

MRI Findings of Extramedullary Hematopoiesis of the Spleen in Patient with Idiopathic Myelofibrosis : 2 Case Report¹

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MRI findings of extramedullary hematopoiesis of the spleen have not been described in the literature. We report the MRI features of this condition, as seen in two patients and confirmed by fine needle biopsy. Three small masses (≤ 3 cm) were isointense on T1WI, hyperintense on T2WI, and enhanced after the injection of gadolinium. Two 6cm-sized masses were hypointense on both T1WI and T2WI, and showed no contrast enhancement.

Index words : Spleen, MR
Spleen, diseases

Idiopathic myelofibrosis is characterized by bone marrow fibrosis and extramedullary hematopoiesis(1). Microscopically, extramedullary hematopoiesis is usually in the form of diffuse infiltration of hematopoietic cells, resulting in organomegaly, although there may be tumor-like masses of hematopoietic tissue(2, 3). Focal masses involving the spleen, which on ultrasound(2, 4, 5) and CT(3, 6) may be seen to mimic a neoplasm, have been reported. To our knowledge, however, the MRI feature of extramedullary hematopoiesis in the spleen has not been described. We report the MRI findings of focal lesions in the spleen associated with idiopathic myelofibrosis.

Case Report

Case 1

A 38-year-old man was admitted to hospital because of a palpable abdominal mass and anemia. The results of analysis were hemoglobin, 10.7g/dl; platelet, 208,000/mm³; and white cells, 4200/mm³; white blood cell differential count showed 1% blast, 2% myelocyte, 15% metamyelocyte, 25% band neutrophil, and 24%

polymorphonuclear leucocyte. Peripheral blood smear showed anisocytosis, poikilocytosis and teardrop-shaped red blood cells, and a shift of neutrophils to the left. Bone marrow aspiration biopsy showed medullary fibrosis (grade IV) and an increased number of megakaryocytes.

Ultrasound revealed massive splenomegaly and three (two homogeneous echogenic, and one mixed echogenic) focal lesions in the spleen, while two enhancing nodular lesions were seen on CT.

On T1-weighted imaging(Fig. 1A), only one large lesion, 6cm in diameter, of slightly low signal intensity was seen; it showed low SI on T2WI(Fig. 1B). Three more small lesions (≤ 3 cm) of high signal intensity were seen on T2-weighted images(Fig. 1C). After gadolinium injection, three small lesions were enhanced (Fig. 1D), but the large lesion was not.

Ultrasound-guided fine needle biopsy was performed on the large lesion and on one small one. Microscopic examination revealed a similar degree of fibrosis and hemosiderin deposit in both lesions, and the pathologic diagnosis was extramedullary hematopoiesis.

Case 2

CT in a 69-year-old man with idiopathic myelofibrosis showed a faintly enhancing 6cm-sized lesion in the spleen; T1- and T2-weighted imaging(Figs. 2A and 2B, respectively) showed it to be hypointense. The pathological diagnosis, subsequent to needle biopsy,

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was extramedullary hematopoiesis.

Discussion

Idiopathic myelofibrosis (agnogenic myeloid metaplasia; myelofibrosis with myeloid metaplasia) is a chronic myeloproliferative disorder of unknown etiology primarily affecting adults. It is characterized by fibrosis and proliferation of all three hematopoietic cell lines in the bone marrow, a leukoerythroblastic peripheral blood smear with teardrop-shaped red blood cells, and progressive hepatosplenomegaly due to extramedullary hematopoiesis(2).

Extramedullary hematopoiesis in patients with myelofibrosis invariably affects the reticuloendothe-

lial organs, mainly the liver and spleen. Other less frequent sites of involvement include the lymph nodes, adrenal glands, thorax, cranial vault, retroperitoneum, lymphatics, breasts, spinal canal, renal pelvis, thymus, and the heart(2, 4–8). Microscopically, there is diffuse infiltration of hematopoietic cells, although there may be tumor-like masses of hematopoietic tissue with a variable degree of fibrosis(4, 6).

Imaging modalities have allowed evaluation of extramedullary hematopoiesis in various organs. Ultrasound findings of focal form are known to be fairly well defined solid masses of variable echogenicity, but usually echogenic(2, 4, 5, 7). Fat and fibrosis are reported to be related to the increased echogenicity of the lesions(2, 4, 7). On CT, there may be solid, well-

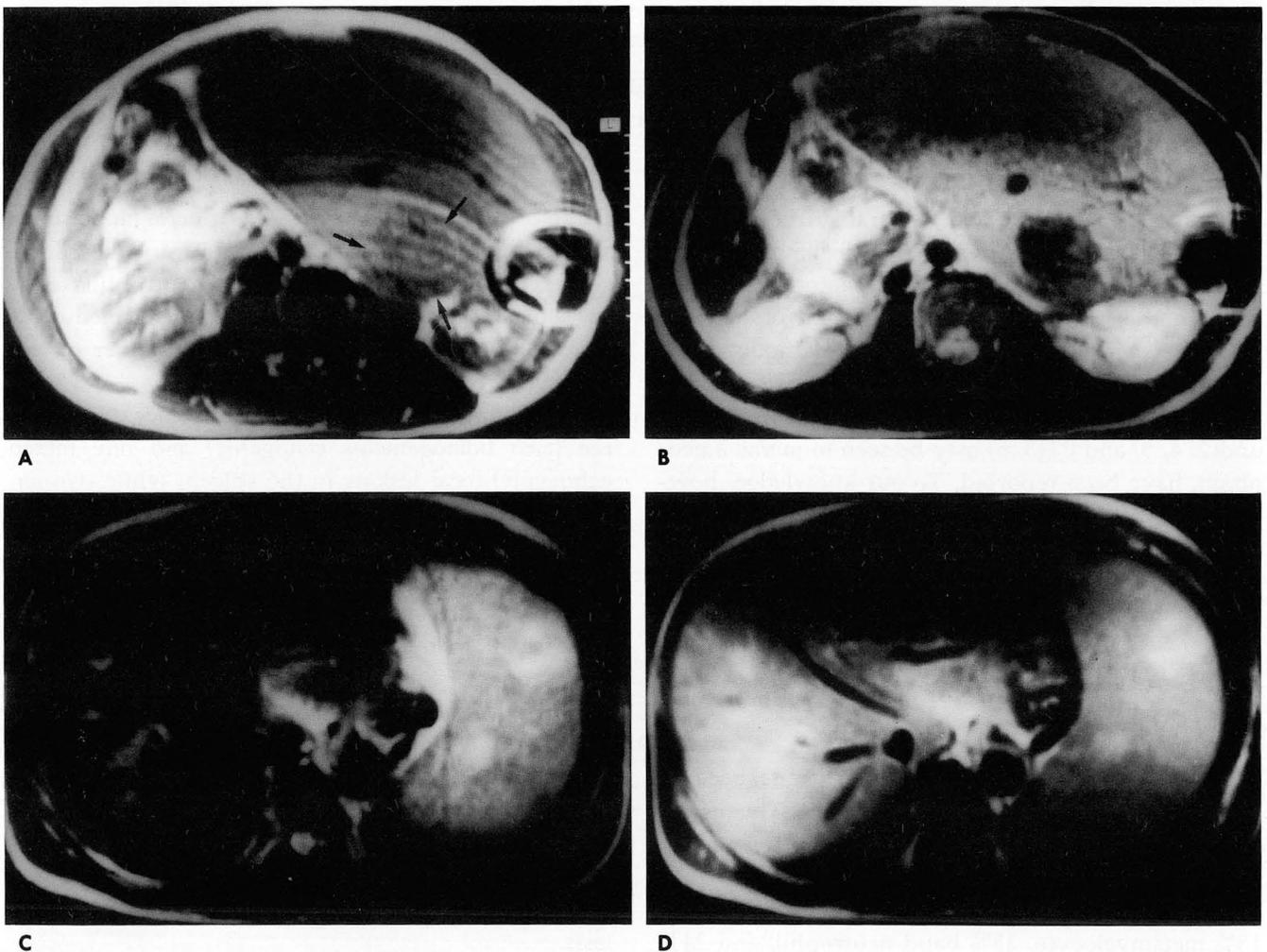


Fig. 1. Extramedullary hematopoiesis of the spleen in 38-year-old man with myelofibrosis.

- A.** T1-weighted image (TR/TE, 600msec/17 msec) shows a 6cm sized, slightly low signal intensity lesion(arrows), in the lower pole of enlarged spleen.
- B.** Same lesion in the lower pole is noted as low signal intensity on T2-weighted image (TR/TE, 3300msec/140msec).
- C.** T2-weighted image (TR/TE, 3300msec/140msec) shows two lesions of high signal intensity with diameters less than 3cm in the upper pole of spleen. Another small lesion of high signal intensity was also noted (not shown).
- D.** Post-Gadolinium enhanced T1-weighted image (TR/TE, 600msec/17msec) shows contrast enhancement of the upper pole lesions. Lower pole lesion was not enhanced (not shown).

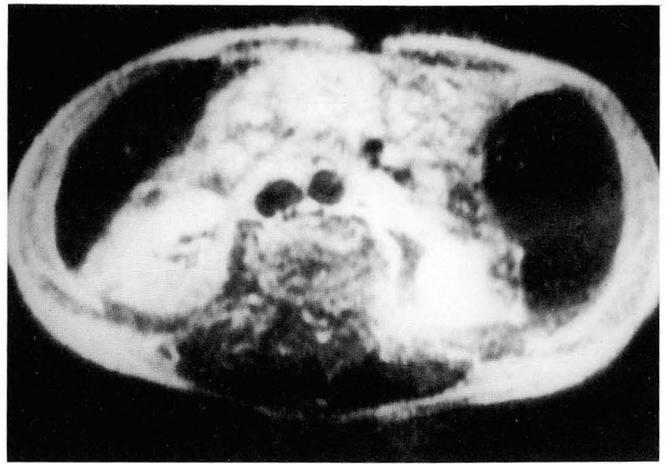
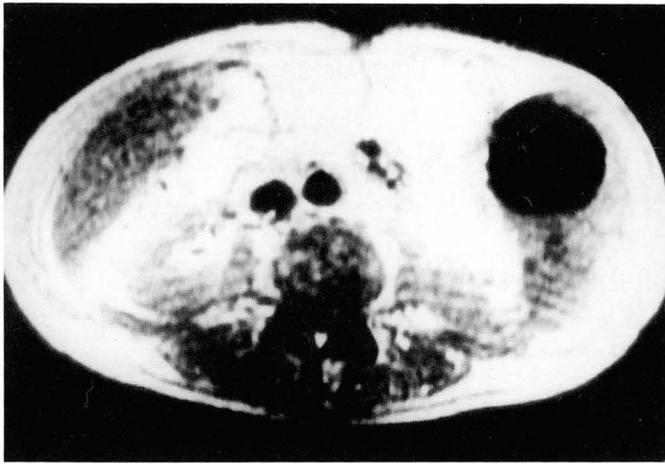


Fig. 2. Extramedullary hematopoiesis of the spleen in 69-year-old man with myelofibrosis.

A. T1-weighted image (TR/TE, 500msec/20msec) shows 6cm-sized focal lesion of low signal intensity.

B. This lesion is demonstrated as low signal intensity on T2-weighted image (TR/TE, 2000msec/80msec).

marginated, dense soft tissue masses(3, 6). Ultrasound and CT findings are usually non-specific, and on the basis of imaging, extramedullary hematopoiesis cannot be differentiated from malignancy(2-4). MR findings of extramedullary hematopoiesis in the spleen have not been reported, although the MR appearance of intrahepatic extramedullary hematopoiesis has been noted (9). In this case, the masses were of intermediate signal intensity on T1-weighted images, and mildly hyperintense relative to the remaining liver parenchyma on T2-weighted images; one lesion showed heterogeneous enhancement on gadolinium-enhanced T1-weighted images, while another showed no contrast enhancement.

In case 1, MRI detected more lesions(four) than ultrasound(three) or CT(two). In both cases, two large lesions, 6cm in diameter, showed low signal intensity on both T1-weighted and T2-weighted images, and there was no contrast enhancement after gadolinium injection. Three small lesions (≤ 3 cm) were not detected by T1-weighted imaging, but were seen as high signal-intensity masses on T2-weighted images. These small lesions were enhanced after gadolinium injection. Fibrosis and hemosiderin deposits were documented on microscopic examination, but on the basis of the small pieces of biopsy specimen, pathologic correlation with MRI findings was not possible.

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특발성 골수섬유증 환자에서 비장에 종괴를 형성한 골수외조혈의 자기공명영상 소견 : 2예 보고¹

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특발성 골수섬유증 환자에서 비장에 종괴를 형성한 골수외조혈의 자기공명영상 소견은 문헌에 보고된 적이 없다. 저자들은 세침생검술로 확진된 2예의 비장에 종괴를 형성한 골수외조혈의 자기공명영상 소견을 보고한다. 2명의 환자에서 5개의 종괴가 발견되었고, 그중에서 3cm 이하의 3개의 종괴들은 T1강조영상에서는 동등신호를, T2강조영상에서는 고신호를 나타내었으며, 조영증강은 잘 되었다. 6cm 크기의 두 종괴는 T1과 T2강조영상에서 모두 저신호로 보였고, 조영증강은 되지 않았다.