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CASE REPORT

장관의 골수성 육종 1예

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A Case of Myeloid Sarcoma of Intestine

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Myeloid sarcoma (MS) is an extramedullary involvement of immature myeloid proliferation. An isolated MS is defined as a myeloblastic tumor when it arises without any concomitant circulating disease. A diagnosis of MS is established using pathologic features including infiltration of myeloblasts and strong myeloperoxidase expression with negative cytokeratin immunohistochemical staining. We report a rare case of colonic MS without any peripheral blood abnormality. If the affected patient were left untreated, the MS could evolve into acute myeloid leukemia (AML) within one year. Several studies recommend the same regimens of chemotherapy as used for circulating AML to treat isolated MS. We focused on the diagnosis of MS in this study. The correct diagnosis of MS is important for adequate treatment. In conclusion, MS should be considered in the differential diagnosis of intestinal tumor. **(Korean J Gastroenterol 2016;68:148-151)**

Key Words: Sarcoma, myeloid; Colonic neoplasms

INTRODUCTION

Myeloid sarcoma (MS) is an extramedullary involvement of immature myeloid proliferation that is also referred to as granulocytic sarcoma or chloroma. An isolated MS is defined as a myeloblastic tumor that arises without any comorbid circulating disease, such as acute myeloid leukemia (AML) or bone marrow disease. However, sometimes MS can be the initial presenting manifestation of AML or a leukemic relapse.[1] The five year cumulative incidence of isolated extramedullary relapse is 9%.[2] About 40% to 47% of MS is misdiagnosed as lymphoma.[3,4] Therefore an accurate diagnosis is important. A diagnosis of MS is established using pathologic features including infiltration of myeloblasts and strong myeloperoxidase (MPO) expression with negative cytokeratin immunohistochemical staining. These findings differentiate MS from lymphoma or other non-hematopoietic tumors.

Isolated MS is a rare tumor whose description is largely based on case reports; only 345 cases were diagnosed between 1973 and 2010 in the United States.[5] MS is reported at every anatomic site, although the most common site of involvement is the soft tissue (27%). The rate of involvement of the gastrointestinal tract is only 8%.[5] Here, we report a rare case of colonic MS without any peripheral blood abnormality.

CASE REPORT

A previously healthy 55-year-old man who complained of

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abdominal fullness and dyspepsia visited our outpatient Department of Gastroenterology. On physical examination, abdominal distention was observed, and overall abdominal tenderness was present without rebound tenderness. A simple X-ray of the abdomen indicated gaseous distension of small bowel. To rule out mechanical obstruction, a CT scan was ordered. A mass of the terminal ileal area was observed on CT (Fig. 1), and multiple lymphadenopathy in both the para-aortic and aorto-caval areas was suspected. Endoscopic examination revealed a huge mass in the ascending colon (Fig. 2). The mass was a fragile exophytic tumor. Because of colonic obstruction, the endoscope could not pass the ascending colon.

To resolve the mechanical obstruction of the colon, a right hemicolectomy was performed. Grossly, the polypoid protruding mass involved the entire wall of the cecum and the adjacent attached mesentery, distorting the cecum; the size of the mass was 8.1×5.5×5.0 cm. Histologically, the cecum had been replaced by the diffuse infiltration of myeloblasts with little differentiation. The terminal ileum and the lymph nodes from pericolic, midcolic, and ileocolic areas were not involved. The tumor cells were large and morphologically similar to lymphoma cells, but they had more variable cytology. Many had an indented nucleus with fine chromatin (Fig. 3). There were also a few immature eosinophils. The neoplastic tumor cells were negative for cytokeratin and lymphoma markers (CD3, CD20, CD30, CD56, and EBER mRNA according to *in situ* hybridization) and positive for leukocyte common antigen, CD34, MPO, CD99, and CD117. The peripheral complete blood count was within normal limits: white blood cells, 7,900/mm^3; hemoglobin, 14.8 g/dL, and platelets, 371,000/mm^3. No blast cells were seen. Therefore, the patient was diagnosed with MS without peripheral blood abnormality. After a definitive diagnosis of MS was confirmed, he was referred to a hematologist for chemotherapy and bone marrow biopsy. However, he did not continue the treatment because he

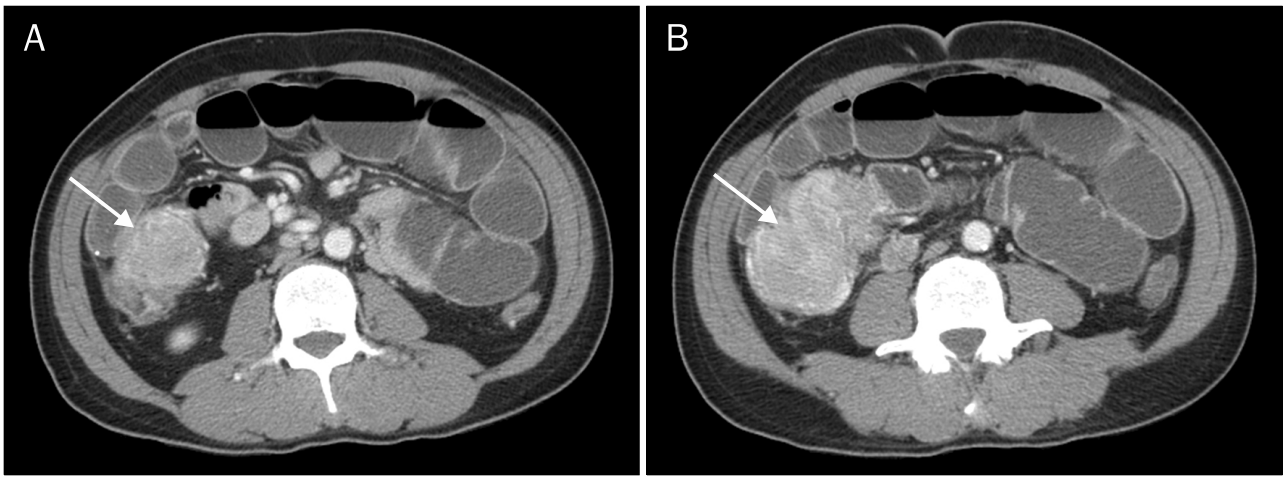

Fig. 1. The CT scan reveals an intraluminal annular enhancing mass (arrows) in the terminal ileum to proximal colon area with small bowel dilatation.

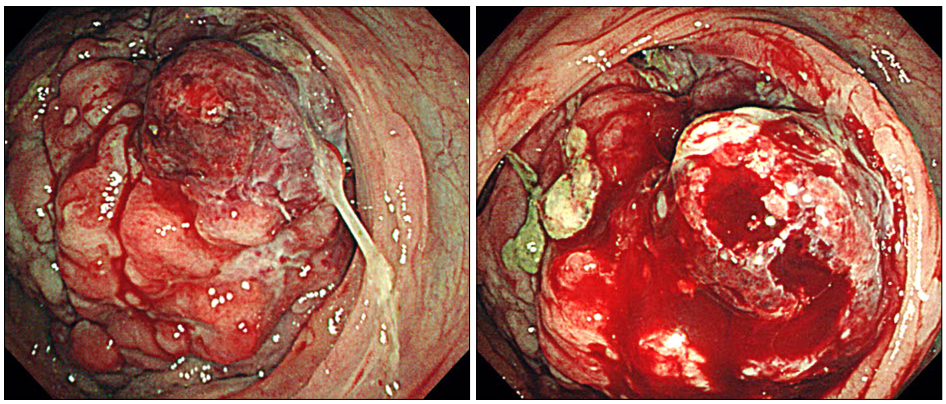

Fig. 2. A huge, obstructive mass of the ascending colon was found upon colonoscopy. The mass was a fragile exophytic tumor.

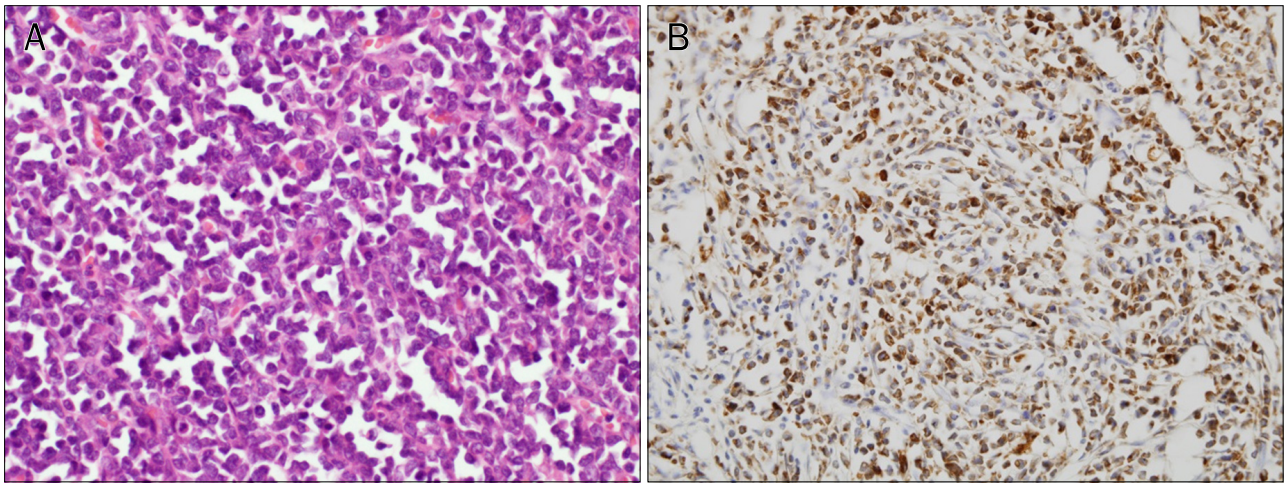

Fig. 3. Histology revealed myeloid sarcoma involving the colon. (A) Neoblastic cells have dispersed chromatin and inconspicuous nucleoli with an indented nuclear configuration (H&E, ×400). (B) The neoplastic cells are positive for myeloperoxidase (×400).

moved abroad.

DISCUSSION

Movassaghian et al.[5] analyzed 345 patients diagnosed with isolated MS between 1973 and 2010 using the SEER (Survival, Epidemiology, and End Results) database. The median overall survival time was eight months for patients with isolated MS in their study. When MS is suspected, evaluation for circulating diseases, bone marrow study, complete blood count and chromosomal analysis are needed. Neiman et al.[6] reported that, among 15 patients who showed no discernible hematologic abnormality at the initial diagnosis and received no chemotherapy, 13 (86.7%) developed acute non-lymphocytic leukemia (ANLL) at a mean of 10.5 months after the diagnosis of MS. Yamauchi and Yasuda[4] also reported 88% of MS patients developed ANLL without systemic chemotherapy, but 58% of MS patients given chemotherapy remained in the non-leukemic stage for more than 11 months. Although there are no treatment or follow up guidelines, several studies recommend the same regimens of chemotherapy as used for circulating AML to treat isolated MS.[1,4,7] Event-free survival was longer in the patients with isolated MS treated with cytarabine containing regimens.[1] Relapse rates are higher and overall survival is poor when treatment is limited to surgical or radiation therapy.[4,8]

Few cases of isolated MS arising from the gastrointestinal tract without circulating or bone marrow disease have been reported.[9] The ileum is the most affected area in the intestine with MS,[10] while isolated primary involvement of the colon and rectum is exceedingly rare with secondary extension from the peritoneum being more common. The specific endoscopic manifestation of MS is unclear. Several reported cases of MS presented as an intraluminal polypoid mass, diffuse polyposis, or rarely, coexisting with adenoma.[11,12] Our case is one of the few cases that presented as large polyp in the ascending colon.

Our patient was a previously healthy man with a normal peripheral blood count. However, the lack of bone marrow aspiration results was a major concern; the patient did not receive further management. So it is unclear whether it was a *de novo* MS or not. Because the clinical diagnosis of MS as an initial manifestation is challenging, we focused on the diagnosis of MS in this case. MS is typically diagnosed based upon pathologic and immune-phenotypic features that are consistent with a high-grade hematopoietic neoplasm with the expression of MPO or other markers of myeloid differentiation. It is important to distinguish MS from lymphoma or other tumors. In immunohistochemistry, CD68/KP1 was the most commonly expressed marker (100%), followed by MPO (83.6%), CD117 (80.4%), CD99 (54.3%), CD68/PG-M1 (51.0%), CD34 (43.4%).[3] With conventional light microscopy, the picture can be misdiagnosed as lymphoma or even as a non-hematopoietic tumor. As this leads to a wrong treatment, immune-phenotying is mandatory.[3] In conclusion, the intestinal mass could be an initial manifestation of AML, a sign of AML relapse, or just isolated MS. In stem cell transplant recipients, MS must be considered in the differential diagnosis of

colorectal tumor.

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