International Journal of Radiology and Diagnostic Imaging

E-ISSN: 2664-4444
P-ISSN: 2664-4436
www.radiologypaper.com
IJRDI 2024; 7(2): 51-52
Received: 22-02-2024
Accepted: 26-03-2024

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Focal extramedullary hematopoiesis in the spleen: An uncommon MRI presentation

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DOI: https://doi.org/10.33545/26644436.2024.v7.i2a.383

Abstract
In this article, we present a case study of a 51-year-old male patient with thrombocytopenia, showcasing a rare magnetic resonance (MR) imaging presentation of focal extramedullary hematopoiesis (EMH) in the spleen. The diverse MR signal characteristics of EMH, influenced by the presence of fat and the degree of hematopoietic activity, are well-documented. However, the observed delayed central enhancement in our patient's MR imaging is novel and, to our knowledge, has not been previously reported. Following a detailed pathological examination, we propose that the presence of hypocellular collagenous stroma in the central zone of the EMH lesion could explain this unique enhancement pattern.

Keywords: Focal extramedullary hematopoiesis, spleen, MRI

Introduction
Extramedullary hematopoiesis (EMH) occurs as a compensatory mechanism when the bone marrow's ability to produce blood cells is inadequate, leading to its manifestation in various organs. However, its appearance as a localized mass within the spleen is uncommon. Prior studies have shown that such focal splenic masses from EMH may exhibit minimal to no enhancement following the administration of contrast material. In this report, we describe a case of EMH confirmed through pathology, where a single mass in the spleen was isointense on T_1-weighted scans and slightly hypointense on T_2-weighted scans. The MRI with contrast enhancement showed that this hypervascular splenic lesion presented with delayed enhancement resembling a central scar. To the best of our knowledge, this distinctive pattern of enhancement in EMH on MRI, paired with pathological evidence, has not been previously documented.

Case report
A 51-year-old male with a history of hypertension was admitted to the hospital following two weeks of developing petechiae and ecchymoses on both his forearms and lower legs, accompanied by severe thrombocytopenia detected in an outpatient setting (White blood cell count: 8000/mm^3; hemoglobin: 9.6 g/dL; platelets: 4 x 10^9/L; prothrombin time: 11.4/11.5 seconds; partial thromboplastin time: 22.8/31.5 seconds). Despite unremarkable further laboratory results except for ongoing thrombocytopenia of unidentified cause, he was treated with a blood transfusion. A subsequent bone marrow biopsy showed normal cellularity with enough megakaryocytes, erythroid, and myeloid cells, leading to a diagnosis of idiopathic thrombocytopenic purpura.
The patient underwent several imaging tests, including MRI. Sonography identified a clearly defined, solitary hyperechoic lesion in the spleen measuring approximately 5 cm. MRI imaging characterized the lesions as a round, thin-walled encapsulated mass within the spleen. It was isointense in T_1-weighted images and slightly hypointense in T_2-weighted images. Post-gadolinium, the tow lesions showed consistent enhancement across arterial, portal venous, and delayed phases, except for its central zone, which did not enhance during the arterial phase but showed delayed contrast filling in later phases. Given these findings, the initial differential diagnosis considered inflammatory pseudotumor, splenic hamartoma, and low-grade lymphoma.

Due to concerns over the possibility of cancer and the presentation of thrombocytopenia, the patient was subjected to a splenectomy. Histological analysis of the removed spleen mass revealed it consisted of clusters of immature myeloid and erythroid cells, encased within a thin fibrous capsule, and featured a central area that was eosinophilic and displayed low cell density. Detailed examination using high-power microscopy identified the composition of the mass as including blood vessels, precursors to myeloid and erythroid cells, megakaryocytes, and extracellular hemosiderin deposits. The central area with low cell density was notable for containing numerous thick collagen bundles. No malignant cells were detected in the sample. The diagnosis established from the pathology was extramedullary hematopoiesis (EMH) in the spleen.

Following the procedure, the patient was monitored regularly. Over time, the symptoms of petechiae and ecchymoses diminished, and the platelet count normalized.

Discussion

Extramedullary hematopoiesis (EMH) serves as a backup process that kicks in when the bone marrow fails to produce an adequate amount of blood cells, leading to the formation of hematopoietic precursors in non-typical locations. EMH is most commonly observed as microscopic diffuse infiltration in various sites, particularly in the liver and spleen. The occurrence of EMH as a distinct mass within the spleen is unusual [1]. There are limited reports in literature regarding the magnetic resonance (MR) characteristics of EMH, and to our knowledge, the presentation of EMH with delayed central enhancement in MR imaging has not been previously documented. This unique MR imaging signature of EMH has been confirmed through pathological examination [1, 3].

The MR signal characteristics of EMH vary based on the stage of hematopoiesis and the activity level of the hematopoietic lesions. Lesions with active hematopoiesis, containing either immature or mature erythroid or myeloid cells, typically show intermediate signal intensity on T_1-weighted images and high signal intensity on T_2-weighted images [2]. Conversely, older inactive lesions may demonstrate low signal intensity on both T_1 and T_2-weighted images due to iron deposition, or high signal intensity in cases of predominant fatty infiltration. In the inactive phase of EMH, the lesions do not enhance with gadolinium [4, 5].

The atypical enhancement pattern observed in our case after gadolinium administration is attributed to the histological characteristics of the splenic mass, which included neovascular content likely responsible for the contrast enhancement in arterial phase images. The central zone's thick collagen bundles did not enhance in arterial phase images; however, contrast material gradually diffused into these hypocellular collagenous areas in delayed images. Due to the rarity and unique MR features of this condition, diagnosing EMH preoperatively was challenging. A definitive diagnosis of focal EMH in the spleen requires histopathological confirmation from a fine needle biopsy or surgical resection. Treatment may not always be necessary unless symptomatic. Options such as resection, radiotherapy, erythropoietin administration, and regular blood transfusions have been explored in managing EMH [5].

In summary, gadolinium-enhanced MRI plays a crucial role in the assessment of splenic masses, particularly when atypical imaging features are present. EMH, especially with peculiar delayed central enhancement, should be considered in the differential diagnosis of a splenic mass. The unique enhancement pattern observed in this case can be explained by the presence of a central hypocellular collagenous stroma in the EMH [6, 7].

Conflict of Interest
Not available

Financial Support
Not available

References

1. Bradley MJ, Metreweli C. Ultrasound appearance of extramedullary hematopoiesis in the liver and spleen. Br J Radiol. 1990;63:816-818.
2. Gabata T, Kadoya M, Mori A, Kobayashi S, Sanada J, Matsui O. MR imaging of focal splenic extramedullary hematopoiesis in polycythemia vera: Case report. Abdom Imaging. 2000;25:514-516.
3. Singer A, Maldjian P, Simmons MZ. Extramedullary hematopoiesis presenting as a focal splenic mass: case report. Abdom Imaging. 2004;29:710-712.
4. Katz D. Splenic extramedullary hematopoiesis: large focal lesion in a patient with thalassemia. AJR Am J Roentgenol. 1999;173:1715-1716.
5. Yokoyama T, Saigo K, Maeda N, Hamada Y, Yano Y, Mori T, *et al*. Tumor-like splenic extramedullary hematopoiesis in a patient with myelofibrosis. Intern Med. 2000;39:416-418.
6. Singounas EG, Sakas DE, Hadley DM, Chalevelakis G, Sfakianos G, Raptis S, *et al*. Paraplegia in apregnant thalassemic woman due to extramedullary hematopoiesis: successful management with transfusions. Surg Neurol. 1991;36:210-215.
7. Azua-Romeo J, Moreno E, Gomollon JP. Images incardiology: right atrial lithomyxoma with extramedullary hematopoiesis. Heart. 2002;88:10-12.

How to Cite This Article
Bouktib Y, Odongo S, Hajjami EA, Boutakioute B, Ouali M, Idrissi N. Focal extramedullary hematopoiesis in the spleen: An uncommon MRI presentation. International Journal of Radiology and Diagnostic Imaging. 2024;7(2):51-52.

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