Case Report
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Imaging Findings of Isolated Myeloid Sarcoma of the Stomach in a Nonleukemic Child: A Case Report and Literature Review
비 백혈병 환아에서 위의 단일 과립세포육종의 영상소견: 증례 보고

Yong Kyun Kim, MD[1], Jung Hyun Kim, MD[1], Hee Jo Baek MD[1], Jin Woong Kim, MD[2], Sang Soo Shin, MD[2], Suk Hee Heo, MD[1]*

[1]Department of Radiology, Chonnam National University Hospital, Chonnam National University Medical School, Hwasun, Korea
[2]Department of Radiology, Chonnam National University Hwasun Hospital, Chonnam National University Medical School, Hwasun, Korea

Myeloid sarcoma is an extramedullary solid neoplasm composed of myeloid precursor cells. This tumor usually occurs simultaneously with or following the onset of acute leukemia. Rarely, it can be the first manifestation of acute myeloid leukemia. The tumor can occur anywhere in the body. However, primary involvement of the stomach without evidence of leukemia is exceedingly rare, and to the best of our knowledge, imaging features of isolated myeloid sarcoma of the stomach have not been reported in children. This case illustrates the imaging appearances of isolated myeloid sarcoma that initially manifested as gastric submucosal wall thickening and discusses the differential diagnosis, in a 15-year-old girl without evidence of hematologic malignancy.

Index terms
Myeloid Sarcoma
Stomach
Multimodal Imaging

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***Corresponding author:** Suk Hee Heo, MD
Department of Radiology, Chonnam National University Hwasun Hospital, 322 Seoyang-ro, Hwasun-eup, Hwasun-gun 58128, Korea.
Tel. 82-61-379-7112 Fax. 82-61-379-7133
E-mail: sheo@jnu.ac.kr

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INTRODUCTION

Myeloid sarcoma (MS), also known as chloroma, granulocytic sarcoma or extramedullary myeloblastoma, is a collection of myeloid precursor cells that include myeloblasts, promyelocytes, and myelocytes (1, 2). MS usually occurs in the course of acute or chronic myeloid leukemia or leukemic transformation in myeloproliferative or myelodysplastic disorders. But it has rarely been reported as an isolated manifestation in non-leukemic patients (1). MS has been found in almost every anatomic location, and it more commonly occurs in the skin, bone, soft tissue, lymph nodes and central nervous system (CNS) (1-3). However, involvement of the gastrointestinal tract, especially the stomach, is uncommon, and radiologic findings of stomach involvement have been rarely reported in the literature (4). We report the imaging features, including those of ultrasonography, CT and positron emission tomography (PET)-CT, in an unusual case of isolated MS of the stomach in a 15-year-old girl without evidence of any hematologic disorder.

CASE REPORT

A 15-year-old girl was admitted to our hospital with intermittent epigastric pain for 3 months. She had no specific medical or surgical history. Physical examination revealed mild tenderness in the epigastric and left upper quadrant areas of the abdomen. There was no hepatosplenomegaly or lymphadenopathy. Laboratory evaluations showed a white blood cell count of 6300/mm^3, a hemoglobin level of 10.9 g/dL, and a platelet count of 337000/mm^3. Serum lactate dehydrogenase level was 498 IU/L,

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which was slightly elevated.

An abdominal ultrasonogram (GE LOGIQ 9; GE Healthcare, Milwaukee, WI, USA) showed marked hypoechoic wall thickening of the gastric body (Fig. 1A). Abdominal CT (LightSpeed VCT; GE Healthcare) revealed marked and eccentric wall thickening with homogeneous contrast enhancement in the stomach, particularly the lesser curvature of the body portion (Fig. 1B, C). Mucosa of the thickened gastric wall and perigastric fat planes were smooth and preserved. There was no perigastric or distant lymphadenopathy. ^{18}F-fluorodeoxyglucose (FDG) PET-CT (GE Discovery 600; GE Healthcare) was performed for further staging because the presumptive diagnosis was a lymphoma. On the PET-CT images, increased ^{18}F-FDG was found only in the lesser curvature side of the gastric body with a maximal standardized uptake value (SUV) of 15.2, suggestive of a malignant lesion (Fig. 1D). There was no other abnormal FDG uptake lesion. Upper GI endoscopy was performed for biopsies, which showed extensive enlarged and hyperemic fold thickening with scattered hemorrhagic spots in the lesser curvature of the gastric body (Fig. 1E). Histopathological findings from the endoscopic biopsy specimens were consistent with MS based on the presence of cells of the myeloid lineage with positive immunohistochemical staining for myeloperoxidase and myeloid markers (CD 34, CD 43, CD 117), and negative immunohistochemical staining for B-cell (CD 20) and T-cell markers (CD 3 and CD 30) (Fig. 2). Bone marrow examination demonstrated no evidence of marrow involvement with acute myeloid leukemia (AML) blasts.

The patient received cytarabine-based induction and consolidation chemotherapy. Unfortunately, 6 months later, the patient

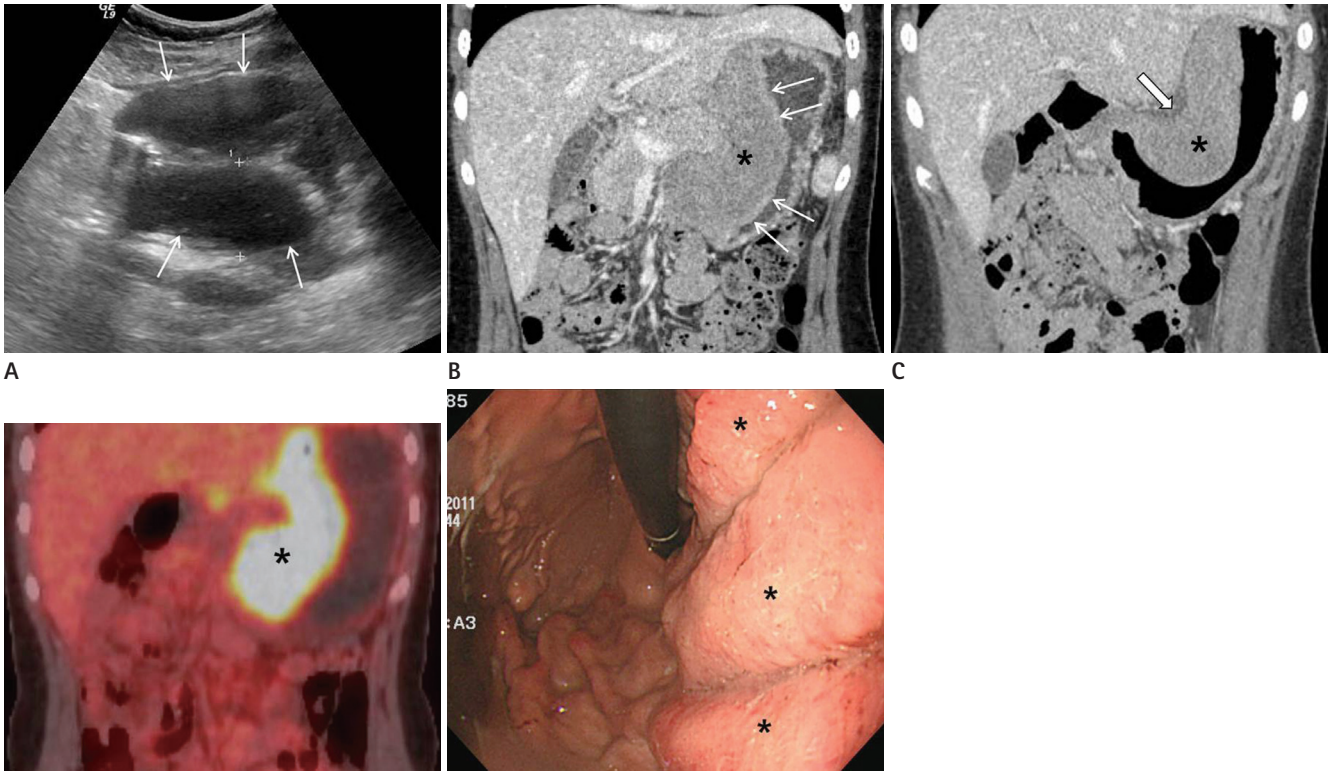

Fig. 1. Multimodal imaging of isolated myeloid sarcoma in a nonleukemic child.
A. Oblique axial gray-scale ultrasonogram shows marked and hypoechoic thickening of the stomach wall (arrows) with loss of stratification.
B, C. Coronal reformatted, contrast-enhanced CT images demonstrate eccentric, marked and homogeneous wall thickening (asterisks) in the lesser curvature of the gastric body. Mucosa of the thickened gastric wall (thin arrows) and perigastric fat planes (thick arrows) are smooth and preserved, which are suggestive of submucosal disease involvement. There is no regional or distant lymphadenopathy.
D. Coronal ^{18}F-FDG PET-CT image shows a diffuse hypermetabolic area (maximal standardized uptake value of 15.2) corresponding to the thickened gastric wall (asterisk).
E. Upper GI endoscopy reveals extensive enlarged and hyperemic fold thickening (asterisks) with scattered hemorrhagic spots.
GI = gastrointestinal, ^{18}F-FDG PET-CT = ^{18}F-fluorodeoxyglucose PET-CT

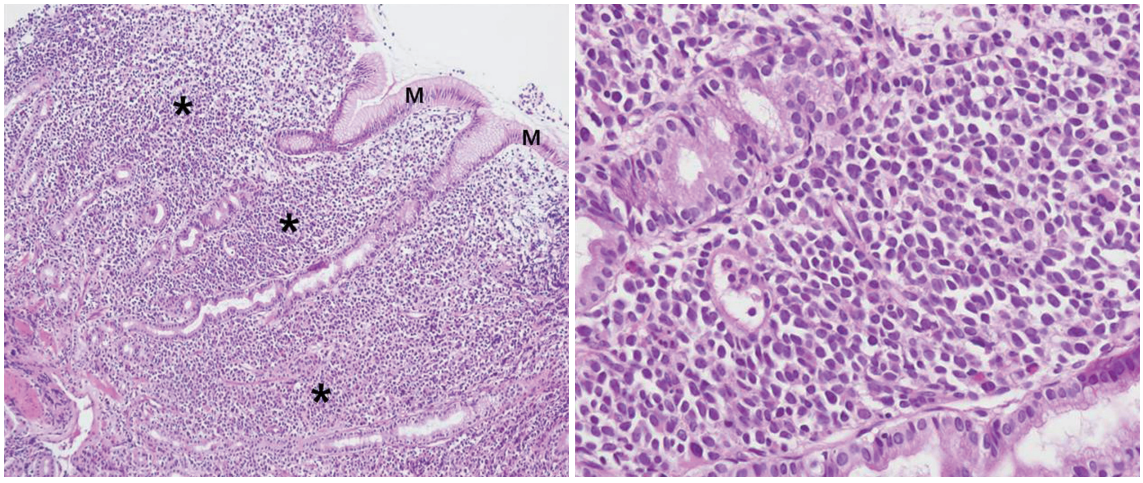

Fig. 2. Histopathologic examination of isolated myeloid sarcoma in a nonleukemic child.
A. Photomicrograph (hematoxylin-eosin stain, × 100 magnification) shows diffuse infiltrates of leukemic blasts and some maturing granulocytes (asterisks) in the submucosal tissue and intact overlying mucosa (M) of the stomach.
B. Microscopic image (hematoxylin-eosin stain, × 400 magnification) demonstrates blastic, hyperchromatic and pleomorphic cells of the granulocytic series, consistent with granulocytic sarcoma.

noted relapse in the stomach, breast and peritoneum and showed AML M4 type with a blast count of 33% on bone marrow examination. Second-line chemotherapy and radiation therapy were applied quickly, but the patient died due to disease dissemination and septic shock.

DISCUSSION

MS is a rare, malignant localized solid tumor composed of immature granulocytic precursor cells occurring in extramedullary sites. In the majority of cases, it is diagnosed at presentation or during the course of AML. The overall prevalence of MS was 2.5–8% in one autopsy series of acute leukemia (1). In a multicenter study of 1832 children with AML, 199 (10.9%) patients had MS and only 13 (0.7%) patients had isolated MS without involvement of the bone marrow (2). Another literature has reported that isolated MS without any blood or bone marrow involvement at the time of diagnosis is a rare disease with an incidence of 2/1000000 in adults (5). MS can occur virtually anywhere in the body; the most common sites are skin, orbit, head and neck, bone and CNS (1, 3). MS involving the gastrointestinal tract is relatively rare and it occurs mostly in the small bowel (1, 4). The involvement of the stomach is very rare, and to the best of our knowledge, the imaging features of isolated MS of the stomach have not been reported in children.

There are a few reports on the radiological features of MS in the gastrointestinal tract and the imaging features of MS are variable; lesions can appear as an intramural or exophytic polypoid mass or bowel wall thickening or a combination of these manifestations (4, 6). In our case, the wall of the stomach was markedly thickened and showed homogeneous contrast enhancement. The gastric wall thickening was eccentric, and it was not circumferential. Overlying mucosa of the thickened gastric wall and perigastric fat planes were smooth on CT and gastric fold was preserved on gastroscopy, suggestive of submucosal disease involvement. There was no regional or distant lymphadenopathy. In ^{18}F-FDG PET-CT images, the thickened gastric wall showed diffuse hypermetabolism with a maximal SUV of 15.2. These imaging features of the gastric lesion were quite similar to those of a primary gastric lymphoma and these features cannot be distinguished from those of a primary gastric lymphoma, although circumferential wall thickening and regional or mesenteric lymphadenopathy are frequently seen in the majority of lymphomas (7). Inflammatory myofibroblastic tumor and gastrointestinal stromal tumor (GIST) should also be included in the differential diagnosis because both gastric lesions may show a well-defined exophytic or submucosal mass (8, 9). The gastric inflammatory myofibroblastic tumor shows various appearances, but it usually shows aggressive features such as ulceration, adjacent wall infiltration and extraluminal extension,

which were not seen in this case (8, 10). Gastric GIST is an uncommon mesenchymal tumor in adolescents and children and it appears as a submucosal mass of varying size with normal overlying mucosa. But, GIST usually shows central necrosis, cavitation, cyst formation, and hemorrhage, when it presents as a large tumor (9).

In an unusual manifestation like our case, in which the tumor precedes the hematologic evidence of leukemia, it may be impossible to diagnose MS of the stomach initially and it can lead to diagnostic challenges. This case illustrates the radiologic findings including those of ultrasonography, CT, and PET-CT of isolated MS that presented as eccentric and smooth wall thickening of the stomach without regional or distant lymphadenopathy.

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비 백혈병 환아에서 위의 단일 과립세포육종의 영상소견: 증례 보고

김용균[1] · 김정현[1] · 백희조[1] · 김진웅[2] · 신상수[2] · 허숙희[1]*

골수성육종은 골수성 전구세포들로 구성된 골수 외 고형암이다. 이는 대부분 급성 백혈병과 병발하거나, 급성 백혈병 발병 후에 발생하며 급성 골수성 백혈병 발병 전에 선행하여 첫 번째 징후로 나타나는 경우는 매우 드물다. 골수성육종은 우리 몸 어느 곳에서나 발생할 수 있으며 피부, 뼈, 연부조직과 중추신경계에 주로 발생한다. 위장관계의 침범, 특히 위의 원발성 침범은 극히 드물고 특히 소아에서 급성 골수성 백혈병에 선행하는 단일 위 골수성육종에 대한 영상소견은 지금까지 잘 알려져 있지 않다. 이 증례는 기저질환으로 혈액종양이 없는 15세 여자 환자에서 위벽의 비대칭적 점막하 비후로 나타난 단일성 위 골수성육종의 초음파, 전산화단층촬영 및 PET-CT를 포함하는 영상소견과 감별진단에 대해 기술하고자 한다.

[1]전남대학교 의과대학 전남대학교병원 영상의학과, [2]전남대학교 의과대학 화순전남대학교병원 영상의학과