Imaging studies in extramedullary hematopoiesis of the spleen

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LETTER TO THE EDITOR

Imaging studies in extramedullary hematopoiesis of the spleen

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Dear Editor,

A 63-year-old man underwent CT studies before surgery for an aortic aneurysm. Contrast medium-enhanced CT scan showed an increased spleen size with a solid, 10 cm large, round, well-defined highly enhanced lesion, suggesting high vascularity with nonhomogeneous density due to the presence of some pseudocystic areas (Fig. 1a) [1]. An increased spleen size had been discovered 13 years before without any symptoms and/or abnormality in complete blood counts (CBCs). Three years later, MRI studies confirmed a slightly increased lesion (12 cm) with a thin hypointense halo and a heterogeneous hyperintense signal intensity on T2-weighted images (i.e., well hydrated) (Fig. 1b, c). Diffusion-weighted images showed patterns compatible with a high cellularity (Fig. 1d). So, malignancy could not be excluded, and the patient underwent splenectomy.

Histological examination was consistent with extramedullary hematopoiesis, characterized by sinusoidal dilatation and hypolobated megakaryocytes often gathered in clusters (Fig. 1e, f). CBC and blood smears remained within the normal range. However, bone marrow examination showed an increased cellularity of 70 % with trilineage hyperplasia, with no increased fibrosis or blasts. The search of JAK2 V617F mutation was negative. Even if clinical and hematological findings did not fulfill the WHO 2008 diagnostic criteria for any of the myeloproliferative neoplasms [2], histological pattern of the bone marrow was suggestive for polycythemia vera, and hematopoiesis in the spleen had features resembling myelofibrosis. Two years after splenectomy, evidence of a myeloproliferative neoplasm did not occur.

Extramedullary hematopoiesis can be a compensatory response to hyperproliferating bone marrow cells, as is the case of hemoglobinopathies such as thalassemia syndromes, sickle-cell anemia, and other chronic hemolytic anemias, or can occur in patients with chronic myeloproliferative neoplasms, especially myelofibrosis. Foci of extramedullary hematopoiesis can be found in various organs, even if it predominantly affects the liver and spleen. The localizations of extramedullary hematopoiesis may be also in the thoracic paravertebral areas, lymph nodes, adrenal gland, retroperitoneal fat, and renal pelvis [3, 4].

To our knowledge, only a few cases of focal extramedullary hematopoiesis have been reported with imaging descriptions [5]. In fact, spleen tumors are usually detectable but not easily characterizable by imaging due to overlapping patterns between benign and malignant nodules. The greatest part of neoplasms of the spleen is benign: hemangiomas, inflammatory

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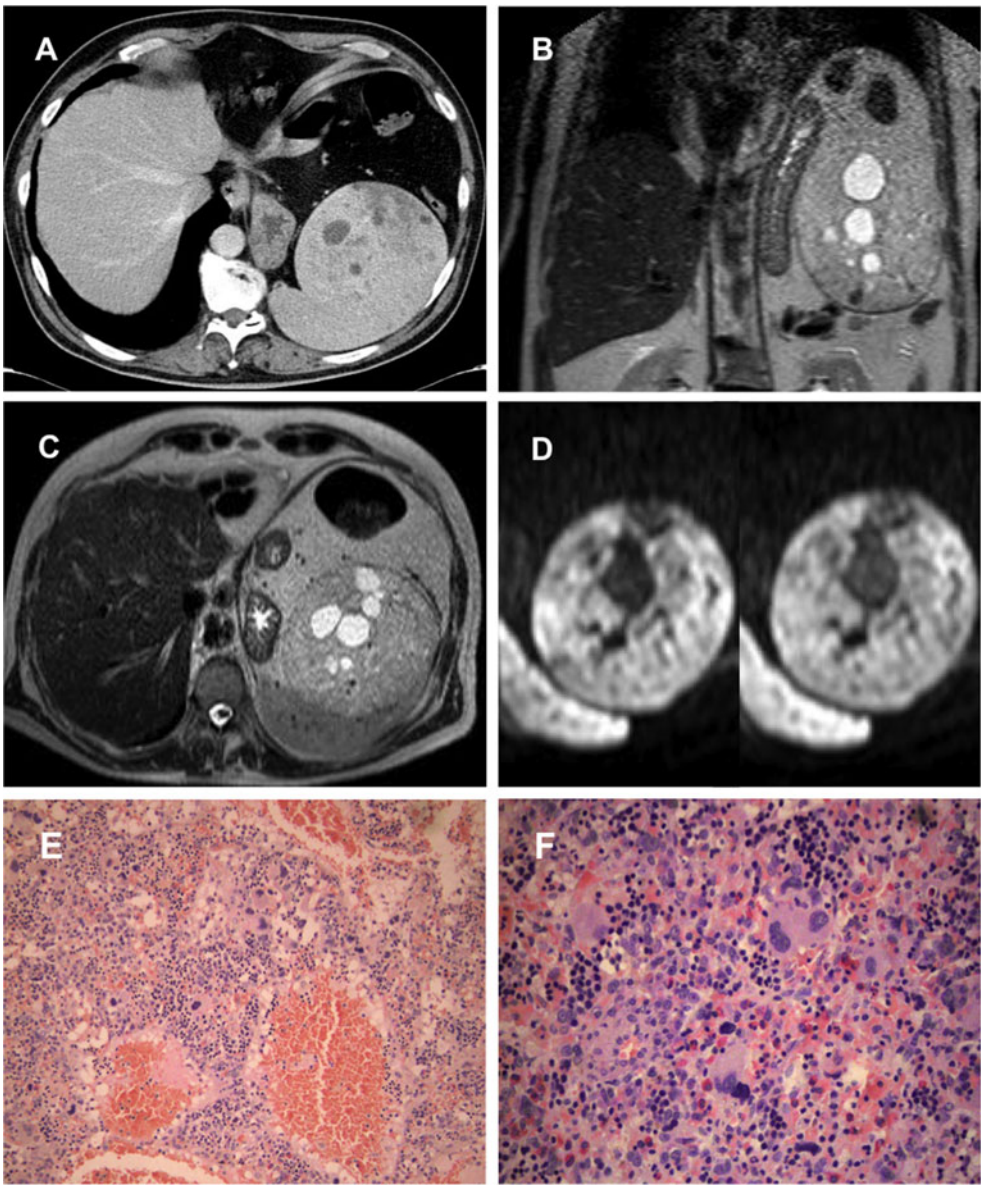

Fig. 1 **a** CT scan after administration of intravenous contrast medium shows a large focal lesion with well-defined margin and heterogeneous density due to internal colliquative areas. **b**, **c** MRI, T2-weighted coronal (**b**), and axial (**c**) scans confirm the lesion with heterogeneous signal intensity, slightly hyperintense on T2-weighted images, with a thin hypointense halo. **d** MRI and heavily diffusion-weighted axial scans show high signal intensity and then a pattern suggestive for crammed cells. **e**, **f** Histological examination of the nodule, showing extramedullary hematopoiesis with ectasic sinusoids and clustered hypolobated megakaryocytes

pseudotumors, and splenic cysts are the most frequent [6, 7], while lymphoma is the most common of malignancies, followed by other less frequent histologic types as hemangioendotheliomas, hemangiosarcomas, and metastases [8]. Our report emphasizes the role of radiologic features for the diagnosis of intrasplenic extramedullary hemopoiesis, bearing in mind that splenic biopsy is a very risky procedure.

Even if none of the above-described findings is neither sufficient nor necessary for definitive diagnosis, the association of well-defined lesion, highly and inhomogeneously enhancing, with pseudocystic areas, and medium-high signal intensity in MRI—T2 and diffusion weighted—not resembling hemangioma or pseudotumor, could suggest radiologists a diagnosis of intrasplenic extramedullary hemopoiesis [4, 5] and then clinicians to perform tests for hematological disorders including myeloproliferative neoplasms, such as bone marrow biopsy, JAK2V617F of MPL mutations in addition to CBC, before referring the patient for splenectomy which can bring some risks of complications.

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Conflict of interest The authors declare that they have no conflict of interest.

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