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CASE REPORT | COLON

Colonic Myeloid Sarcoma as a Rare Presentation of Relapsed Acute Myeloid Leukemia

Parkpoom Phatharacharukul, MD[1], Nabil Fayad, MD[1], and Robert Siwiec, MD[1]

[1]*Division of Gastroenterology and Hepatology, Indiana University School of Medicine, Indianapolis, IN*

ABSTRACT

Myeloid sarcoma (MS), an extramedullary tumor of immature granulocytic cells, affects the gastrointestinal tract in approximately 10% of cases. MS involvement of the colon and rectum is considered to be extremely rare. We present a 36-year-old woman with acute myeloid leukemia and allogenic hematopoietic stem cell transplant 2 years before who was admitted with abdominal pain and nonmucous, nonbloody diarrhea. Colonoscopy revealed an ulcerated mass in the proximal colon, and biopsies showed MS compatible with acute myeloid leukemia relapse.

INTRODUCTION

Patients with hematologic malignancy who undergo bone marrow transplantation may develop various gastrointestinal (GI) tract system disturbances. Symptoms may include nausea, vomiting, abdominal pain, and diarrhea. In patients with diarrhea and findings of colitis, the differential diagnosis should include infection (bacterial, viral, and parasitic), acute or chronic graft-vs-host disease, medication side effects, and cancer relapse. We present a case of relapsed acute myeloid leukemia (AML) after bone marrow transplantation with GI tract manifestations.

CASE REPORT

A 36-year-old white woman presented to the emergency department with abdominal pain and nonmucous, nonbloody diarrhea of 1-week duration. Her medical history was significant for AML status postallogeneic hematopoietic stem cell transplant 2 years earlier, which was complicated by chronic graft-vs-host disease involving the skin, liver, and eyes. Her medications included prednisone 20 mg per day, sirolimus, mycophenolate mofetil, and ibrutinib. She denied any fever, chills, or sick contacts. Physical examination was pertinent for mild abdominal distention with generalized tenderness to deep palpation.

Abdominal and pelvic computed tomography with intravenous contrast did not show any mass or lymphadenopathy but was worrisome for new ascites, along with increased edema and wall thickening involving the ascending colon and proximal transverse colon (Figure 1). Thus, she was admitted for further evaluation. On admission, her hemoglobin was 7.3 g/dL and her white blood cell count was 12,400 cells/mL^3 without blast cells. Because of her immunocompromised status and colon wall thickening, colonoscopy revealed an ulcerated, friable mass with contact bleeding from the cecum to the midtransverse colon (Figure 2). Biopsies of the mass showed prominent diffuse proliferation composed of large immature cells with delicate chromatin and prominent nucleoli with frequent mitotic figures (Figure 3). Immunohistochemistry stains were positive for CD34 and myeloperoxidase and negative for PAX5 and CD3 findings compatible with myeloid sarcoma (MS).

Bone marrow biopsy showed a hypocellular marrow with no morphologic evidence of acute leukemia. However, ascites fluid cytology showed the presence of blast cells, with immunohistochemistry stains positive for CD7, CD33, CD34, CD38, CD71, CD117, and CD123 compatible with AML.

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Correspondence: Parkpoom Phatharacharukul, MD (pphatha@iu.edu).

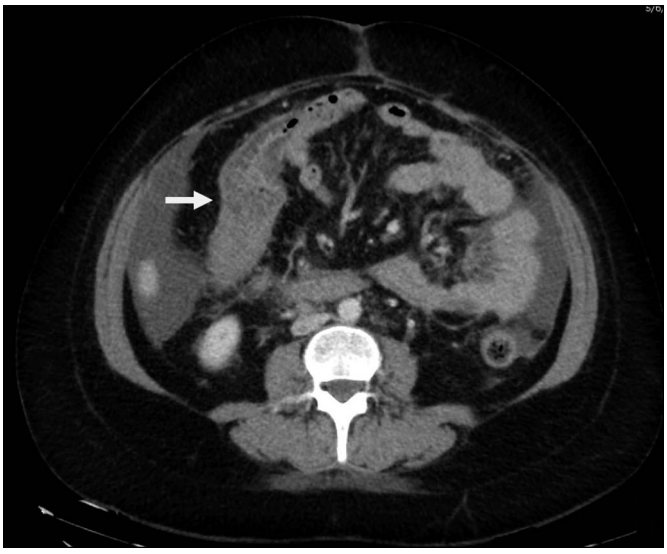

Figure 1. Abdominal and pelvic comuted tomography with intravenous contrast showed increased edema and wall thickening in the ascending and proximal transverse colon (arrow) and ascites.

After a multidisciplinary team discussion, given that her extramedullary relapse of AML conferred a poor prognosis, the patient was transitioned to hospice care and died 1 month later.

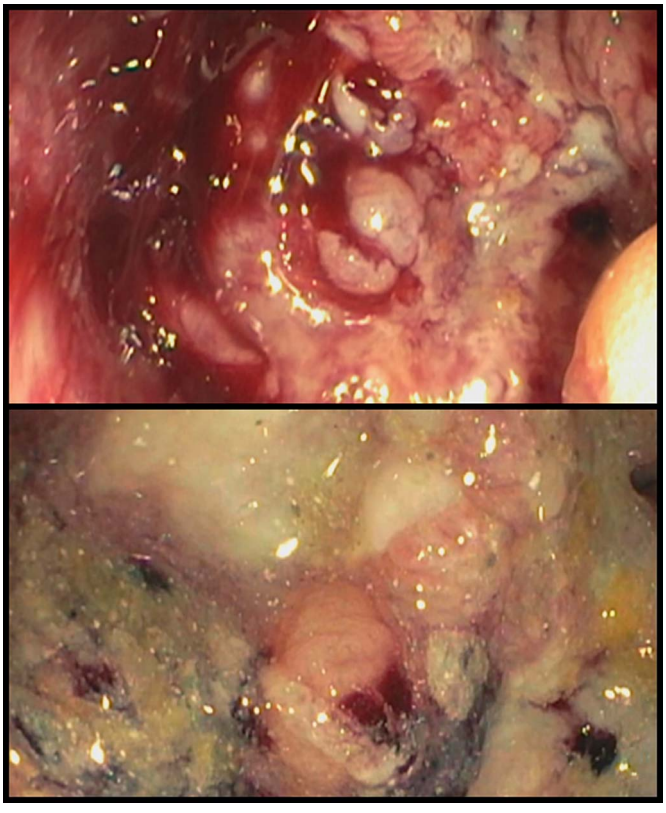

Figure 2. Colonoscopy revealed an ulcerated friable mass with contact bleeding in the cecum, ascending colon and transverse colon.

DISCUSSION

MS, also known as granulocytic sarcoma or chloroma, is an extramedullary tumor of immature granulocytic cells with an incidence of 2%–9% in patients with AML. In addition to AML, MS is also associated with myeloproliferative neoplasms, myelodysplastic syndrome, blast phase of chronic myeloid leukemia, and, in rare cases, in nonleukemic patients (primary or isolated MS).[1]

MS can occur at any age and has a slightly higher male predominance. It can involve any site of the body with the most common sites being lymphatic nodes, skin and soft tissues, testes, bone, and the peritoneum. The GI tract can be involved in approximately 10% of cases.[2] Colorectal MS is very rare with less than 20 reported cases in the literature.[3] Clinical presentations of MS in the GI tract are varied, ranging from asymptomatic to abdominal pain and constipation, bowel intussusception, hemorrhage, or diarrhea, as in our case.[3–5] The diagnosis requires a high index of suspicion because radiographic and endoscopic findings are nonspecific, including peritoneal or mesenteric infiltration on computed tomography scan and nodularity, friability, and focal ulceration on endoscopy. This commonly results in misdiagnosis in up to 25%–47% of patients because of inadequate immunophenotyping of the underlying MS lesion.[1]

Confirmation of the diagnosis is based on histology and immunohistochemistry profiling, including positive staining for myeloperoxidase, CD43, CD68, lysozyme, and CD117. Every patient diagnosed with MS should undergo a bone marrow biopsy to differentiate isolated MS from other forms of myeloid neoplasms, including AML or chronic myeloid leukemia, which have different management and prognosis.[1]

Treatment options depend on the tumor site and size, patient's age, performance status, and previous treatment for AML (if any). First-line treatment for MS is chemotherapy which could lead to complete remission in up to 65% of patients.[1] Appropriate patients should undergo evaluation for bone marrow transplantation. In addition to systemic chemotherapy, local therapy, including debulking surgery and/or radiotherapy, could also be offered to select patients who are symptomatic because of the mass effects of the lesions.[1] The prognosis for patients with MS is poor, with a median overall survival of 8 months. Interestingly though, a retrospective population-based study showed that patients with AML who had MS had a higher survival rate than patients with AML without MS.[2]

Colonic MS, although rare, should be on the differential diagnosis in patients with a history of hematologic malignancy presenting with GI symptoms and abnormal endoscopic or radiographic findings of the colon. Confirmation of the diagnosis requires histology with specific immunohistochemistry. Once the diagnosis is confirmed, the patient should be

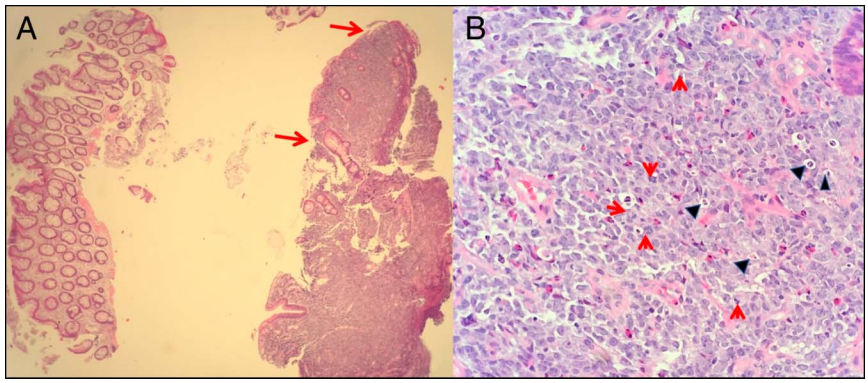

Figure 3. Biopsy of the mass found in the colon showed (A) a sharp contrast between unremarkable (left) and abnormal (right) mucosal fragments with surface erosions (arrows) (hematoxylin and eosin stain, 40× magnification) and (B) cytomorphologic features of malignant mucosal infiltrate to include increased nuclear to cytoplasmic ratios, delicate chromatin, and prominent nucleoli. Many atypical mitotic figures are present (arrows), and numerous apoptotic bodies are identified (arrowheads) (hematoxylin and eosin stain, 400× magnification).

referred back to hematologist/oncologist for further management and treatment.

DISCLOSURES

Author contributions: P. Phatharacharukul wrote the manuscript. N. Fayad and R. Siwiec edited the manuscript. R. Siwiec is the article guarantor.

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Informed consent was obtained for this case report.

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