolated MS occurring in the gastrointestinal tract has been estimated to approximately 6.5%[11] and the ileum is usually prone to recurrence [12, 13]. Due to its rarity and difficulty in diagnosis, MS is often misdiagnosed as other diseases [14], with a misdiagnosis rate of 25-47% and the most common misdiagnosis being lymphoma [15-17]. The case reported in the present study involved a tumor that could not be misdiagnosed. In the preoperative imaging report, MS was not listed in the differential diagnosis. When the tumor was explored by laparoscopy, the intraoperative frozen section was also diagnosed as lymphoma and the immunohistochemical staining of the paraffin sections following surgery led to the final diagnosis of MS.

MS, also known as "green tumor" or granulocytic sarcoma, was not named "MS" by the world health organization until 2002 [18]. MS is most commonly seen in patients with malignant blood diseases [3, 8] and is present in approximately 1-2% of patients with acute myeloid leukemia [1-3, 7].

50% of MS cases are asymptomatic or produce nonspecific manifestations of organ dysfunction or pain related to the mass effect at the affected site. Isolated small intestinal MS is often treated with intestinal obstruction as the first symptom [19]. The majority of the complications include hemorrhage, perforation, necrosis, intussusceptions and obstruction. In the present case, the patient repeatedly stopped venting and defecating for 2 weeks and was accompanied by upper abdominal pain, mild tenderness and no symptoms of fever, anemia, enlarged lymph nodes or any other type of leukemia, which resulted in a considerably difficult clinical diagnosis. Abdominal CT can only indicate local ileal wall ring thickening, intestinal lumen stenosis, progressive homogeneous moderate enhancement, ileal proximal obstruction and enlarged lymph node shadow in the area of the superior mesenteric artery. In the current study, it was found that the CT manifestations of MS were mostly equal to the density of the lesions, with moderate enhancement

following enhancement and additional homogeneous enhancement compared with heterogeneous enhancement [8, 20, 21]. The CT findings of this case are consistent with those reported in the literature to a large extent. However, some of these clinical signs are not considered a unique sign of MS and similar imaging findings have been reported in inflammatory lesions and lymphoma [22, 23].

Shinagare *et al* [24] described the magnetic resonance characteristics of 25 patients with 41 different MS localization patterns. The T1-weighted images indicated that the percentage of isosignal cases was 75.6% and that of low signal cases 24.4%. In the T2-weighted images, the high signal cases corresponded to 95.1% and the equal signal cases to 4.9%. In another study different results were reported. Meyer *et al* [20] performed magnetic resonance imaging (MRI) on 28 cases of MS in different parts and found that 60.7% of the T1-weighted images were isosignals, while 82.1% of the T2-weighted images were high signals. The enhancement degree of MRI is similar to that of CT. The imaging findings of CT and MRI are not characteristic of MS, which undoubtedly increases the diagnostic difficulty of this disease.

It has been reported that 18F-fluorodeoxyglucose-positron emission tomography (FDG-PET/CT) is highly sensitive to the diagnosis of extramedullary MS [25-27]. MS has a moderate to high level of glucose intake for FDG, which can be used to provide improved diagnosis for this disease and also assess the situation prior to and following treatment. This can address queries involving the recurrence of the tumor as well as its reduction or enhancement. This is considered an optimal diagnostic method. However, it is not particularly cost-effective and increases the exposure of the patient to radiation. Therefore, it is necessary to identify improved methods to diagnose MS using CT and MR.

We also collected the case reports and comprehensive reports of CT findings of myelosarcoma from 2002 to 2020 for retrospective analysis. The high risk leukemia patients in the small intestine of marrow cell sarcoma are presented with summarized clinical characteristics and CT imaging findings. The data indicated 12 cases (4 female, 8 male) involving leukemia with MS, of which 10 were small intestine isolation MS cases. A total of 8 cases included the jejunum and 6 the ileum (table 1).

**Table 1.** Clinical and CT features of myelogenic sarcoma from literatures of review.

Study	age/sex	Chief complaint	CT finding			Presentations of lymph nodes
			Location and presentation of lesions	Enhanced Scanning	Changes of lumen	
Palanivelu <i>et al</i> [28]	52/male	Abdominal distention and Pain with a high fever	Mass in the jejunum	Homogeneous moderately enhanced	Narrow bowel cavity and proximal intestinal dilatation	ND
Kumar <i>et al</i> [19]	55/female	Left lower abdominal pain	The wall of the jejunum thickened irregularly with a mesenteric mass	ND	Narrow bowel cavity and proximal intestinal dilatation.	ND

Study	age/sex	Chief complaint	CT finding			
			Location and presentation of lesions	Enhanced Scanning	Changes of lumen	Presentations of lymph nodes
Yoldaş et al [29]	44/male	Abdominal pain	Ileum wall thickened irregularly	Homogeneous moderately enhanced	proximal intestinal dilatation	ND
Lee et al [30] 45/male	45/male	Intermittent abdominal cramps	Ileum mass with ileum intussusception,	Homogeneous moderately enhanced	ND	Multiple enlarged lymph nodes in the mesenteric root and perimesenteric vessels
Cicilet et al [31]	45/female	Severe abdominal pain and vomiting	Eccentric focal ileum wall thickening	Homogeneous moderately enhanced	proximal intestinal dilatation slightly	ND
Wang et al [32]	23/male	Intermittent upper abdominal pain	The wall of the jejunum was thickened in concentric circles	Homogeneous moderately enhanced	ND	The adjacent mesenteric soft tissue mass wrapped the branch of the superior mesenteric artery
Aslan et al [33]	57/female	Abdominal pain, nausea, vomiting and constipation	The wall of ileum was thickened	Homogeneous moderately enhanced	proximal ileum significantly expanded	Multiple small lymph nodes adjacent to mesenteric vessels
Mizumoto et al [34]	54/male	Abdominal pain and vomiting	The wall of jejunum was thickened	ND	Narrow bowel cavity and proximal intestinal dilatation.	A number of small lymph nodes were noted beside the mesenteric vessels.
Plowman et al [35]	41/female	Persistent abdominal pain and constipation	The wall of the jejunum was thickened to the annular area	Homogeneous moderately enhanced	Narrow bowel cavity and proximal intestinal dilatation.	ND
Singh et al [22]	41/male	Abdominal pain and bloating	Jejunum ring thickened	enhanced homogeneity	proximal intestinal dilatation	mesenteric roots with apparent vascular submasses
Choi et al [23]	38/male	Abdominal pain and diarrhea	An ectogenic mass of ileum with thickened intestinal wall,	Homogeneous moderately enhanced	proximal intestinal dilatation	Enlarged mesenteric lymph nodes
Choi et al [23]	48/male	Diffuse abdominal pain	The wall of the ileum was thickened and formed a mass	Homogeneous moderately enhanced	ND	Adjacent enlarged lymph nodes of intestine and mesangial.

SET: ND=no data.

The table indicated that the clinical manifestations of the small intestinal solitary MS in non-leukemia patients were basically abdominal pain and abdominal distension caused by complete or incomplete intestinal obstruction, while the CT manifestations included intestinal lumen stenosis caused by intestinal wall thickening. Contrast enhancement was mainly homogeneous enhancement and mesenteric lymph node enlargement. This was not consistent with the CT manifestations of MS found by Ooi [21] and Meyer et al. [20], which may be due to the MS involving multiple parts of the whole body. Although all the cases collected by Choi et al. [23] were from the gastrointestinal tract, they were mixed with myeloid sarcomas caused by non-leukemia patients. Therefore, it was impossible to analyze and summarize the CT signs of the myeloid sarcomas caused by non-leukemia patients. The 12 cases in table 1 were all in the small intestine and were all non-leukemia patients. The aforementioned data were combined with our own cases and the conclusions led to the speculation that

the CT manifestations of isolated small intestinal myeloid cells in non-leukemia patients were the thickening of intestinal wall and the tumor formation. Moreover, we deduced that in such cases, the intestinal cavity was narrow and that the expansion of the proximal small intestine was readily caused, whereas the mesenteric lymph nodes were also affected. The contrast enhancement was mainly homogeneous enhancement. The imaging signs appeared to be different from the CT findings of patients with inflammatory lesions and lymphoma. The latter requires more cases to support our hypothesis.

## 5. Conclusion

Isolated small intestinal MS is a rare malignant tumor, which usually appears with intestinal obstruction as the first symptom. The patients who have never had a malignant blood disease are considered a major diagnostic challenge. To

improve radiologists' awareness of isolated small intestinal MS, MS must be used as the differential diagnosis of small intestinal tumors. There were some limitations in this literature. Firstly, a part of case reports were conference papers so we could not get detail, so we had to give up, and secondly we did not make a clinical or radiological comparison with the articles about the MS patients with myeloid leukemia.

## Compliance with Ethical Standards

This study was approved by the ethics committee of Guangdong Hospital of Traditional Chinese Medicine.

## Conflict of Interest

The authors declare that they have no conflict of interest.

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