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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.

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.

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 11.2×10^9/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);

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

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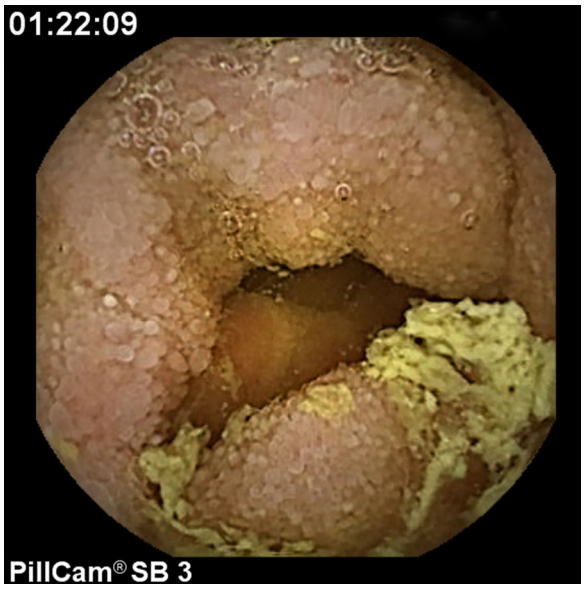

Fig. 1. Small-bowel capsule endoscopy frame showing a luminal stricture with thickened mucosa and short, swollen villi.

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 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

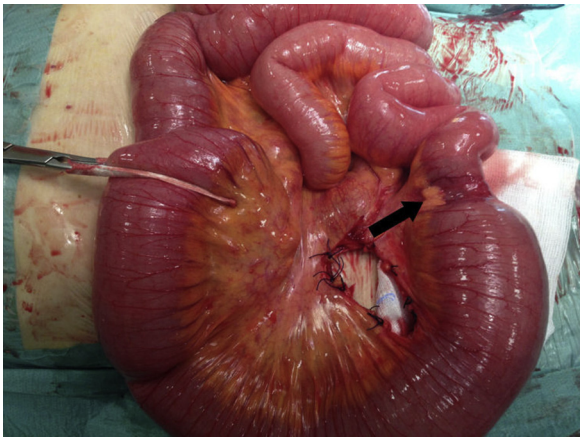

Fig. 2. Laparotomic finding of an annular, stricturing mass lesion of the ileum (arrow).

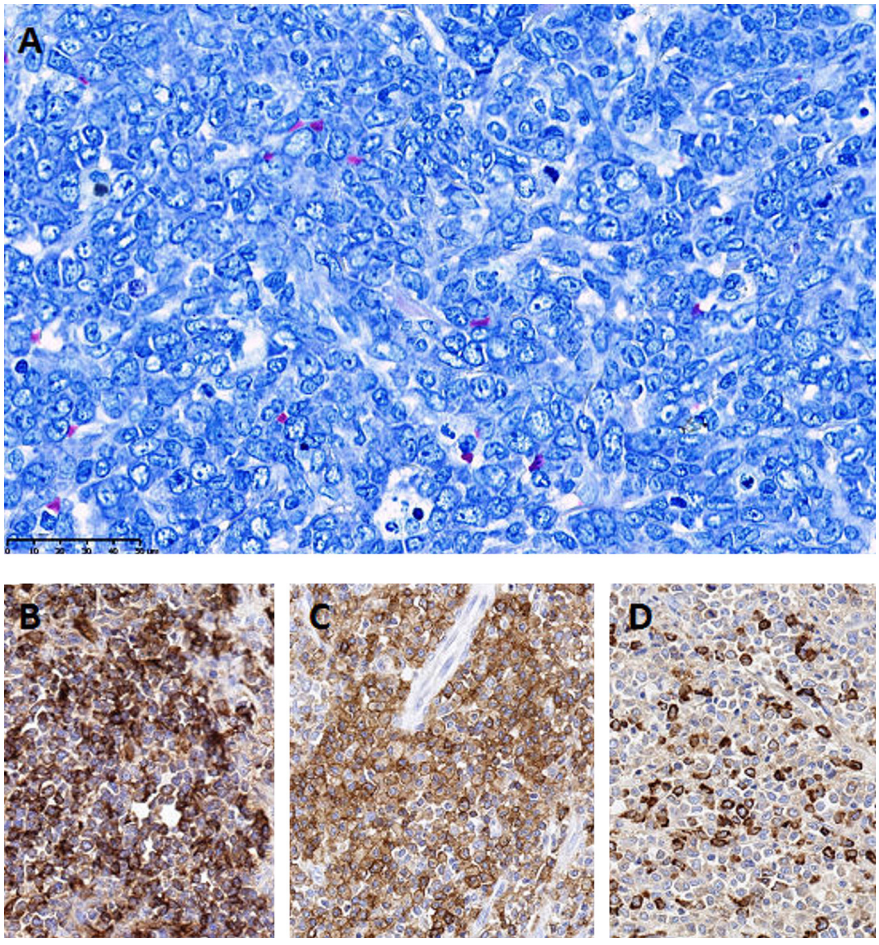

Fig. 3. Histopathology of the resected bowel specimen. (A) Diffuse infiltration by round, small- to medium-sized cells with moderate basophilic cytoplasm. The cells had round or oval folded nuclei containing dispersed chromatin and exhibited strongly positive staining for CD34 (B), CD117 (C), and myeloperoxidase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