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## Case Report

# De novo myeloid sarcoma as a rare cause of small bowel obstruction: GT findings and histopathologic correlation

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## ABSTRACT

Small bowel obstruction caused by myeloid sarcoma in a patient with any hematological abnormality is very rare. Myeloid sarcoma occurs most commonly in patients with acute myelogenous leukemia (AML) and less with other hematological disorders. A 57-year-old female presented with abdominal pain, nausea, vomiting, and constipation. Radiological studies showed concentric bowel thickening in distal ileum that caused nearly total luminal compromise and signs of obstruction in proximal ileal bowel loops. She underwent laparotomic surgery and ileal resection was done. Diagnosis of myeloid sarcoma was made by histopathological examination of surgical specimens. Bone marrow biopsy was done to rule out systemic acute myelogenous leukemia (AML). Results of bone marrow biopsy were within normal limits. Finally, the patient was diagnosed as de novo myeloid sarcoma. Although the histopathological examination makes a definitive diagnosis, imaging allows to locate the lesion, evaluate its complications, and guide for correct biopsy. Accurate diagnosis of myeloid sarcoma has important prognostic value as transformation to AML can happen without chemotherapy or stem cell transplantation.

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## Introduction

Myeloid sarcoma is one of the rarest malignant solid tumors. It is an extramedullary solid tumor composed of myeloid precursor cells. It is also known as granulocytic sarcoma. The tumor was also known as a “chloroma” because of a

characteristic greenish color noted on exposure to air that is caused by high levels of the enzyme myeloperoxidase within immature myeloid cells in initial case reports [1].

It occurs most commonly in patients with acute myelogenous leukemia (AML) and less with other hematological disorders such as myelodysplastic syndrome, chronic myelogenous leukemia, or other myeloproliferative diseases. It

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also can occur as a sign of AML in a nonleukemic patients, as a sign of blast crisis that about to happen in chronic myelogenous leukemia or as an isolated manifestation etc. [2,3].

Although myeloid sarcoma virtually can appear in any part of body, it is most commonly seen in skin, soft tissues and bones [4]. In our case report we present a patient, without any hematological disease history, had an obstructive lesion in small bowel on imaging that pathologically proven as myeloid sarcoma after surgical resection.

## Case report

A 57-year-old woman with no relevant medical history presented with abdominal pain, nausea, vomiting, and constipation. On physical examination, she was found to have tenderness on umbilical region and right lower abdominal quadrant. Results of blood parameters were within normal limits except LDH which was elevated (LDH: 327 U/L. Normal range: 0248 U/L). As clinicians put preliminary diagnosis of acute abdomen, the patient was sent to computed tomography (CT) examination. Intravenous (IV) contrast-enhanced CT of the abdomen was performed. It revealed bowel wall thickening (mean 10 mm) involving nearly 3 cm of distal ileum with homogenous enhancement on post contrast images causing nearly total luminal compromise (Fig. 1). Obvious dilatation of proximal ileal loops with "feces sign" proximal to level of obstruction was seen (Fig. 2). A few minimally enlarged perilesional lymph nodes were detected on CT (Fig. 3). Differential diagnosis of lymphoma and small bowel carcinoma was thought with respect to imaging findings.

The patient underwent laparotomy and intraoperatively lesion was identified nearly 40 cm proximal to ileocecal junction. Resection was performed and the resected specimen was sent for pathological examination.

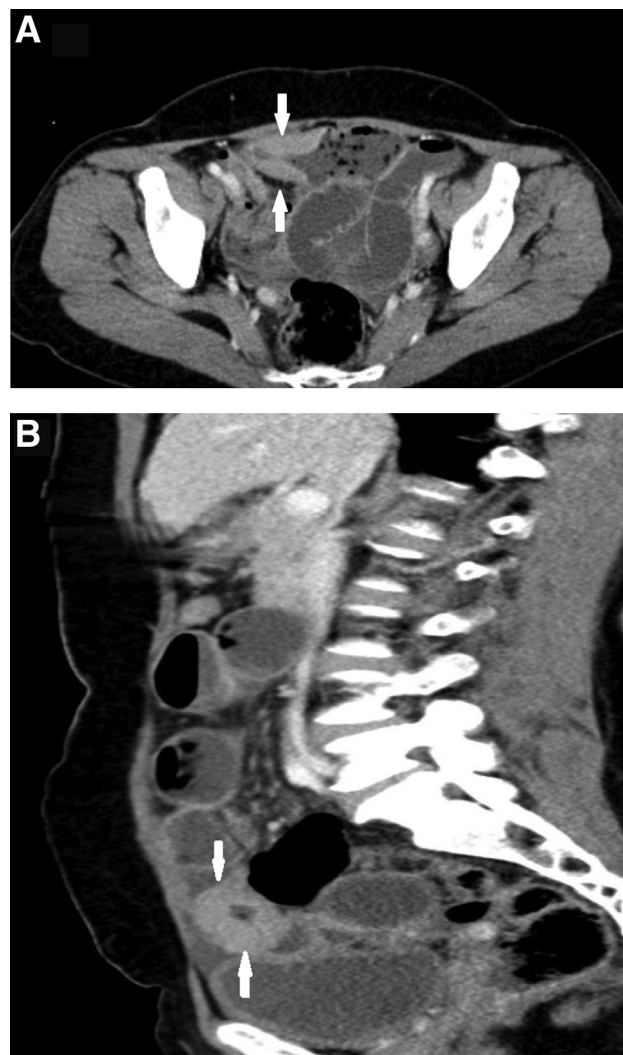
Resected small bowel specimen had a circular mass, narrowing the lumen and flattening but not ulcerating mucosal surface. Sections showed transmural white, solid tumor reaching serosal surface. No green or brown discoloration was observed at fresh or fixated specimen (Fig. 4).

Microscopic examination revealed diffuse infiltration of all layers of small bowel. Mucosal epithelia were intact. Proliferation was consisting of large pleomorphic cells with narrow eosinophilic cytoplasm and high nucleocytoplasmic ratio. Diffusely proliferating cells were discohesive and were not forming any architectural pattern. Apoptotic bodies and mitotic figures were evident (Fig. 5).

Immunohistochemical analysis showed these cells were negative for PANCK (AE1/AE3), S100 and were positive for LCA. Further evaluation revealed that these cells were in myeloid origin with diffuse MPO, Lysozyme, and CD43 positivity. CD117 showed variable expression, CD34 was negative (Fig. 6).

With the aid of this immunophenotype and blastic features of proliferating cells, the diagnosis of myeloid sarcoma was made.

Based on this diagnosis the patient underwent a bone marrow biopsy to rule out systemic acute myelogenous leukemia (AML). Results of bone marrow biopsy were within normal lim-



**Fig. 1 – Axial and sagittal contrast-enhanced CT scans of abdomen showing concentric focal bowel wall thickening with homogenous enhancement and nearly total luminal compromise (Arrows).**

its. Finally, the patient was diagnosed as de novo myeloid sarcoma.

## Discussion

Early diagnosis of myeloid sarcoma is important because the disease alter the treatment regimen. If patients without underlying hematologic disorder at the time of diagnosis are not treated with systemic induction chemotherapy, acute leukemia develops in up to 71% of them at a mean of 6 months after diagnosis of myeloid sarcoma [5]. However, if patients are treated with standard induction chemotherapy at diagnosis, there is lower probability of AML. Radiation therapy may also increase survival of patients [5,6].

Most of small bowel tumors are metastases (50%). Primary malignant lesions in decreasing frequency are



**Fig. 2 – Coronal reformatted contrast-enhanced CT image showing concentric bowel wall thickening in distal ileum (thick long arrows) causing luminal narrowing and dilatation of proximal bowel loops (thick short arrows) with clear “feces sign” (thin long arrow).**



**Fig. 3 – Axial contrast-enhanced CT image showing a minimally enlarged lymph node (arrow) proximal to bowel wall thickening.**

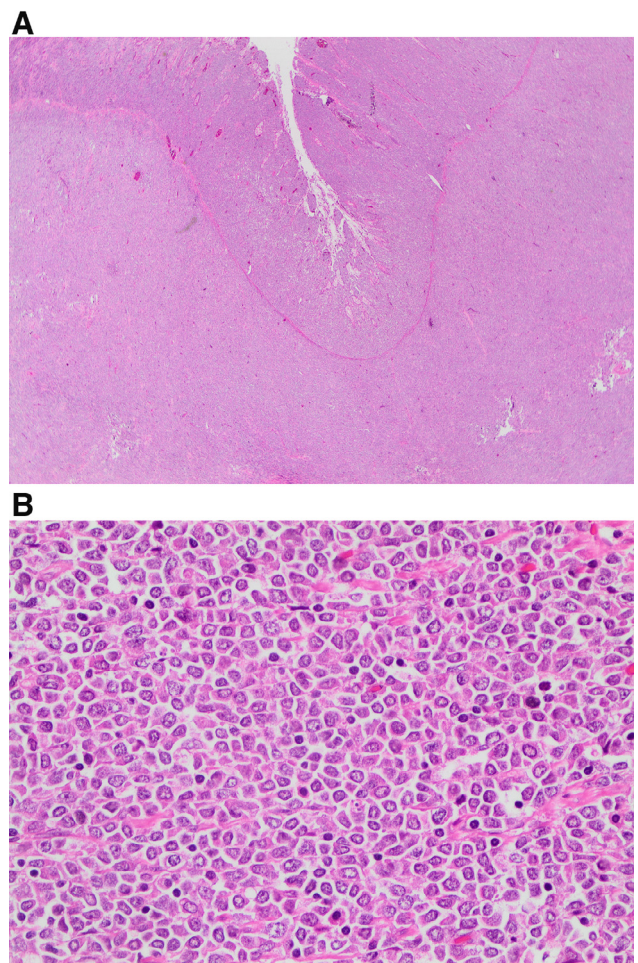
carcinoids (44.3%), adenocarcinomas (32.6%), lymphomas (14.7%), GISTs (7.2%), and sarcomas (1.2%) [7].

Myeloid sarcoma is generally extramedullary manifestation of AML and to a lesser extent of other myeloproliferative disorders. Isolated (or de novo) myeloid sarcoma is extremely rare (two in one million adults) [8]. Because of the extreme rarity and similar immunohistological pattern of this condition, frequently it is misdiagnosed as lymphoma [9].

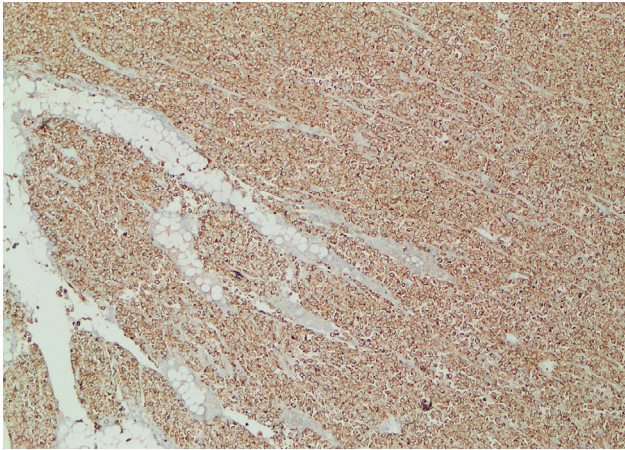
The most common affected tissues are skin, bones, lymph nodes, and soft tissues. On the other hand, gastrointestinal (GI) tract involvement is rare entity. In previous studies the



**Fig. 4 – Resected small bowel gross specimen; a circular mass reaching serosal surface of small bowel.**



**Fig. 5 – Diffuse infiltration of large pleomorphic cells with blastoid features; high nucleocytoplasmic ratio, narrow eosinophilic cytoplasm (20x, 400x, Hematoxylin-Eosin).**



**Fig. 6 – Proliferating cells were diffusely positive for myeloperoxidase (MPO) (100x).**

most common primary site of involvement in GI tract is small bowel, especially the ileum [3].

The CT features of myeloid sarcoma of small bowel and colon are variable. Lesions may appear as an intraluminal or exophytic polypoid mass, as bowel wall thickening or as a combination of these features. Patterns of contrast enhancement are also variable. Thus, myeloid sarcoma cannot reliably be differentiated with CT findings from lymphoma, other neoplastic conditions or inflammatory bowel diseases [10].

Although nearly 50% of intestinal myeloid sarcoma patients are asymptomatic, the most common complications of bowel myeloid sarcoma in the literature are hemorrhage, perforation, necrosis, obstruction and intussusception. There is also conspicuous CT feature of bowel myeloid sarcoma as it infiltrates peritoneum, omentum and mesentery with subsequent soft tissue mass formation [10].

## Conclusion

In conclusion, although the histopathological examination makes a definitive diagnosis, imaging allows to locate the le-

sion, evaluate its complications and guide for correct biopsy. Accurate diagnosis of myeloid sarcoma has important prognostic value as transformation to AML can happen without chemotherapy or stem cell transplantation.

It should be kept in mind that de novo myeloid sarcoma may cause small bowel obstruction and may mimic malignancies such as lymphoma and adenocarcinoma with imaging findings.

## REFERENCES

- [1] Miliauskas JR, Crowley KS. Granulocytic sarcoma (chloroma) of the small intestine with associated megakaryocytes. *Pathology* 1985;17(559):661–2.
- [2] Catalano MF, Levin B, Hart RS, Troncoso P, DuBrow RA, Estey EH. Granulocytic sarcoma of the colon. *Gastroenterology* 1991;100:555–9.
- [3] Corpechot C, Lemann M, Brocheriou I, et al. Granulocytic sarcoma of the jejunum: a rare cause of small bowel obstruction. *Am J Gastroenterol* 1998;93:2586–8.
- [4] Neiman RS, Barcos M, Berard C, et al. Granulocytic sarcoma: a clinicopathologic study of 61 biopsied cases. *Cancer* 1981;48:1426–37.
- [5] Imrie KR, Kovacs MJ, Selby D, et al. Isolated chloroma: the effect of early antileukemic therapy. *Ann Intern Med* 1995;123:351–3.
- [6] Tsimberidou AM, Kantarjian HM, Estey E, et al. Outcome in patients with nonleukemic granulocytic sarcoma treated with chemotherapy with or without radiotherapy. *Leukemia* 2003;17:1100–3.
- [7] Bilimoria KY, Bentrem DJ, Wayne JD, Ko CY, Bennet CL, Talamonti MS. Small bowel cancer in the United States: changes in epidemiology, treatment, and survival over the last 20 years. *Ann Surg* 2009;249(1):63–71. <https://doi.org/10.1097/SLA.0b013e31818e4641>.
- [8] Guermazi A, Feger C, Rousselot P, et al. Granulocytic sarcoma (Chloroma). *Am J Roentgenol* 2002;178(2):319–25. <https://doi.org/10.2214/ajr.178.2.1780319>.
- [9] Antic D, Elezovic I, Milic N, et al. Is there a "gold" standard treatment for patients with isolated myeloid sarcoma? *Biomed Pharmacother* 2013;67(1):72–7. <https://doi.org/10.1016/j.biopha.2012.10.014>.
- [10] Choi EK, Ha HK, Park SH, et al. Granulocytic sarcoma of bowel: CT findings. *Radiology* 2007;243:752–9.