Case Report
A primary myeloid sarcoma involving the small intestine and mesentery: case report and literature review

Tao He¹, Yun Guo², Chao Wang³, Jun Yan⁴, Meiying Zhang⁵, Wei Xu¹, Xuan He¹, Shufang Zheng¹

Departments of ¹Pathology, ²Endocrinology and Hematology, ³Surgical Oncology, The Affiliated Hospital of Logistics University of Chinese People’s Armed Police Force, Tianjin 300162, China; ⁴Department of Pathology, First Center Hospital, Tianjin 300192, China; ⁵Department of Gastroenterology & Hepatology, Chinese PLA General Hospital, # 28 Fuxing Road, Beijing 100853, China

Received April 6, 2018; Accepted July 18, 2018; Epub August 1, 2018; Published August 15, 2018

Abstract: Myeloid sarcoma (MS) is a rare, extramedullary hematological malignant tumor. MS, which is shown to precede acute myeloid leukemia (AML), and in which bone marrow aspiration or biopsy finds no hematological disorder, is classified as primary or isolated MS. Primary MS with no evidence of the cancer in the blood is rare. Herein, we report a case of a primary MS involved the small intestine and mesentery. A 40-year-old man with intermittent upper vague abdominal pain for 1 month was admitted to the hospital on October 20th, 2017. The pain was obviously aggravated after food ingestion, but he had no nausea or vomiting. An abdominal computed tomography (CT) showed a soft tissue density mass in the mesenteric region and a wall thickening of the jejunum. Positron emission tomography (PET)-CT imaging with 18F-fluorodeoxyglucose (18F-FDG) showed a significant uptake in mesenteric regional mass (7.6 cm × 4.1 cm) and jejunum wall. The patient underwent a laparotomy, and the involved part of the small intestine along with the mesentery was resected. Histological examination and immunohistochemical (IHC) staining determined the pathological diagnosis was MS. Clinical laboratory tests and a bone marrow biopsy were used to rule out systemic AML. The patient had been treated with a combination of pirarubicin and cytarabine. A follow-up CT scan and necessary clinical laboratory tests were performed after the surgery and no abnormalities were found. To date, the patient continues to be in complete remission. In conclusion, primary MS is a rare disease, yet the diagnosis of MS should be considered when any mass with diffusely infiltrating tumor cells is observed.

Keywords: Myeloid sarcoma, granulocytic sarcoma, small intestine, AML, extramedullary

Introduction
Myeloid sarcoma (MS), also known as granulocytic sarcoma or chloroma, is a rare extramedullary tumor presenting one or more tumor masses composed of immature myeloid cells [1]. It can arise de novo or occur in association with myelodysplastic syndrome (MDS), acute myeloid leukemia (AML), or other myeloproliferative disorders [2]. MS, which is shown to precede AML, is called primary or isolated MS when bone marrow aspiration or biopsy finds no hematological disorder. It is a rare disease with an incidence of 2/1,000,000 in adults [2]. Herein, we report a case of a primary MS involved the small intestine and mesentery. The diagnosis and treatment issues are analyzed and discussed.

Case report
A 40-year-old man with an intermittent, upper, vague abdominal pain for 1 month was admitted to the hospital on October 20th, 2017. The pain was obviously aggravated after food ingestion, but the patient had no nausea or vomiting. The past history of cerebral infarction and hypertension of the patient occurred more than six months prior, and his highest blood pressure was 180/100 mmHg. With oral felodipine, his blood pressure was under control, and at admission, his blood pressure was 140/85 mmHg.

A general physical examination found that in his deep belly next to the navel one could reach a soft mass, and its size was about 7 cm × 4 cm
A case of primary myeloid sarcoma and a literature review

Figure 1. Initial abdominal computed tomography (CT) and positron emission tomography (PET)-CT. A: CT shows the following: Soft tissue density mass in the mesenteric region and wall thickening of the jejunum. B: (PET)-CT imaging with 18F-fluorodeoxyglucose (18F-FDG) showed a significant uptake in the mesenteric regional mass (7.6 cm × 4.1 cm) and the jejunum wall.

Figure 2. Gross specimen and histology revealed myeloid sarcoma involving the small intestine. (A) The involved part of the small bowel along with the mesentery that was resected. (B) The tumor invaded the intestinal wall and mesentery. (C) The small intestine had a decrease in mucosal glands in the lamina propria and an infiltration of diffusely dispersed primitive cells (H&E, × 100). (D) The tumor cells were round to oval in shape, with mild to moderate basophilic cytoplasm. The cells had a high ratio of nuclei to cytoplasms, round or oval nucleus, and dispersed chromatin (H&E, × 400). Bar = 5 cm (A, B), 50 μm (C, D).

Clinical laboratory tests found that the patient’s red blood cell count (4.13*10^{12}/L) and hemoglobin (117 g/L) were slightly decreased, his uric acid (442 umol/L) slightly increased, and his D-Dimer (2539.98 ng/ml) was significantly increased. The squamous cell carcinoma tumor marker antigen (3.27 ng/ml) was slightly increased. His liver enzymes and electrolytes were within normal limits.

An abdominal computed tomography (CT) showed the following: Soft tissue density mass in the mesenteric region and wall thickening of the intestine. Positron emission tomography (PET)-CT imaging with 18F-fluorodeoxyglucose (18F-FDG) showed a significant uptake in mesenteric regional mass (7.6 cm × 4.1 cm) and intestine wall (Figure 1).

The patient underwent laparotomy, and the operation involved part of the small intestine and the mesentery being resected (Figure 2A, 2B). A histological examination showed that an infiltration of primitive cells was found throughout the mesenteric region and the small intestine. The cells were round to oval in shape, with mild to moderate basophilic cytoplasm. The cells had a high ratio of nuclei to cytoplasms, round or oval nuclei, and dispersed chromatin (Figure 2C, 2D).

Immunohistochemical (IHC) staining was performed on the paraffin-embedded sections. IHC staining was positive for leukocyte common antigen (LCA), myeloperoxidase (MPO), CD34, CD117, CD68-KP1, CD38, and CD4, but negative for CD3 and CD20, and the Ki-67 was expressed in 60% to 80% of the cells (Figure 3).

A bone marrow biopsy of the patient, which was found to be within normal limits, was used to rule out systemic AML. The patient had been treated with a combination of pirarubicin and
cytarabine. A follow-up CT scan and routine clinical laboratory tests were performed after surgery, and no abnormalities were found. To date, he continues to be in complete remission.

Discussion

Myeloid sarcoma (MS) is a rare form (< 1%) of extramedullary acute myeloid leukemia (AML). Although MS can be developed at any anatomic site, the three most common presentation locations are connective/soft tissues, skin/breast, and the digestive system [1]. The median overall survival time is 8 months for primary MS patients [3]. In the digestive system, this sarcoma seems to have a predilection for the small intestine. A literature review reported that the clinicopathologic features of patients with MS involved the small intestine were diverse [4]. The unique features of our patient include a clinical presentation with abdominal pain but without chronic diarrhea, weight loss, or obstructive symptoms. Six months prior to his diagnosis, the patient had a history of cerebral infarction. However, there was no evidence that there was a correlation between the cerebral infarction and the patient’s MS.

MS can develop at any anatomic site, and a diagnosis of MS should be considered when any mass is infiltrated with diffuse tumor cells. In the absence of hematological disease, arriving at a precise diagnosis may be challenging. In fact, the initially diagnosis is often incorrect, with the rate of misdiagnoses running about 25-47% [5, 6]. Light microscopy limits the ability to make a correct diagnosis, and some cases have been misdiagnosed as other malignancies such as non-Hodgkin lymphoma (NHL), undifferentiated cancer, malignant melanoma, extramedullary hematopoiesis, and inflammation [7-9]. In this case, the limitations of diagnosis also led us to initially consider NHL when there was nothing found by peripheral blood examination and bone marrow biopsy. IHC analysis has an important role in the diagnosis. CD68-KP1 is the most frequently expressed
marker in MS, followed by MPO, CD117, CD99, CD68/PG-M1, lysozyme, CD34, TDT, CD56, CD61, CD30, glycophorin A, and CD4 [10]. MS may show weak LCA positivity, but it’s negative for B-cell (CD20) and T-cell (CD3), so these markers may be used to distinguish MS from NHL.

Since primary MS is a rare disease, the optimal treatments for MS have not been established. Nevertheless, when a bone marrow biopsy is negative for acute leukemia in primary MS, primary MS may develop into AML [11]. Hence systemic chemotherapy is recommended for all patients with primary MS [12-14]. In a retrospective study, systemic anti-leukemic chemotherapy decreased the rate of progression to AML and increased survival [15]. Surgery should be considered, especially for tumors which cause organ dysfunction and/or obstruction. However, surgery alone is not an effective treatment strategy for primary MS, and the incidence of relapse is significantly higher in patients who are treated by a localized approach with surgery or radiotherapy [5]. Obviously, the current treatment still needs to be further optimized. It was reported that early systemic chemotherapy was associated with worse outcomes in older patients (> 70 years) as compared to early radiation/surgery or no treatment [1]. AML induction therapy combined with 5-Aza maintenance therapy can achieve a long-term partial response [16], but it is necessary to accumulate more clinical evidence to confirm this. Our patient underwent a laparotomy and then received systemic chemotherapy. To date, he continues to be in complete remission.

Conclusion

Primary MS is a rare disease, and a diagnosis of MS should be considered for any mass with diffusely infiltrating tumor cells. Surgery should be considered, especially for primary MS involving the small intestine or when there is organ dysfunction and/or obstruction. Systemic chemotherapy is recommended.

Acknowledgements

This work was supported by the Natural Science Foundation of Tianjin (No. 18JCYBJC13000) and the Doctoral Start up Foundation of Logistics College of the Chinese People’s Armed Police Force (No. WHB201715).

Disclosure of conflict of interest

None.

Address correspondence to: Shufang Zheng, Department of Pathology, The Affiliated Hospital of Logistics University of Chinese People’s Armed Police Force, 220 Chenglin Road, Hedong District, Tianjin 300162, China. Tel: +86-022-60578814; E-mail: zhengshf@126.com

References

A case of primary myeloid sarcoma and a literature review


