Primary myeloid sarcoma of the jejunum and greater omentum causing small intestine obstruction

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Abstract

Myeloid sarcoma, which is highly associated with acute myeloid leukemia, is defined as an extramedullary discrete tumor mass, consisted by immature myeloid cells or myeloblasts. Myeloid sarcoma usually involves the skin, lymph node, bone, soft tissue and testis, while involvement of the gastrointestinal tract is rather uncommon. The diagnosis depends on histological features and immunohistochemical results. We present a rare case of myeloid sarcoma, with synchronous involvement of the jejunum and the greater omentum, manifesting with small bowel obstruction. (Acta gastroenterol. belg., **2009**, 72, **369-372**).

Key words : chloroma, granulocytic sarcoma, gastrointestinal tract.

Introduction

Myeloid sarcoma is defined as an extramedullary discrete tumor mass, consisting of immature myeloid cells or myeloblasts. It is also known as granulocytic sarcoma, chloroma, myeloblastoma and extramedullary myeloid cell tumor (1-6). It is highly associated with acute myeloid leukemia, and can occur subsequently, arise concurrently or precede the onset of leukemia by months or even years. In the latter case it is called primary sarcoma and occurs in non leukemic patients, the majority of whom, if untreated, progress to acute myeloid leukemia within 11 months (7). Myeloid sarcoma may also develop in patients with myelodysplastic or myeloproliferative disorders (1,2,5,6). Myeloid sarcoma can occur in virtually any anatomic site, with the skin, lymph node, bone soft tissue and testis being more frequently involved (1,3) whereas involvement of the gastrointestinal tract is uncommon (5,6), presenting at multiple anatomic sites in less than 10% of the cases. Myeloid sarcoma shows a predilection towards men, with male to female ratio being 1.2:1 and the median age of onset is 56 years (3). We report a rare case of myeloid sarcoma of the jejunum and greater omentum, which caused small intestine obstruction, in a 48 year old male without any hematological disorder.

Case report

A 48 year old male presented with epigastric pain, distension and vomiting, symptoms indicative of small bowel obstruction. The patient did not have any past medical history. Physical examination did not reveal any pathologic condition. Laboratory examinations and abdominal X-ray were within the normal range. The symptoms subsided within days after admission.

A CT scan was performed showing wall thickening of a small intestine segment and infiltration of the mesentery and the peritoneal adipose tissue (Fig. 1). Push enteroscopy was performed which revealed an obstructive exophytic mass about 50 cm from the ligament of Treitz and biopsies were taken. Colonoscopy did not reveal any mucosal lesions.

Histopathologically, dense and diffuse infiltration by medium and large cells was observed. The neoplastic cells caused total effacement of the tissue architecture. The cells had a faintly stained cytoplasm, indistinct cell boundaries and medium sized nuclei. The neoplasmatic nuclei presented with increased atypia and pleomorphism and had prominent nucleoli and atypical contour (Fig. 2). Among the tumor cells a few inflammatory cells were present, both neutrophil polymorphonuclear leukocytes and small mature B- and T- lymphocytes. Immunohistochemistry revealed intense positivity for myeloperoxidase, CD68 and protein bcl-2 and partial positivity for CD11c, CD34 and CD117 (Fig. 3). The neoplasmatic cells were negative for T-cell markers (CD3, CD4, CD5, CD8, CD45RO), for B-cell markers (CD20, CD79a) and for CD56, Tdt, bcl-6, CD30, CD10, PAX5, MUM-1, AE1/AE3, Calretinin, WT-1, Ber-EP-4, Plasma cell, CD138, CD15, Cyclin D1. The Ki-67 was approximately 70-80%. These findings indicated myeloid sarcoma.

A capsule enteroscopy was decided in order to study the small intestine and detect other lesions. Unfortunately the capsule did not pass the exophytic mass and an effort to retrieve it by enteroscopy was unsuccessful, so it had to been removed surgically. An exploratory laparotomy was performed, during which a mass obstructing the jejunum was found and also multiple masses were observed on the greater omentum (Fig. 4). Segmental resection of the small intestine with primary anastomosis and dissection of the greater omentum was performed. The small bowel segment that was

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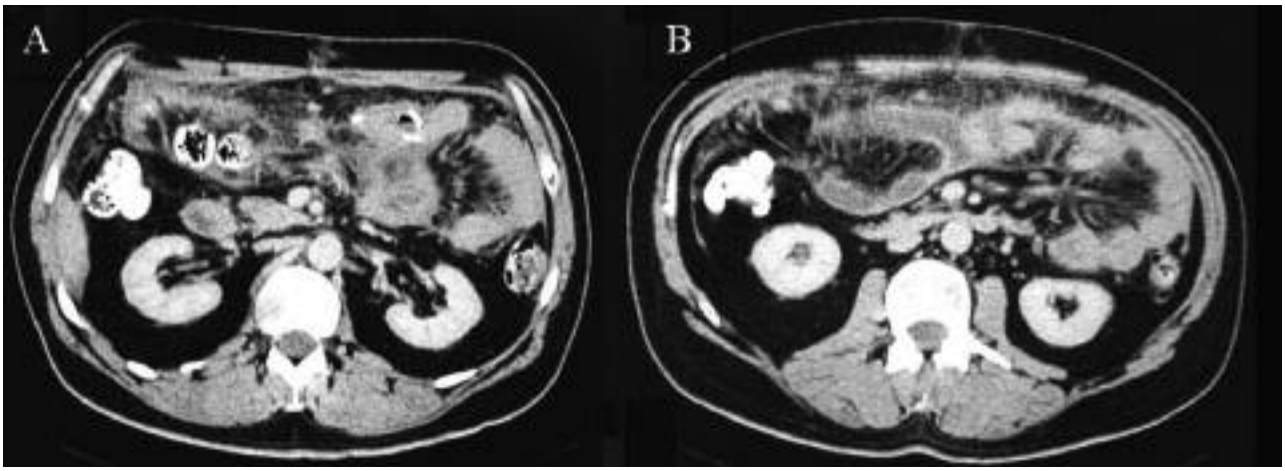

Fig. 1. — CT scan showing A : wall thickening of a small intestine segment and B : infiltration of the mesentery and the peritoneal adipose tissue.

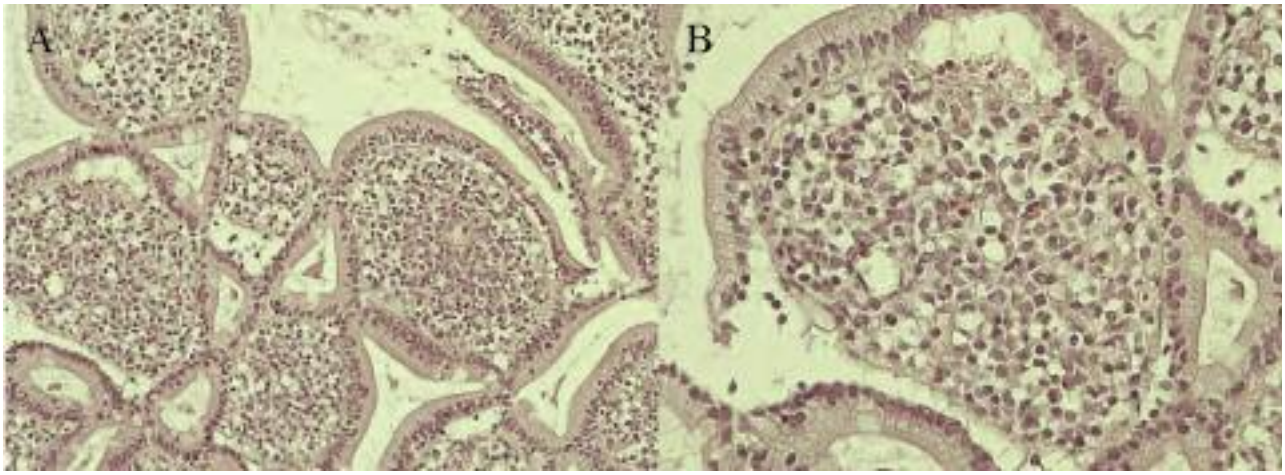

Fig. 2. — Hematoxylin – eosin stain of the mass showing neoplastic cells causing total effacement of the tissue architecture. The cells have a faintly stained cytoplasm, indistinct cell boundaries and medium sized nuclei. (A : ×200, B : ×400).

resected was 58 cm long and had 2 tumors : an orbicular one, that was 4.5 cm long, localized at 50 cm from the ligament of Treitz, and a polypoid one, that was 1.4 cm long at a distance of about 2 cm from the previous lesion. The histological and immunohistochemical findings of the small intestine and the greater omentum were consistent with the previously found. The infiltrate extended to the mucosa, submucosa, muscularis, serosa and even to the adjacent adipose tissue and the small bowel mesentery. From the surgical margins only the serosa and adipose tissue were involved.

There was no evidence of blood involvement suggesting acute leukemia or other myeloproliferative disorders and the bone marrow aspiration was normal. Postoperatively the patient was treated with acute myeloid leukemia chemotherapy regimen course with cytarabine and novantrone followed by a course with cytarabine and idarubicine and another course with cytarabine and novantrone.

After six months there is no evidence of recurrent disease or acute myeloid leukemia.

Discussion

Myeloid sarcoma is a rare disease that more commonly involves the skin, the bones and the lymph nodes whereas involvement of the gastrointestinal tract is more uncommon (5-8). Ileum is the most frequently involved region of the gastrointestinal tract (6,9), while its presence in the jejunum is quite rare. To our knowledge, our case of synchronous involvement of the jejunum and the greater omentum is the first reported in literature. The tumor presents a close relationship with leukemia and may occur as a complication of acute myeloid leukemia, during a blast crisis of chronic myeloid leukemia or leukemic transformation of myelodysplastic disorders, precede the onset of acute myeloid leukemia or finally present as an isolated lesion (8). In our case the patient

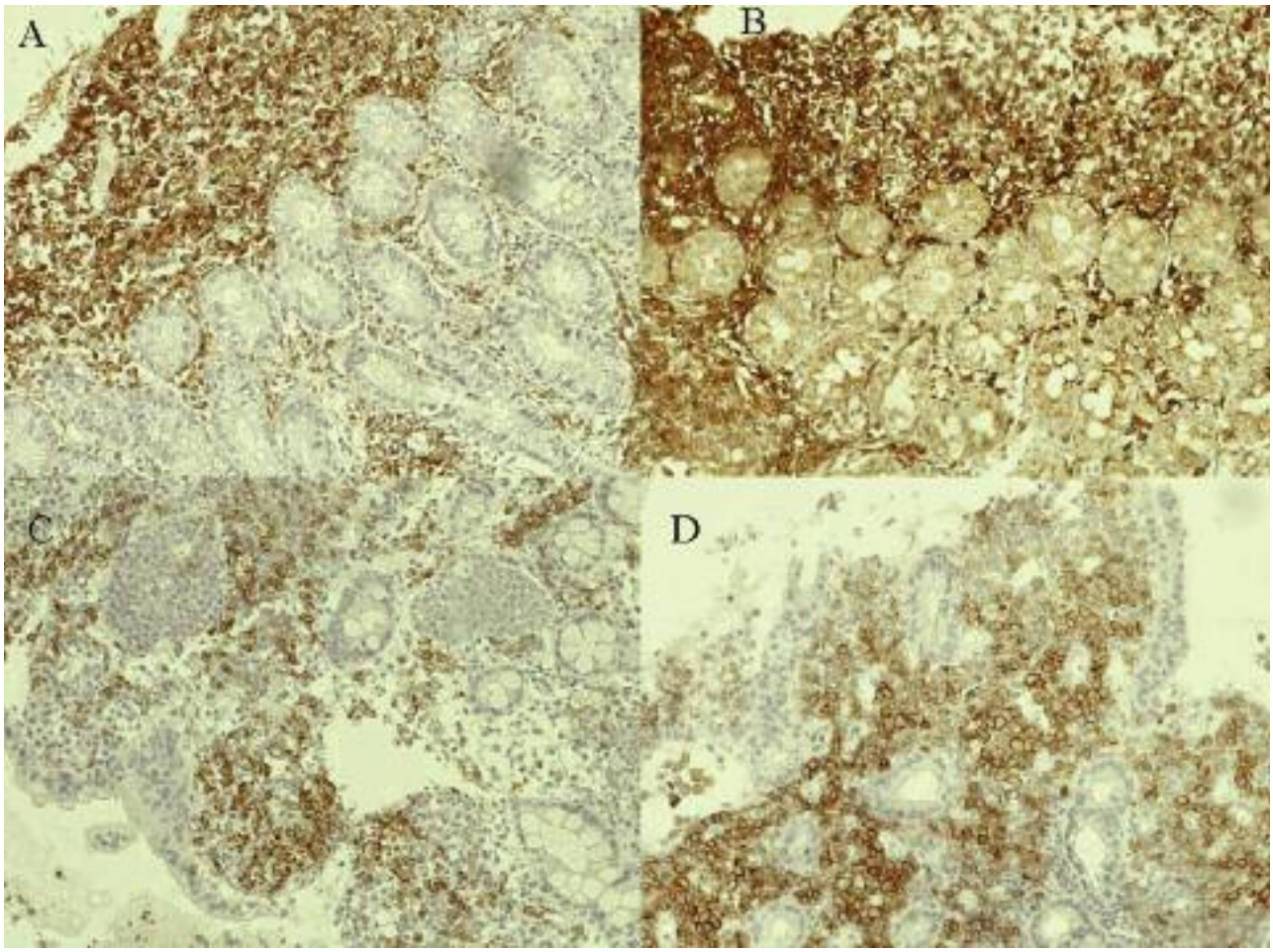

Fig. 3. — Immunohistochemical stain showing that neoplasmatic cells express A : MPO (×200), B : CD68 (×200), C : CD11c (×200), D : CD117 (×200).

did not have any signs of leukemia and the tumor can be considered as primary non leukemic myeloid sarcoma.

Although the majority of these tumors don't produce clinical signs and symptoms, the clinical presentation of myeloid sarcomas of the gastrointestinal tract may include gastrointestinal bleeding, chronic anemia, epigastric pain, perforation or small bowel obstruction, usually manifested with abdominal distention and vomiting, as in our case (5,6,10). Laboratory and imaging examinations have little to offer to the diagnosis. A recent study of Choi et al reported that the CT findings of myeloid sarcoma are characterized by variety of shape and contrast enhancement but there is a high predilection for mesenteric and peritoneal spread (9).

Macroscopically myeloid sarcoma of the gastrointestinal tract may present as exophytic or polypoid masses, wall thickening and ulceration. In our case there were two masses, one exophytic and one polypoid. Although many tumors, especially those with granulocytic differentiation, exhibit a green color, because of the presence of myeloperoxidase (1), this is not frequent in cases of gastrointestinal involvement, where most tumors have a pink to gray-white color (7). Histologically, myeloid sarcoma consists of neoplasmatic myelocytes with or without maturation, that replace the tissue architecture (3,5). The neoplasmatic cells are usually of medium to large size with medium to large nuclei and often prominent nucleoli (1,7). Also there are numerous mitoses (1). Immunohistochemistry shows in most cases positivity for one or more of the following antibodies as myeloperoxidase, lysozyme, napthyl-ASD-chloroacetate esterase, CD34, CD43, CD68 and CD117 (1,3,7). In this case the neoplasmatic cells were not only positive to myeloperoxidase, CD68, CD34 and CD117 but also stained for CD11c and bcl-2.

Differential diagnosis is mainly with malignant lymphoma, especially diffuse large cell lymphoma (1,2) and rarely lymphoblastic lymphoma (1,2) or Burkitt lymphoma (1,3) and depends on histological features and immunohistochemical results.

Myeloid sarcoma can be treated with systematic chemotherapy, irradiation, surgical resection or bone marrow transplantation. In a study comparing treatment modalities in 74 cases of primary myeloid sarcoma it was found that median non leukemic period was 3 months in the patients treated with surgical resection, 6 months in

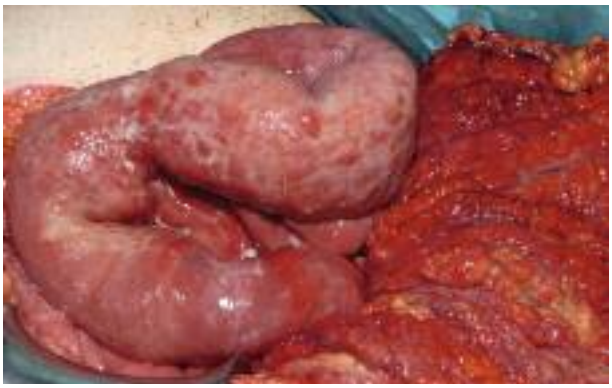

Fig. 4. — Intraoperatively, a mass obstructing the jejunum and also multiple masses on the greater omentum were observed.

the patients that received local radiation and 12 months in the patients treated with systemic chemotherapy. The study also emphasized the observation that a significant longer nonleukemic period occurred after treatment with acute myeloid leukemia chemotherapy regimen than treatment with other agents. Patients undergoing bone marrow transplantation seem to have a higher possibility of prolonged survival or cure (3).

The overall prognosis is poor with the median interval period to development of acute myeloid leukemia being 11 months and the median survival 22 months (6,7) but some patients may remain disease free and not develop acute myeloid leukemia after discontinuation of treatment (6). In the present case, six months after diagnosis, there was no manifestation of acute myeloid leukemia. There are no prognostic factors regarding clinical behavior and response to the treatment (3). Although all patients should be treated with chemotherapy for acute myeloid leukemia, surgical intervention is indicated in cases of complications related to the tumor.

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