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## Case Report

# Isolated primary myeloid sarcoma of small intestine – A case report and review of the literature

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### ARTICLE INFO

#### Article history:

Received 24 February 2012

Accepted 15 June 2012

#### Keywords:

Primary myeloid sarcoma  
 Granulocytic sarcoma  
 Small intestine

### ABSTRACT

**Introduction:** Myeloid sarcoma (MS) is a rare condition that is characterized by the occurrence of an extramedullary tumor consisting of immature myeloid cells – granulocytes, monocytes or both. MS of the small intestine in a nonleukemic patient is rare.

**Aim:** The aim of this work was to report and analyze a rare case of MS of the small intestine in a nonleukemic patient.

**Case study:** A 41-year-old male was admitted to hospital with symptoms of bowel obstruction. He suffered from severe abdominal pains, vomiting and constipation of a 12-h duration. He also reported a 2-week history of nausea and colic. On admission, the patient's general condition was good. He was normotensive and denied fever, weight loss, and allergy. Generalized abdominal tenderness was noted on palpation with hyperactive peristalsis, high bowel sounds and no guarding. There was no palpable lymphadenopathy. Results of laboratory tests, including white blood cell count of  $7.17 \times 10^9/L$ , red blood cell count of  $5820 \times 10^9/L$ , hemoglobin level of 171 g/L, and coagulation factors, were all normal. The patient underwent emergency laparotomy and part of his small intestine with tumor was resected.

**Results and discussion:** The patient underwent exploratory laparotomy which revealed nodular masses in the mesentery and in the wall of the small bowel. The diagnosis of a mechanical obstruction was confirmed. The involved part of the small bowel along with the mesentery was resected and sent for a histological examination. The histological examination of the specimen showed diffuse infiltration of a full thickness of the bowel, extending into the mesentery, by medium-sized neoplastic cells. The cells were round to oval in shape, with mild to moderate basophilic cytoplasm, predominantly agranular. The cells had a high N:C ratio, round or oval nucleus, dispersed chromatin and prominent nucleolus. The infiltration contained eosinophils, including many myelocytes and metamyelocytes. Immunohistochemical staining was performed on the paraffin-embedded sections. MS was diagnosed.

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Conclusions: (1) Correct, prompt diagnosis and appropriate immediate treatment are of crucial importance in nonleukemic MS. (2) If the initiation of treatment is postponed, it is highly probable that the patient will progress to acute myeloid leukemia (AML).

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## 1. Introduction

Myeloid sarcoma (MS), also called extramedullary myeloid tumor, granulocytic sarcoma or chloroma, is a rare solid tumor composed of immature myeloid blasts occurring at an extramedullary site.<sup>1</sup> Usually it involves the bone, skin, lymph node, and soft tissue although any part of the body may be affected. In less than 10% of cases, MS occurs at multiple anatomical sites.<sup>3,7,9,13,14,16,18,22</sup> MS may develop de novo or concurrently with acute myeloid leukemia (AML), chronic myeloproliferative disorder (CMPD), myelodysplastic syndrome (MDS) and myelodysplastic syndrome/myeloproliferative diseases (MDS/MPD).<sup>5,8,21,22</sup> There is a predilection for males (males:females as 1.2:1.0) and for this disease to occur in last decades of life – median age at time of diagnosis is 56 years.<sup>22</sup> An isolated primary MS of the small intestine in a nonleukemic patient is uncommon and often a small bowel obstruction is the first symptom indicating this disease.<sup>11,13</sup>

## 2. Aim

The aim of this work is to report and analyze a rare case of MS of the small intestine in a nonleukemic patient.

## 3. Case study

A 41-year-old male was admitted to hospital with symptoms of bowel obstruction. He suffered from severe abdominal pains, vomiting and constipation of a 12-h duration. He also reported a 2-week history of nausea and colic. On admission, the patient's general condition was good. He was normotensive and denied fever, weight loss, and allergy. Generalized abdominal tenderness was noted on palpation with hyperactive peristalsis, high bowel sounds and no guarding. There was no palpable lymphadenopathy. Results of laboratory tests, including white blood cell count of  $7.17 \times 10^9/L$ , red blood cell count of  $5820 \times 10^9/L$ , hemoglobin level of 171 g/L, and coagulation factors, were all normal. The patient underwent exploratory laparotomy which revealed nodular masses in the mesentery and in the wall of the small bowel (Fig. 1). The diagnosis of a mechanical obstruction was confirmed. The involved part of the small bowel along with the mesentery was resected and sent for a histological examination.

The surgical specimen was fixed in 4% formaldehyde and embedded in paraffin. Five-micron tissue sections were stained with hematoxylin–eosin. Immunohistochemical staining was performed on the paraffin-embedded sections and additionally reaction with myeloperoxidase (MPO), LCA/CD45, CD43, CD34, CD117, TdT, CD68, CD15, CD4, Ki-67, CD3, CD5, CD8, CD2, CD56, CD20, CD79a, CD7, CD30, ALK, CD138 and plasma cell.



Fig. 1 – The infiltration of the bowel and the mesentery (specimen was fixed in 4% formaldehyde).

#### 4. Results and discussion

The histological examination of the specimen showed diffuse infiltration of a full thickness of the bowel, extending into the mesentery, by medium-sized neoplastic cells. The cells were round to oval in shape with mild to moderate basophilic

cytoplasm, predominantly agranular. The cells had a high N:C ratio, round or oval folded nuclei, and dispersed chromatin including small nucleoli (Fig. 2). The infiltration contained eosinophils, including many myelocytes and metamyelocytes (Fig. 3). The tumor cells showed positive immunohistochemical reactivity with MPO (95%), LCA/CD45 (90%), CD43 (80%), CD34 (75%), CD117 (75%), TdT (30%), CD68 (5%), CD15 (3%), CD4 (1%) and

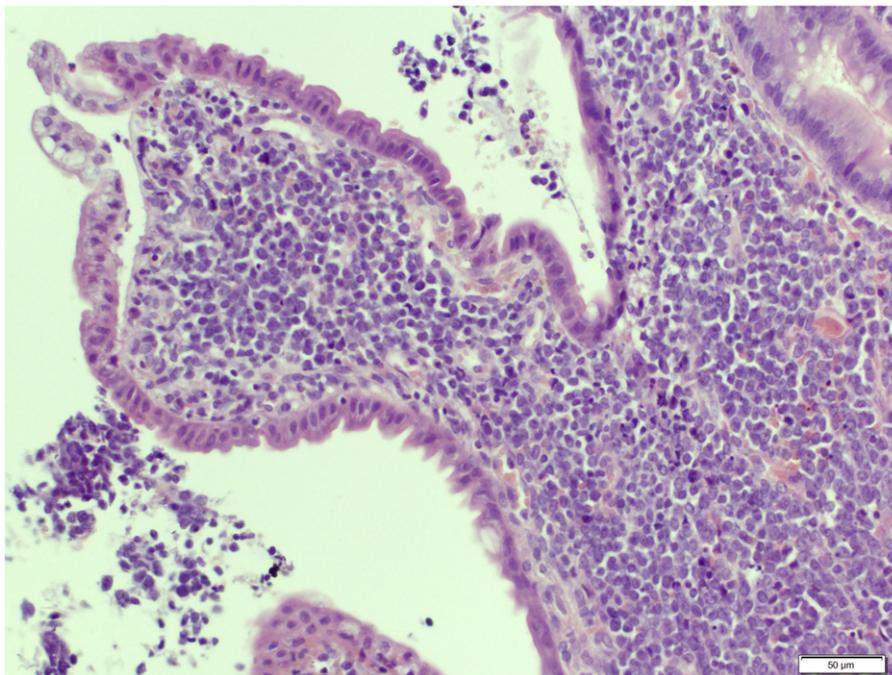


Fig. 2 – The infiltration of MS in the mucous membrane of the bowel (HE, magnification 20 ×).

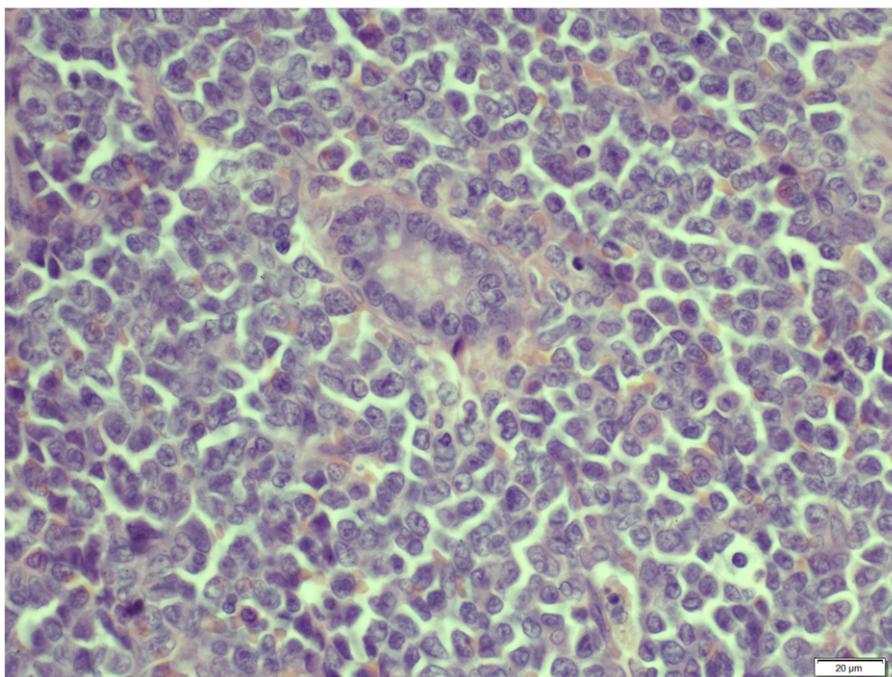


Fig. 3 – Myeloid sarcoma – the architectural features of mucous membrane such as glandular tubules are not destroyed by infiltration of MS. The neoplastic cells have nuclei with thin nuclear membranes and small nucleoli (HE, magnification 40 ×).

mitotic activity Ki-67 (75%). The neoplastic cells did not react with the following antibodies: CD3, CD5, CD8, CD2, CD56, CD20, CD79a, CD7, CD30, ALK, CD138 and plasma cell (Figs. 4-7).

On the basis of these results, the diagnosis of MS was established. After the surgery, the patient was discharged from hospital on the 7th postoperative day in a good general condition. In the following month his condition dramatically worsened.<sup>23</sup> At that time the histopathological diagnosis of MS was made. Although the patient was admitted to the

oncology center, it was too late to save his life. He died from AML, 1.5 months following the surgery.

MS is a neoplasm of blood-forming cells such as myeloblasts, immature granulocytes, monocytes with or without mature granulocytes, presenting itself at any extramedullary site as an isolated tumor without bone marrow infiltration or as a disseminated disease involving many organs. MS can be subclassified into five histological types: immature granulocytic sarcoma (more than 90% of blasts, MPO < 10%),

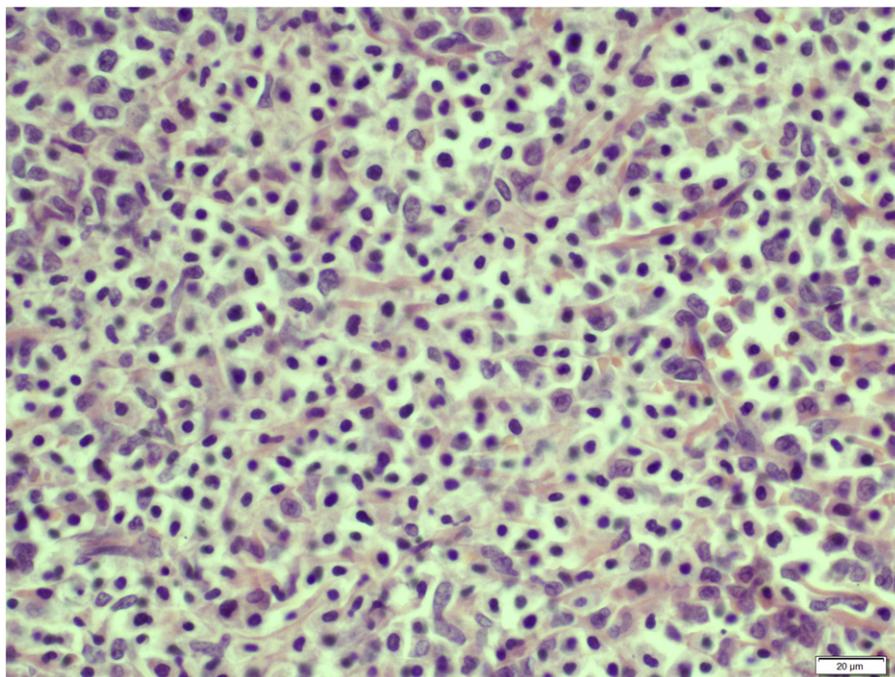


Fig. 4 – Myeloid sarcoma – there are granulocytes in various degree of maturation and blasts (HE, magnification 20 ×).

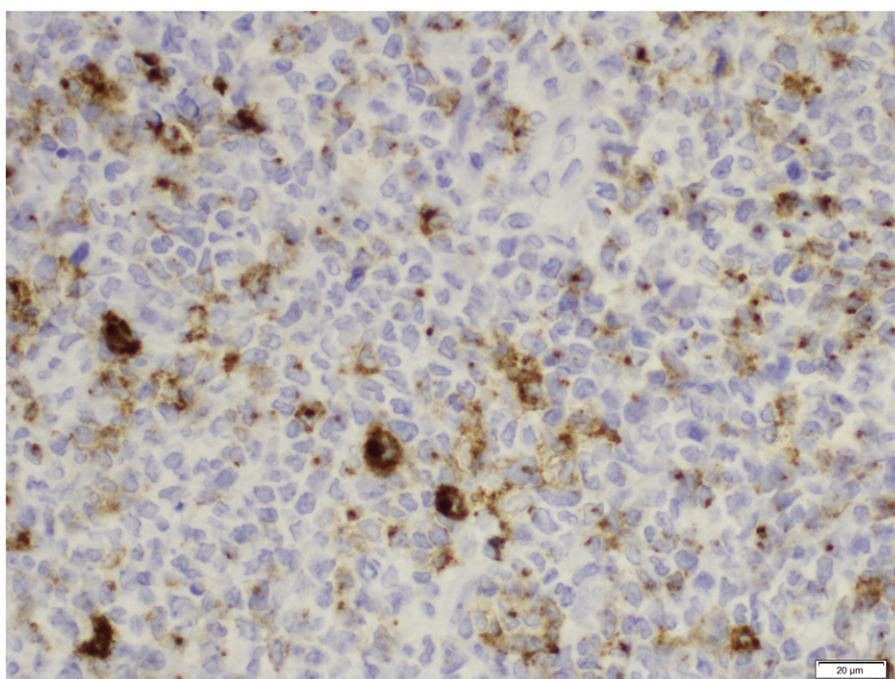
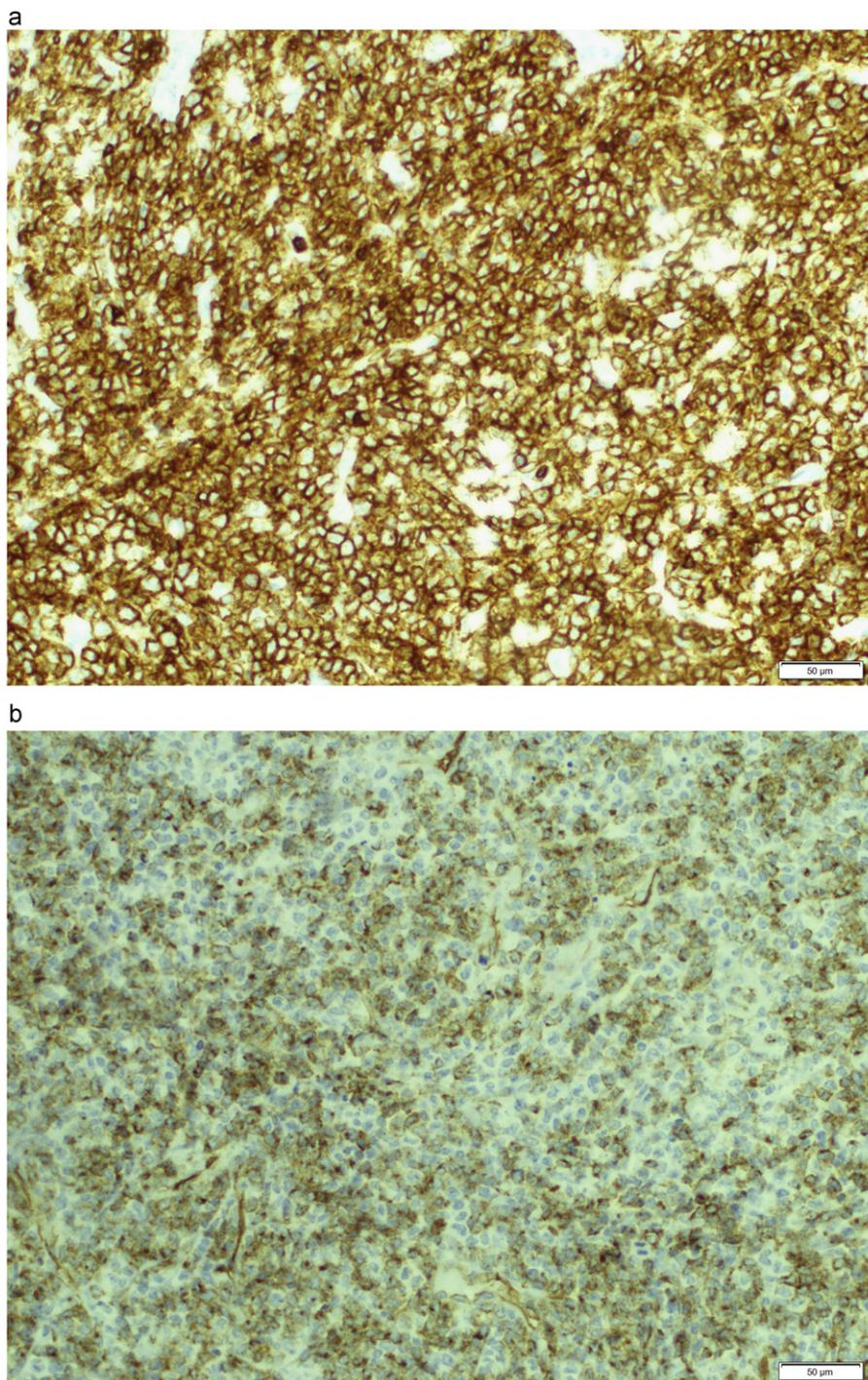


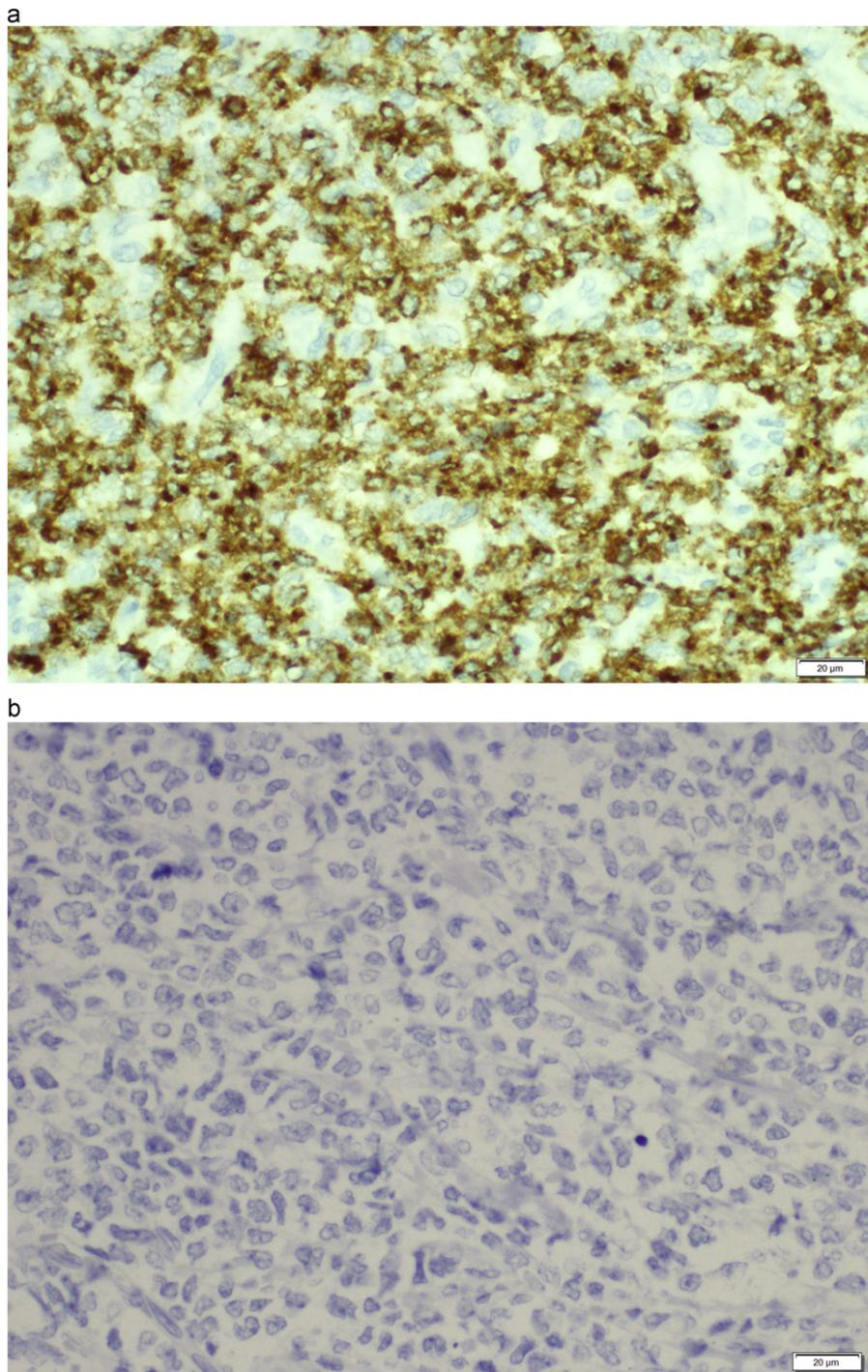
Fig. 5 – Myeloid sarcoma – there are granulocytes positive for CD15 (magnification 20 ×).



**Fig. 6 – The neoplastic cells are strongly positive for LCA (CD45) (a) and positive for CD34 (b).**

differentiated granulocytic sarcoma (mature granulocytes more than 10%), monoblastic sarcoma (more than 80% of monoblasts, CD34–), monocytic sarcoma (CD43+, CD68+), and myelomonocytic sarcoma.<sup>12</sup> Isolated primary MS is an uncommon cause of small bowel obstruction. The patient was admitted to hospital without any significant abnormalities in blood count. Therefore, correct diagnosis is very important. In the absence of hematological disorders the rate of misdiagnosis is very high (50%), particularly when immunohistochemical stains are not employed.<sup>1,4,19</sup> MS is frequently mistaken for non-Hodgkin lymphoma (lymphoblastic type,

Burkitt lymphoma, diffuse large B-cell lymphoma), small round cell tumor (neuroblastoma, rhabdomyosarcoma, Ewing sarcoma or PNET, medulloblastoma) and undifferentiated carcinoma. Myeloid sarcoma can closely resemble lymphomas such as Burkitt lymphoma or diffuse large B cell lymphoma, but cells of MS are negative for CD20 and in contrast cells of that lymphomas are strongly positive for CD20.<sup>24</sup> According to the literature, an immunohistochemical panel including MPO, CD43, CD117, CD34, CD68, CD3, CD20 can successfully identify the majority of MS paraffin-embedded specimens.



**Fig. 7 – The neoplastic cells are strongly positive for MPO (a) and negative for CD20 (b).**

MS has been described in association with a variety of chromosomal abnormalities such as monosomy of 7 and 16, trisomy of 4, 8, 11 chromosomes and  $t(8;21)(q22;q22)$  and  $inv(16)(p13;q22)$ .<sup>24</sup> Normal karyotype is frequently observed in monoblastic sarcoma.<sup>17</sup> The optimal therapy for primary MS has not yet been clearly defined. Surgical resection and/or radiation therapy, traditional chemotherapy for AML may result in longer remissions.<sup>4</sup> Targeting c-kit (CD117) expressing tumors with tyrosine kinase inhibitors is a new therapeutic option.<sup>11</sup> Patients who undergo allogenic or autologous bone marrow

transplantation seem to have a higher probability of prolonged survival or cure.<sup>22</sup> Untreated patients present with a median nonleukemic period of 3 months.<sup>24</sup> Once AML develops, the prognosis for patients with granulocytic sarcoma becomes poor; the median survival period was reported to be between 6 and 14 months.<sup>4,12,17</sup> Thus, it is most important for the clinicians to have a high index of suspicion and to perform a bone marrow biopsy, as soon as possible, when the patient presents with atypical tumors, macroscopically greenish on cross sections.

## 5. Conclusions

1. Correct, prompt diagnosis and appropriate immediate treatment are of crucial importance in nonleukemic MS.
2. If the initiation of treatment is postponed, it is highly probable that the patient will progress to AML.

## Conflict of interest

None declared.

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