Clinical Case Report

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A myeloid sarcoma involving the small intestine, kidneys, mesentery, and mesenteric lymph nodes

A case report and literature review

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Abstract

Rationale: Myeloid sarcomas (MSs) are rare malignant hematological tumors. They most commonly occur in patients with acute or chronic myeloid leukemia. A de novo MS with no evidence of blood system disease is rare, but may represent the first sign of a systemic illness that precedes a full-blown disease. Herein, we report the computed tomography (CT) findings of an extremely rare case of a nonleukemic MS that progressed to acute myelogenous leukemia (AML) and simultaneously involved the small intestine, kidneys, mesentery, and mesenteric lymph nodes. Moreover, we provide CT findings before and after AML chemotherapy, which have not been reported previously.

Patient concerns: A 25-year-old man with intermittent upper abdominal pain for 6 months was admitted to the hospital on November 28, 2015. Initial CT showed concentric wall thickening of the jejunum with an adjacent mesenteric soft tissue mass and mesenteric lymph nodes enlargement. Both kidneys were involved as indicated by the presence of well-defined mildly dilated lesions. During the laparoscopic surgery, the small intestinal tumor, mesenteric soft tissue mass, and mesenteric lymph nodes were removed.

Diagnoses: The pathological diagnosis was an MS.

Interventions: The patient refused systemic chemotherapy and was rehospitalized with persistently aggravated abdominal distension on February 17, 2016. Follow-up CT showed diffuse small bowel wall thickening, widespread infiltration of the peritoneum, omentum, and mesentery, mesenteric lymph node enlargement, and large amounts of ascites fluid. The lesions in both kidneys were substantially larger and more numerous than on initial CT. Then the patient was treated with conventional AML chemotherapy.

Outcomes: The patient achieved complete hematological remission on bone marrow examination. Follow-up CT in September 4, 2016, showed none of the abnormalities seen on initial CT. Currently, the patient is in complete remission.

Lessons: If the radiological examination shows lesions at multiple sites, and these lesions are soft tissue masses with homogenous enhancement, MS should be considered in the differential diagnosis, and an aspiration biopsy should be performed to provide a definitive pathological diagnosis. If MS is diagnosed, systemic chemotherapy is crucial to recovery; otherwise, the disease may progress rapidly. Medical imaging is helpful for diagnosing MS and for monitoring treatment response.

Abbreviations: AML = acute myeloid leukemia, CT = computed tomography, MS = myeloid sarcoma.

Keywords: computed tomography, kidney, mesentery, myeloid sarcoma, small intestine

1. Introduction

Myeloid sarcomas (MSs) are rare extramedullary tumors composed of immature myeloid precursor cells.[1–3] They generally occur during the natural course of acute myeloid leukemia (AML) or the blastic phase of chronic myelogenous leukemia, myeloproliferative disorder, or myelodysplastic disorder, or after remission. However, on rare occasions, their appearance precedes peripheral blood or bone marrow manifestations of these diseases.[1–3] Herein, we report a case of a nonleukemic MS that progressed to AML and simultaneously involved the small intestine, kidneys, mesentery, and mesenteric lymph nodes. Diagnostic and treatment issues are discussed.

2. Consent

The patient consented to the use of his data for teaching and publication purposes in writing.

3. Case report

A 25-year-old man with intermittent upper abdominal pain for 6 months and nausea and vomiting in the last 4 days was admitted to the hospital on November 28, 2015. The patient was otherwise healthy previously. A general physical examination revealed no abnormalities, whereas an abdominal examination revealed

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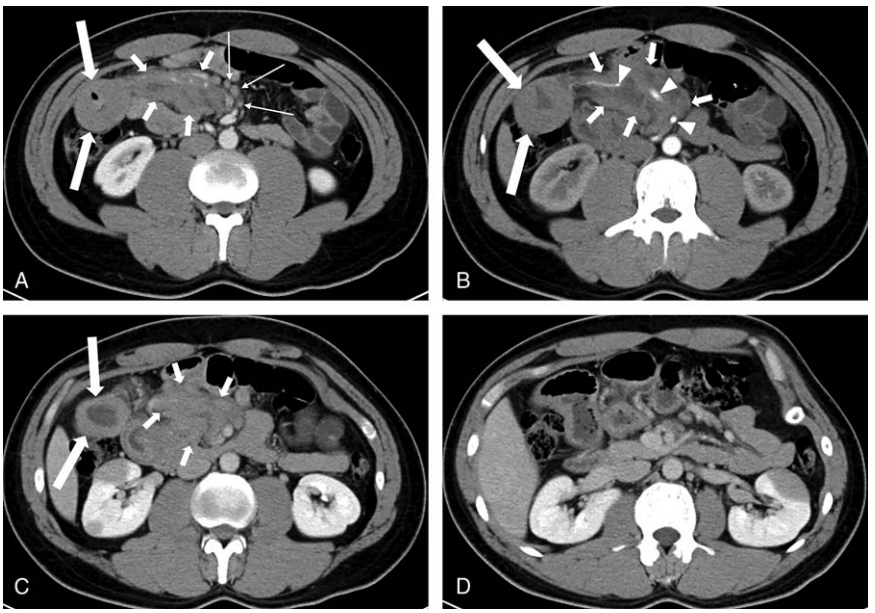

Figure 1. Initial transverse contrast-enhanced abdominal computed tomography. The images show concentric wall thickening of the jejunum (A–C, thick long arrows), an adjacent mesenteric soft tissue mass (A–C, short arrows) encasing a branch of the superior mesenteric artery (B, arrowheads), and mesenteric lymph nodes enlargement (A, thin long arrows). (C and D) the images show well-defined mildly dilated lesions with homogeneous enhancement indicative of the involvement of both kidneys.

moderate tenderness and rebound pain under the xiphoid. Laboratory test results were all within normal limits.

Initial contrast-enhanced abdominal computed tomography (CT) showed the following: concentric wall thickening of the jejunum with progressive homogeneous enhancement (average CT values: arterial phase, 44.6 HU; portal phase, 57.7 HU; equilibrium phase, 61.7 HU), an adjacent mesenteric soft tissue mass encasing a branch of the superior mesenteric artery, and mesenteric lymph node enlargement (Fig. 1A–C). Both kidneys were involved as indicated by the presence of well-defined mildly dilated lesions with progressive homogeneous enhancement (average CT values: arterial phase, 52.1 HU; portal phase, 73.4 HU; equilibrium phase, 79.7 HU) (Fig. 1C and D).

During laparoscopic surgery, a small intestinal tumor measuring 5×5 cm was found in the middle of the small intestine. In addition, a mesenteric soft tissue mass measuring 10×8 cm and closely associated with the superior mesenteric artery was found, along with prominent mesenteric lymph nodes, adjacent to the small intestinal tumor. The small intestinal tumor, the mesenteric soft tissue mass, and the mesenteric lymph nodes were all removed.

Histological examination showed infiltrates of primitive cells throughout the small bowel (Fig. 2 A and B). The tumor cells were large with moderate amounts of cytoplasm, and the nucleoli were visible. Immunohistochemical staining was positive for myeloperoxidase (Fig. 2C), CD34 (Fig. 2D), CD117 (Fig. 2E), CD68, CD99, CD4, CD10, CD43, and leukocyte common antigen. Ki-67 was expressed in 50% to 60% of the cells. The final diagnosis was an MS, with the bone marrow showing no hematological malignancy at that time. The patient refused systemic chemotherapy and other treatments for an MS and was discharged from the hospital.

The patient was readmitted to the hospital with persistent aggravated abdominal distension on February 17, 2016. His blood test results were as follows: white blood cells, 25.9×10^9/L; red blood cells, 5.01×10^{12}/L, hemoglobin, 13.9 g/dL; and platelets, 78×10^9/L. Bone marrow examination revealed that 65.5% of the cells in the bone marrow were myeloblasts, indicating AML.

Follow-up contrast-enhanced abdominal CT on February 17, 2016, showed the following: diffuse small bowel wall thickening, widespread infiltration of the peritoneum, omentum, and mesentery, mesenteric lymph node enlargement, and large amounts of ascites fluid (Fig. 3A–D). The lesions in both kidneys were substantially larger and more numerous than on initial CT, resulting in enlargement of both kidneys (Fig. 3B and C). After conventional AML chemotherapy with daunorubicin and cytarabine, the patient achieved complete hematological remission on bone marrow examination.

Follow-up contrast-enhanced abdominal CT on September 4, 2016, showed no diffuse small bowel wall thickening, peritoneal, omental, or mesenteric lesions, enlarged mesenteric lymph nodes, or ascites. The lesions in both kidneys had disappeared, and the renal parenchyma in the lesion areas had slightly atrophied (Fig. 4).

The patient later received a consolidation therapy. To date, he continues to be in complete remission.

4. Discussion

MSs, also referred to as granulocytic sarcomas, chloromas, myelosarcomas, and extramedullary myeloid cell tumors, most commonly occur in patients with acute or chronic myeloid leukemia.[1–3] A de novo MS with no evidence of blood system disease is rare, but may represent the first sign of a systemic illness that precedes a full-blown disease. An untreated primary MS ultimately progresses to AML 10–12 months on average after its diagnosis.[4,5] In the present case, the MS appeared de novo with no evidence of blood system disease and progressed to AML within 3 months. The absence of systemic signs and symptoms complicates the diagnosis of an MS. In general, 46% to 75% of MS cases in nonleukemic patients are initially misdiagnosed as large cell lymphomas.[5]

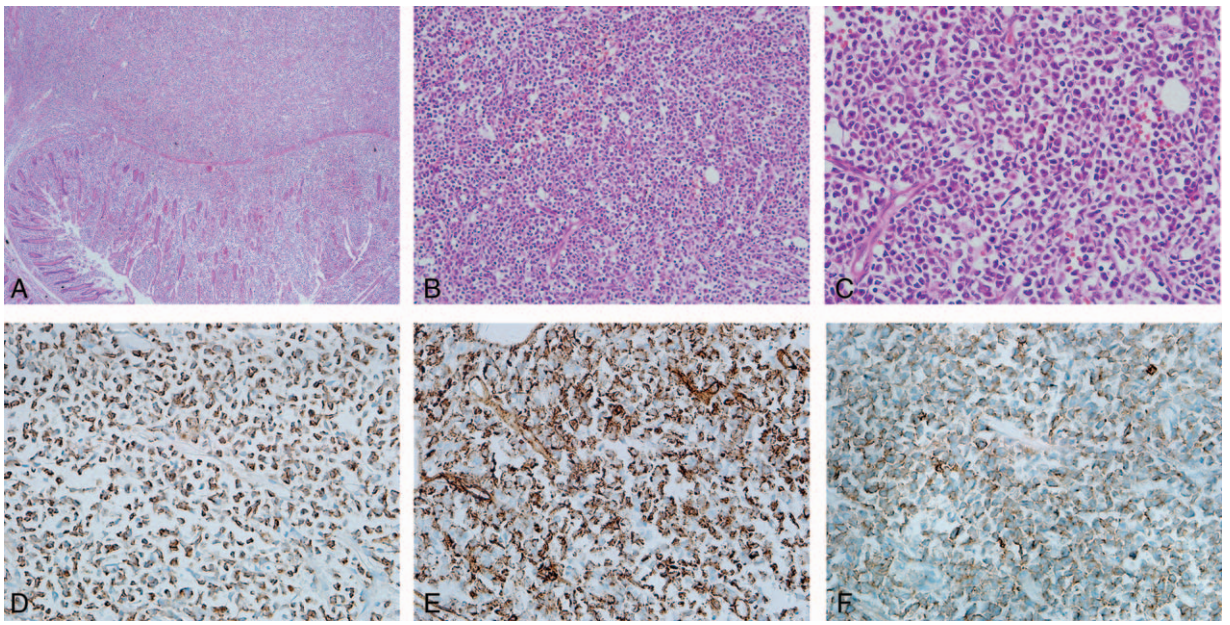

Figure 2. Pathologic analysis of the small intestinal tumor. (A–C) The tumor cells are diffusely dispersed in the intestinal wall, and the smooth muscle tissue is destroyed. Hematoxylin and eosin, A × 40, B × 200, C × 400. Immunohistochemistry shows positive staining of the tumor cells for myeloperoxidase (D), CD34 (E), and CD117 (F). (D–F) × 400. Staining was performed using the EliVision system (Maixin Biotech. Co. Ltd., Fuzhou, China).

A small bowel MS can appear as concentric wall thickening, an intraluminal or exophytic polypoid mass, or a combination of both, with variable contrast enhancement and a high predilection for mesenteric and peritoneal spread.[6–10] Initial CT in the present case showed concentric wall thickening of the jejunum involving the mesentery and mesenteric lymph nodes. Laparoscopic surgery was performed, but the patient refused additional treatment. Owing to disease progression, follow-up CT 2.5 months after surgery showed widespread infiltration of the small intestine, peritoneum, omentum, and mesentery. Moreover, the lesions in both kidneys were substantially larger and more numerous on follow-up CT versus initial CT. Six and a half months after conventional AML chemotherapy, the lesions were no longer detectable on CT, and slight atrophy of the renal parenchyma in the lesion areas was observed. Our initial results are similar to those of previous cases[6–10]; however, post-

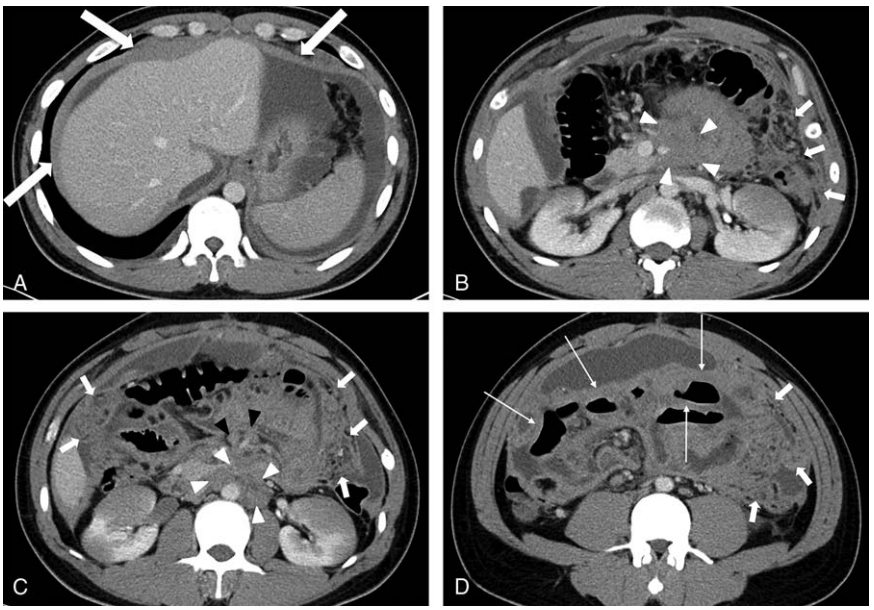

Figure 3. Follow-up postoperative transverse contrast-enhanced abdominal computed tomography before chemotherapy. The images show large amounts of ascites fluid, widespread infiltration of the peritoneum (A, thick long arrows), omentum (B–D, short arrows), and mesentery (B and C, white arrowheads), diffuse small bowel wall thickening (C, thin long arrows), and mesenteric lymph nodes enlargement (C, black arrowheads). Comparison of panels B and C with panels C and D in Fig. 1 shows that the lesions in both kidneys were substantially larger and more numerous in the follow-up versus initial CT images; hence, both kidneys were enlarged.

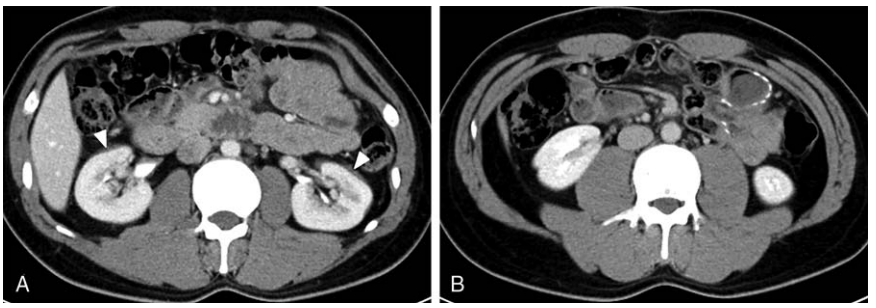

Figure 4. Follow-up postoperative transverse contrast-enhanced abdominal computed tomography after systemic chemotherapy. (A and B) The ascites fluid is completely absorbed, and the peritoneal, omental, and mesenteric lesions, the diffuse small bowel wall thickening, and enlarged mesenteric lymph nodes are absent. There are no lesions in either kidney, and the renal parenchyma of the lesion areas is slightly atrophied (A, arrowheads).

operative CT before and after chemotherapy was not performed in those studies as it was in ours. The main differential diagnosis of the small bowel MS in the present case is lymphoma, which typically exhibits an infiltrative pattern that causes diffuse bowel wall thickening; the thickening is moderately enhanced with or without aneurysmal dilation on CT images.[11,12] Small intestine MSs involving the mesentery have been reported in a few previous studies,[6,13,14] and the common CT finding is mesenteric lymphadenopathy.

To the best of our knowledge, renal MSs have been radiologically described in only 4 case studies.[15–18] They appear as soft tissue masses on plain CT[15] and as a well-defined homogeneously enhanced masses on enhanced CT.[16,17] Similarly, enhanced CT in our case showed well-defined mildly dilated lesions with progressive homogeneous enhancement. On T1- and T2-weighted magnetic resonance images, renal MSs are hypointense relative to the renal cortex with weak homogeneous gadolinium enhancement.[18] Based on these findings and those in our case, we suggest that homogeneous enhancement is an imaging feature of renal MS.[15–18]Differential diagnoses of renal MSs on CT mainly include insignificantly enhanced renal cell carcinomas, renal infarctions, and renal lymphomas. Unlike renal MSs, insignificantly enhanced renal cell carcinomas are usually singular and prone to hemorrhage, necrosis, and cystic changes in the lesions. Renal infarctions are usually wedge-shaped with no mass effect, and enhanced cortical rim signs representative of cortical rims and renal vessel abnormalities on the lesion side may be observed on contrast-enhanced CT images.[19] The CT characteristics of renal lymphomas include multiple lesions, which appear as masses with mild homogenous enhancement, and rare with hemorrhage, necrosis, cystic changes, and calcification in the lesions[20]; these characteristics complicate the differential diagnosis of renal MSs.

Treatments for MSs mainly include surgical resection, local radiotherapy, and systemic chemotherapy.[5,21] Surgical resection and local radiotherapy do not delay the transformation of MS to AML or improve prognosis.[5] Delay, however, can be achieved via chemotherapy, especially AML-type induction chemotherapy, which can also prolong the survival period. Timely diagnosis of MSs allows early treatment of MSs, which is associated with better event-free survival and overall survival rates compared with those for acute leukemia.[5,21] Therefore, a correct diagnosis, or at least differential diagnoses by radiologists that consider the lesion, may expedite appropriate therapy and prevent unnecessary surgeries. MSs have several radiological features. Enhanced homogenous soft tissue masses at multiple sites strongly suggest the presence of an MS, especially when a hematological disorder is present.[3,4,6,22,23]

5. Conclusions

As surgical treatment was chosen and the patient refused postoperative chemotherapy, the present MS case differs from previous MS cases and, therefore, provides important information. First, if the radiological examination shows lesions at multiple sites, and these lesions are soft tissue masses with homogenous enhancement, MS should be considered in the differential diagnosis, and an aspiration biopsy should be performed to provide a definitive pathological diagnosis. Second, if MS is diagnosed, systemic chemotherapy is crucial to recovery; otherwise, the disease may progress rapidly. Third, medical imaging is helpful in the differential diagnosis of MS, as well as in evaluating and monitoring the response of patients to treatments.

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