

MR imaging of focal splenic extramedullary hematopoiesis in polycythemia vera: case report

T. Gabata,¹ M. Kadoya,¹ A. Mori,² S. Kobayashi,¹ J. Sanada,¹ O. Matsui¹

¹Department of Radiology, Kanazawa University, School of Medicine, 13-1, Takara-machi, Kanazawa City, 920-8641, Japan

²Department of Surgery, Minamigaoka Hospital, 2-125 Magae, Kanazawa City 921-8141, Japan

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Abstract

We report a case with focal mass-like lesion of the spleen in polycythemia vera. Magnetic resonance imaging of the lesion showed homogeneous hyperintensity on T2-weighted images, with progressive enhancement on dynamic magnetic resonance images after bolus injection of gadolinium. The magnetic resonance features of focal splenic extramedullary hematopoiesis have not been previously reported.

Key words: Spleen—Hematopoiesis—Magnetic resonance imaging.

Extramedullary hematopoiesis (EMH) is a compensatory response to deficient bone marrow blood cell production, leading to the production of myeloid and erythroid elements at ectopic sites. EMH is most common in patients with marked destruction of red blood cells such as congenital hemolytic anemia, including thalassemia major, sickle cell anemia, and hereditary spherocytosis. EMH is also seen in patients who have organs that cannot form blood normally to produce erythrocytes, in conditions such as myelofibrosis, polycythemia vera, and carcinomatous or lymphomatous replacement of bone marrow (leukemia, Hodgkin disease) [1].

The sites of EMH include the spleen, liver, lymph nodes, adrenal glands, mediastinum (paraspinal), heart, thymus, lung, renal pelvis, retroperitoneum, gastrointestinal lymphatics, and dura mater of cranium and spine [1]. Although foci of EMH are commonly seen in spleen and liver parenchyma microscopically, focal mass lesions of EMH in the liver and spleen are rare [2–7].

Whereas the computed tomographic (CT) and ultrasonographic findings of focal splenic EMH have been reported, magnetic resonance (MR) imaging findings in intrasplenic EMH have not. We present the MR imaging findings of a focal mass-like extramedullary hematopoiesis of the spleen in a patient with polycythemia vera.

Case report

A 73-year-old woman was admitted to undergo surgery for carcinoma of the transverse colon. She had been diagnosed with liver cirrhosis and polycythemia vera, for which she frequently received bloodletting. Red blood cell count and hemoglobin levels were $510 \times 10^9/\text{mm}^3$ and 8.8 g/dL, respectively.

Unenhanced CT of the abdomen showed marked splenomegaly without focal abnormality. Fat-suppressed turbo spin-echo T2-weighted MR images and half-Fourier single-shot turbo spin echo (HASTE) T2-weighted images showed splenomegaly with a hyperintense mass, 5 cm in diameter (Fig. 1A). Unenhanced gradient-echo T1-weighted images showed slight hypointensity (Fig. 1B). The mass showed progressive enhancement on gadolinium-enhanced dynamic MR images (Figs. 1C,D).

The patient underwent colonic resection and a needle biopsy of the splenic mass under direct visualization. The microscopic diagnosis was extramedullary hematopoiesis (Fig. 1E).

Discussion

EMH predominantly affects the spleen and liver. Although it usually shows diffuse infiltration microscopically, there may be focal mass-like involvement of the liver and spleen [2–7]. Some studies have reported on the MR findings of EMH at various sites. The hematopoietic mass is usually visualized with intermediate intensity on T1-weighted images and high intensity on T2-weighted images in relation to muscle, with slight enhancement shown after infusion of gadolinium [8–10]. However,

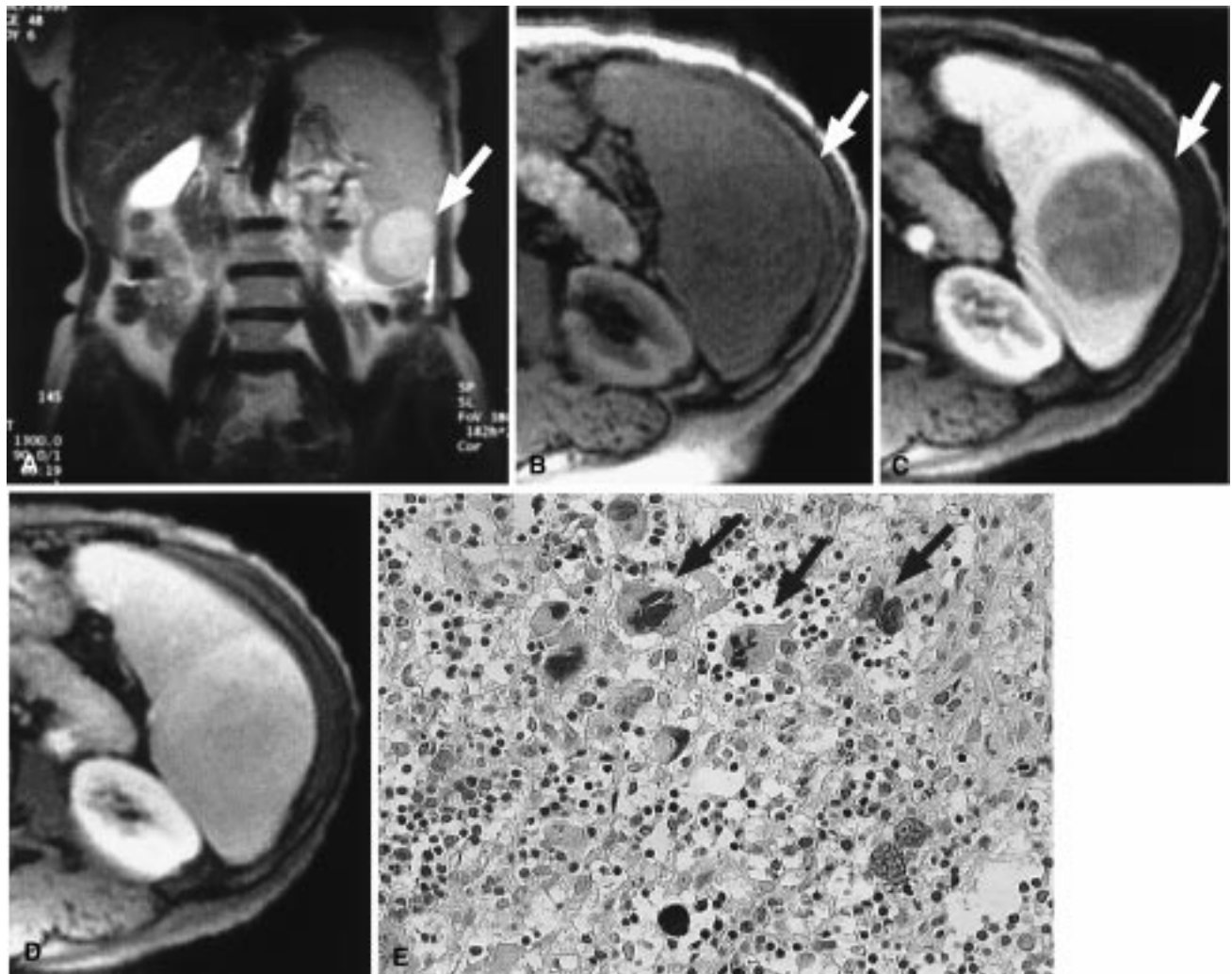


Fig. 1. Extramedullary hematopoiesis of the spleen in a 73-year-old woman. **A** Half-Fourier single-shot turbo spin echo (HASTE) T2-weighted image (TR = 1300 ms, TE = 90 ms) shows hepatosplenomegaly, with a hyperintense round mass, 5 cm in diameter, within the lower pole of the spleen (*arrow*). **B** Unenhanced gradient echo T1-weighted image (TR = 157 ms, TE = 4.7 ms, FA = 90°) shows slight hypointensity in relation to the spleen (*arrow*). **C** On the arterial dom-

inant phase of gadopentetate dimeglumine-enhanced dynamic MR images, the mass shows enhancement to a slight degree (*arrow*). **D** On the venous phase, the mass shows isointensity in relation to the spleen because of the prolonged enhancement. **E** Microscopic diagnosis is extramedullary hematopoiesis including magakaryocytes (*arrows*), erythroblasts, and immature granulocytes. HE stain $\times 100$.

Mesurole et al. reported a hematopoietic mass showing hypointensity on T2-weighted MR images [11].

The signal intensity of the mass depends on the evolution of the hematopoiesis. Tsitouridis et al. [12] showed that active recent hematopoietic lesions have immature and mature cells consisting mainly of erythroid and myeloid cells, whereas inactive older lesions have fatty tissue and massive iron deposition. Active hematopoietic lesions show intermediate signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and some enhancement after contrast medium injection. Older inactive lesions may show low signal intensity on both T1- and T2-weighted images due to iron

deposition or high signal intensity on both sequences due to fatty infiltration. Older lesions with fatty infiltration or iron deposition may show no enhancement.

In our case, the hematopoietic mass of the spleen showed low signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and progressive enhancement on dynamic MR images. These signal intensity and enhancement patterns indicated an active splenic hematopoietic lesion, which was confirmed pathologically.

The differential diagnosis of focal splenic mass lesions includes hemangioma, hamartoma, lymphangioma, lymphoma, metastasis, and angiosarcoma. Although focal

splenic mass lesion is rare, EMH should be included in the differential diagnosis, particularly if a hematological disorder is present.

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