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Small-bowel myeloid sarcoma: Report of a case with atypical presentation



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ABSTRACT

INTRODUCTION: Small-bowel myeloid sarcoma is rare. Acute bowel obstruction is its usual clinical presentation.

PRESENTATION OF CASE: We report a case of small-bowel myeloid sarcoma that occurred in a 64-year-old woman who presented chronic secretory diarrhoea, hypokalaemia, and weight loss. Immature white blood cells in a peripheral smear and small-bowel capsule endoscopic features were the main diagnostic clues. The patient experienced capsule retention and developed acute bowel obstruction. Urgent laparotomy showed a stricturing ileal mass and pathology of the resected bowel specimen unveiled a CD34+, CD117+, and myeloperoxidase-positive myeloid sarcoma. The diarrhoea promptly resolved after surgery, and the patient is now undergoing chemotherapy.

DISCUSSION: Secretory diarrhoea can be the first manifestation of small-bowel myeloid sarcoma. Capsule endoscopy may provide a diagnostic clue, but it can trigger an acute bowel obstruction. Differential diagnosis of the pathologic specimen may be difficult and a high suspicion index of is mandatory to perform immunophenotyping to determine the correct management.

CONCLUSION: Chronic diarrhoea with alarm features can be the first manifestation of small-bowel myeloid sarcoma.

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1. Introduction

Myeloid sarcoma (MS) is a rare malignant solid tumour composed of myeloblasts and immature/aberrant myeloid cells located outside the bone marrow.¹ Generally, MS predates the development of acute myeloid leukaemia (AML), although MS has been reported simultaneous to haematologic malignancy or during a relapse. Any organ or tissue can be affected by MS, and multifocal localisations have been reported.² In the largest published series to date, the gastrointestinal tract was involved in only 6.5% of cases, with a predilection for the small bowel.³ In this paper, we report an additional case of small-bowel MS with atypical clinical presentation.

Abbreviations: AML, acute myeloid leukaemia; MS, myeloid sarcoma; SBCE, small bowel capsule endoscopy.

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2. Case report

A previously healthy 64-year-old woman was admitted to our gastrointestinal unit for unexplained chronic diarrhoea and a loss of more than 10% of her usual body weight. Watery, non-bloody diarrhoea had started six weeks before admission. Blood analyses performed four weeks before admission showed mild normocytic anaemia, low serum albumin, and hypokalaemia. Total serum immunoglobulin A was normal. Anti-transglutaminase antibodies were negative, along with stool examination (including culture and a search for ova and parasites), oesophagogastroduodenoscopy with duodenal biopsies, and ileocolonoscopy with biopsies performed two weeks before admission. The patient appeared pale and thin, with dry skin and mucous membranes. Her abdomen was flat and soft, and deep palpation did not arouse pain or guarding. Her liver and spleen were not felt, and bowel sounds were present. A blood cell count showed $11.2 \times 10^9/L$ white blood cells, with 22% of immature cells in a peripheral smear. After fluid and lytes replacement, a bone marrow biopsy was performed, which disclosed immature cells: namely, 30% of blasts, CD45+, CD34+, CD117+, CD33+, CD13+, and HLADR+; normal karyotype (46XX);

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Fig. 1. Small-bowel capsule endoscopy frame showing a luminal stricture with thickened mucosa and short, swollen villi.

FLT3 0; and NPM: absence of mutations. A diagnosis of *de novo* AML, FAB M1 was made. As chemotherapy was not possible due to the persisting large volume of diarrhoea, small-bowel capsule endoscopy (SBCE) was performed (PillCam SB3, *Given Imaging*). One hour and twenty-two minutes after ingestion, the capsule did not

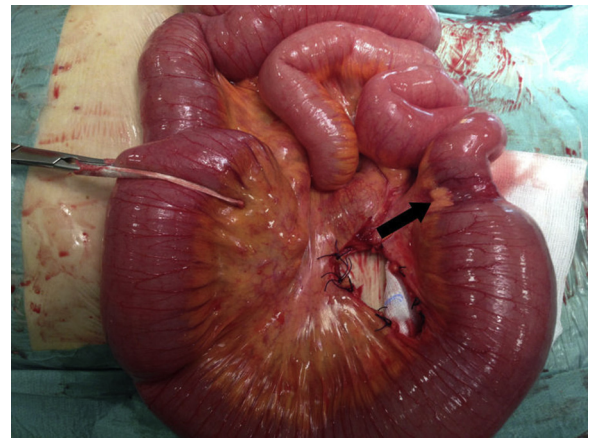


Fig. 2. Laparotomic finding of an annular, stricturing mass lesion of the ileum (arrow).

pass through an ileal stricture next to the dilated lumen, filled with luminal debris. The mucosa was thickened and pale, with short and swollen villi (**Fig. 1**). The following day, vomiting, abdominal pain, and distension ensued, and abdominal CT disclosed a stricture of the distal ileum next to the retained capsule, with proximal bowel dilation. On the 10th day of the hospital stay, laparotomy was performed. When the peritoneum was opened, a marked enlargement of the small bowel adjacent to an annular stenosis, 15 cm proximal to the ileocaecal valve, was found (**Fig. 2**). Segmental ileal resection with manual latero-lateral anastomosis was

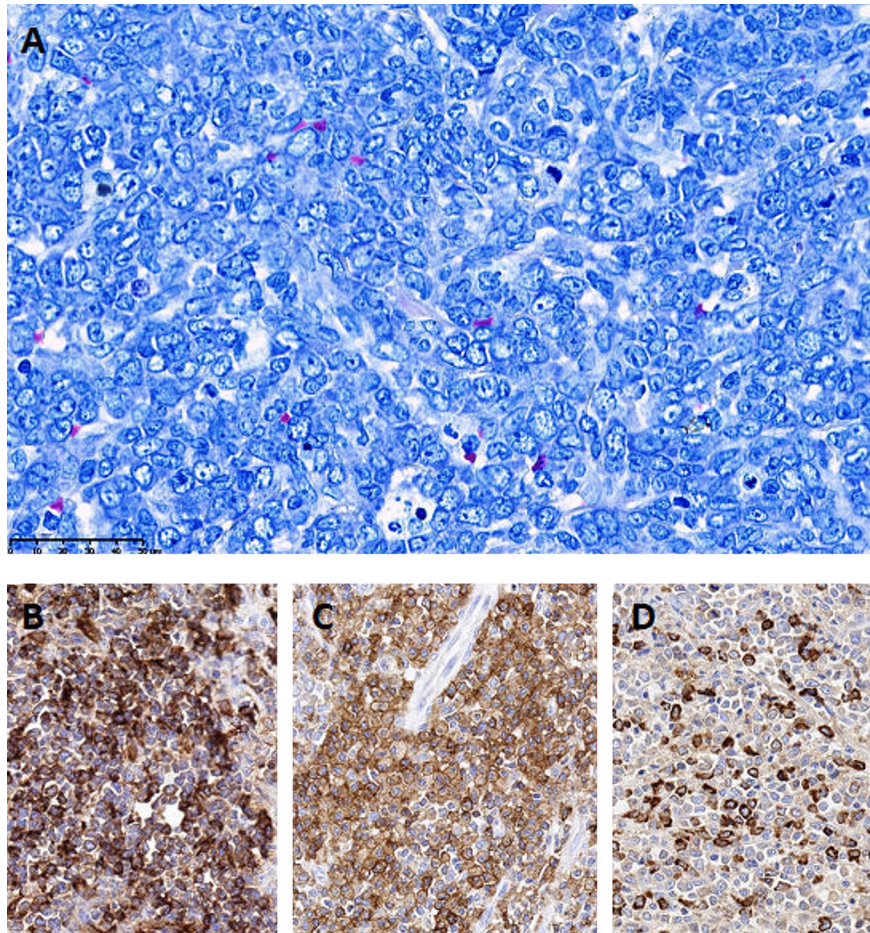


Fig. 3. Histopathology of the resected bowel specimen. (A) Diffuse infiltration by round, small- to medium-sized cells with moderate basophilic cytoplasm. The cells had round or oval folded nuclei containing dispersed chromatin and exhibited strongly positive staining for CD34 (B), CD117 (C), and myeloperoxidase (D).

performed. The still-flashing capsule was retrieved within the resected bowel. The postoperative course was uneventful, with early oral feeding, bowel canalisation, and full resolution of the diarrhoea. Pathologic examination of the resected specimen disclosed diffuse infiltration by round, small- to medium-sized cells with moderate basophilic cytoplasm. The cells had round or oval folded nuclei containing dispersed chromatin. The blastic features were compatible with very immature myeloid cells, and the cells were strongly positive for CD34, CD117, and myeloperoxidase, a pathologic picture diagnostic of MS (Fig. 3). At the time of writing, the patient was being treated with a combination of cytarabine and anthracyclines (3-plus-5 scheduled regimen), and stem cell mobilisation was obtained for eventual rescue treatment.

3. Discussion

MS is a tumour mass consisting of myeloid blasts occurring at an anatomical site other than the bone marrow. Infiltration of any site in the body by myeloid blasts in leukemic patients is not considered as MS unless it presents with tumour masses in which the tissue architecture is effaced. In the past, the gross greenish appearance that occasionally characterised this curious neoplasm was given the name “chloroma.” It is a rare condition; less than 1% of patients with AML will present with MS.⁴ MS most commonly consists of myeloblasts with or without features of promyelocytic or neutrophilic maturation. In a significant proportion of cases, MS displays myelomonocytic or pure monoblastic morphology. Regarding immunohistochemistry, MS has been well characterised, with CD68/KP1 being the most commonly expressed marker, followed by, in order of decreasing frequency, MPO, CD117, CD99, CD68/PG-M1, lysozyme, CD34, TdT, CD56, CD61, CD30, glycophorin, and CD4.³ Although a variety of chromosomal abnormalities have been reported in 55% of cases, none of these alterations was found in our patient. Any organ system may be involved in MS, although the skin, soft tissues, and nodes account for up to half of all localisations. MS of the gastrointestinal tract is reported in 6.5% of cases³ at any site from the mouth to the anal verge, but this sarcoma seems to have a predilection for the small bowel, as other neoplasms with elevated haematogenous spread, such as melanoma and lung cancer. The reason for this small-bowel tropism is unclear, but it may be due to the large vascular and lymphatic supplies of the absorptive portion of the gastrointestinal tract. A unique feature of this case is the clinical presentation with chronic diarrhoea and weight loss and without abdominal pain or obstructive symptoms. In a literature review performed by Kohl and co-workers, none of the 19 patients diagnosed with small-bowel MS had the same clinical presentation.⁵ From a pathophysiologic standpoint, in experimental animals, mechanical bowel wall stimulation, such as mucosal stroking and distension of the intestine, initiates luminal chloride secretion by myenteric and submucosal neural mechanisms; thereafter, the release of vasoactive compounds such as VIP may mediate hypersecretion and splanchnic vasodilation, as demonstrated in an elegant

experimental canine model of partial small-bowel obstruction.⁶ Differential diagnosis of the pathologic specimen may be difficult, particularly in the case of lymphoblastic, Burkitt's, or diffuse large B-cell lymphoma or even a non-haematologic malignancy,¹ and a high suspicion index of MS is mandatory to perform immunophenotyping to determine the correct management. Finally, we will make a short comment on the use of SBCE to investigate the unexplained diarrhoea of our patient. As outlined in a recent review article, SBCE is considered to be appropriate in the clinical setting of chronic diarrhoea associated with at least one “red-flag” symptom (weight loss, anaemia, and increased C-reactive protein levels).⁷ In the largest unselected, multicentre, prospective study of SBCE, chronic diarrhoea was the indication in 140 of 2921 (4.8%) consecutive examinations, and SBCE was diagnostic and retained in 39 (27.8%) and 2 (1.4%) cases, respectively.⁸ Our patient developed acute intestinal obstruction after capsule retention, representing the 0.07% in the quoted series. Furthermore, to the best of our knowledge, no SBCE image of small-bowel MS has been previously published. Interestingly, using retrograde double-balloon enteroscopy, Hotta and Kunieda recently described the endoscopic picture of ileal MS, consisting of swollen villi of different sizes and mucosal thickening, a picture overlapping with the SBCE findings of our patient.⁹ In conclusion, we reported an additional case of small-bowel MS presenting with chronic diarrhoea and weight loss, and, for the first time, we showed the SBCE findings of this rare tumour. Although our patient experienced a complication of SBCE, mandating surgery, her diarrhoea resolved after tumour resection, and chemotherapy was ultimately feasible.

Conflicts of interest

The authors declare no conflicts of interest.

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Ethical approval

None.

Author contributions

Each author has participated sufficiently, either intellectually or practically, in the work to take public responsibility for the content of the article, including the conception, design, and data interpretation.

CMG, VC, and ML provided clinical advice and consultation and participated in the design of the report; VC was the treating surgeon; ML was the referring haematologist; AA and GS performed the pathologic studies; and CMG and GS drafted the manuscript. All authors have read and approved the final manuscript.

Key learning points

- Small-bowel myeloid sarcoma can be the first presentation of acute leukaemia.
- Chronic diarrhoea and weight loss can be the only symptoms of small-bowel myeloid sarcoma.
- Capsule endoscopy may provide a diagnostic clue, but it can trigger an acute bowel occlusion.
- To address the right treatment, high suspicion index is paramount to perform appropriate immunohistochemistry on the pathological specimen.

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