

Letters to the Editor

Myeloid sarcoma of gastrointestinal tract: A rare cause of obstruction

Key words: Obstruction. Myeloid sarcoma.

Dear Editor,

Myeloid sarcoma (MS) is a rare disease presented as an extramedullary tumor composed of immature myeloid cells (1-3). Gastrointestinal involvement is rare.

Case report

A 27-year-old woman was admitted with a 10-days history of abdominal pain and intermittent vomiting. Her health was generally good, with no prior abdominal surgery, no history of weight loss, night sweats, anorexia, or family history of malignancy.

On admission, the patient was dehydrated. The abdominal examination revealed a mildly distended abdomen with generalized abdominal tenderness, with hyperactive bowel sounds. No rebound tenderness was noted.

Laboratory examination was unremarkable. Abdominal X-ray revealed a few distended small bowel loops with air fluid levels. Abdominal ultrasound and enhanced computed tomography (CT) documented dilated jejunal loops attributable to parietal mass with 4.6x4.5 cm, infiltration of the mesentery and the greater omentum (Fig. 1A). The tumor markers and viral serologies were negative.

In this context, an ultrasound-guided biopsy of the lesion was performed, showing on hematoxylin and eosin staining diffuse

infiltration of large cells, with occasional prominent nucleoli and moderate eosinophilic cytoplasm. Immunohistochemistry was positive for CD34, CD117, myeloperoxidase, lysozyme and Bcl-2.

The peripheral blood did not show circulating blasts, and the bone marrow aspirate excluded infiltration by malignant cells. The molecular analysis did not identify any cytogenetic abnormalities.

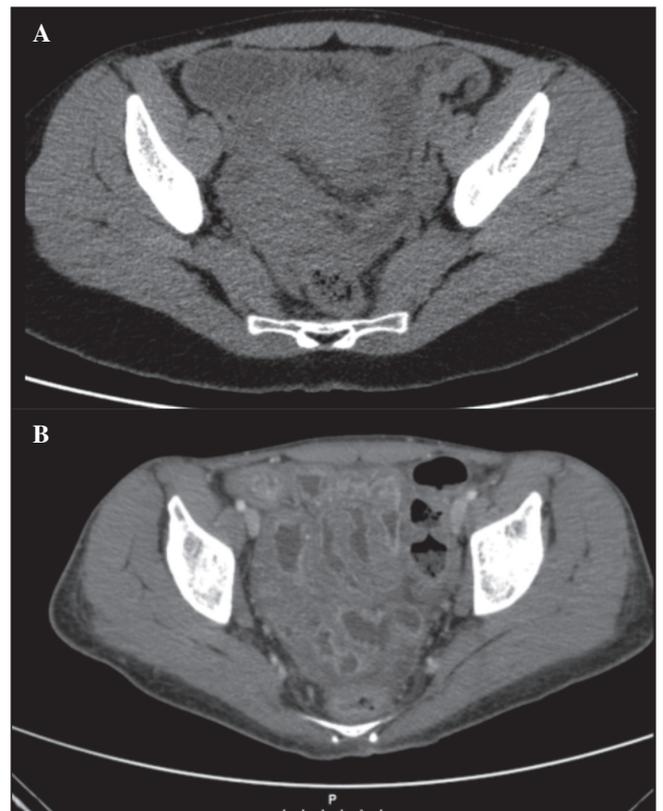


Fig. 1. Abdominal CT scan showing a mass and tumor infiltration of the mesentery (A). Abdominal CT scan showing disappearance of the mass. No evidence of bowel obstruction (B).

The patient initiated systemic chemotherapy with idarubicine and cytarabine, without response, and started mitoxantrone and etoposide with good response. The patient was proposed to allogeneic transplant by a donor family member.

An abdominal CT scan done one month later revealed the disappearance of the tumor (Fig. 1B). One year after allogeneic transplant the patient is clinically well without evidence of recurrence.

Discussion

Myeloid sarcoma (MS) is an extra medullar tumor of immature myeloid cells. More than 50% of the tumors are misdiagnosed as non-Hodgkin lymphoma. The prevalence rate is difficult to determine but it has been estimated in 2/1,000,000, with a predilection for males (2:1) (4,5).

It generally occurs in association with a myeloproliferative disorder and in this latter setting the majority (88%) of untreated patients progress to acute myeloid leukemia within 11 months (6).

MS can be present as either isolated or multiple lesions that may be synchronous or metachronous. The most common sites are the skin, bone, and lymph nodes (6,7). The synchronous involvement of the jejunum and the greater omentum is rare. Immunohistochemical stains and flow cytometry led to the correct diagnosis (2,6).

MS has been associated with various cytogenetic abnormalities, particularly t (8:21) (54%) and inv (16) (p13; q22) (25%). The prognostic significance of these chromosomal rearrangements remains uncertain (2).

The optimal therapy for primary MS still undetermined (2). Systemic chemotherapy, surgical resection, radiotherapy, bone marrow transplantation, or a combination of these approaches, are used (4).

In conclusion, this case is noteworthy because of the rarity of gastrointestinal involvement. The prompt recognition and early institution of therapy was crucial to avoid its progression to a systemic disease. The disappearance of the tumor after chemotherapy and allogeneic transplant makes this case a success.

Liliane C. Meireles¹, Ana Catarina Lagos², Inês Marques³,
Fátima Serejo¹ and José Velosa¹

¹Centro Hospitalar Lisboa Norte. Lisbon, Portugal. ²Hospital das Forças Armadas. Lisbon, Portugal. ³Centro Hospitalar das Caldas da Rainha. Caldas da Rainha, Portugal

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