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Massive intrahepatic extramedullary hematopoiesis in myelofibrosis

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Abstract
We describe the sonographic (US) and computed tomographic appearances of a large, solitary tumor in the liver produced by extramedullary hematopoiesis in an 82-year-old patient with agnogenic myeloid metaplasia. Confirmation of this diagnosis was made by US-guided fine-needle aspiration biopsy.

Key words: Extramedullary hematopoiesis—Myelofibrosis—Liver, CT, US.

Extramedullary hematopoiesis (EMH) is a compensatory mechanism by which blood cells are produced outside the bone marrow when marrow production is unable to maintain the needs of the organism.

The most common sites of EMH are liver, spleen, and lymph nodes, but this condition has also been reported in other locations (kidney, adrenal gland, lung, pleura, skin, breast, ovary, thymus, gastrointestinal tract, presacral masses, central nervous system, and dura mater). The most common manifestations of EMH are hepatosplenomegaly and lymphadenopathy. Although macroscopic nodular foci of hematopoietic cells are occasionally found on autopsy examination of the liver, the radiologic demostration of the focal intrahepatic masses of EMH is unusual and these masses do not have specific imaging features.

We report on the radiologic findings (sonography, US; computed tomography, CT) of a focal intrahepatic lesion caused by EMH in a patient with idiopathic myelofibrois. The diagnosis was established by US-guided fine-needle aspiration biopsy (FNA).

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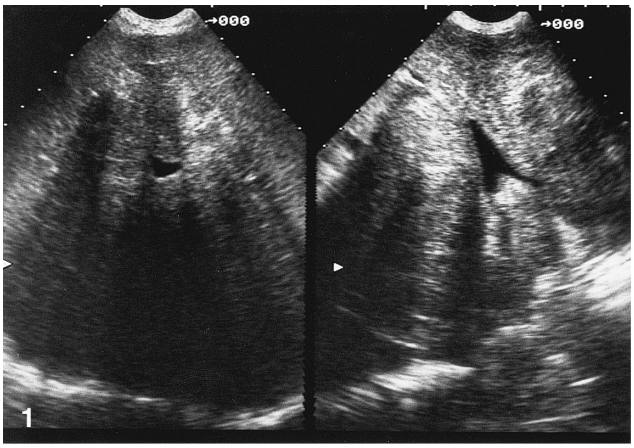
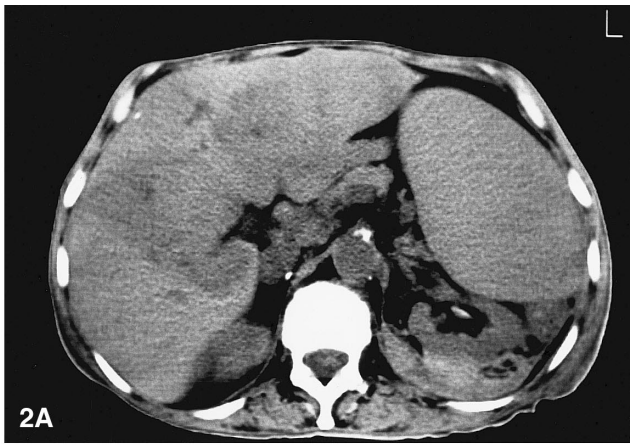
Case report

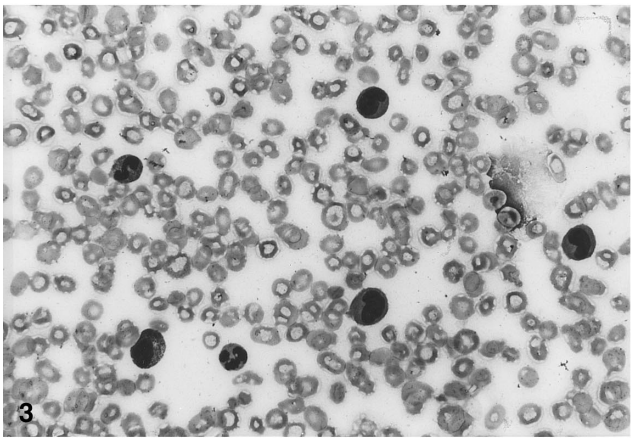
An 82-year-old man was admitted to be operated upon for bilateral inguinal hernias. Anorexia and weight loss had been present. Physical examination showed an enlarged liver and spleen. Abdominal US showed hepatosplenomegaly and a large intrahepatic mass (Fig. 1). This was a solid hyperechoic lesion that attenuated the sound and had a periportal location, affecting both lobes. The spleen was normal in echotexture, with a craniocaudal measurement of 19 cm. Noncontrast CT showed a well-defined, lobulated, hypodense, intrahepatic lesion (15 cm in mean diameter), without invasion of vessels and without signs of cirrhosis (Fig. 2A,B). The lesion had no enhancement when intravenous contrast agent was given. A retroperitoneal hematoma involving the left perirenal, posterior pararenal, and iliopsoas spaces was also found and was attributed to the previous bone marrow biopsy. The chest radiograph was normal. Laboratory studies showed a white blood cell count of 32,000/mm^3, hemoglobin of 9.8 g/dL, hematocrit of 31%, mean corpuscular volume of 96; platelet count of 72,000/mm^3, total bilirubin of 1.85 mg/dL and indirect bilirubin of 1.2 mg/dL, alkaline phosphatase of 656 U/L, lactate dehydrogenase of 1314 U/L, and gamma glutamyl transpeptidase of 156 U/L. Bone marrow biopsy was consistent with myelofibrosis. Two aspirations were obtained with a 22-gauge needle, each time documenting correct positioning of the needle within the hepatic mass. The microscopic examination of FNA material and the patient's medical history led to the definitive diagnosis of EMH (Fig. 3).

Discussion

EMH is a compensatory mechanism by which blood cells are produced outside the bone marrow when marrow production is insufficient to meet the body's demands [1]. Erythropoiesis occurs normally in an extramedullary fashion during gestation. In postnatal life, EMH can occur (a) in congenital hemolytic anemias such as thalassemia and hereditary spherocytosis, (b) in severe hemolytic and ineffective erythroietic states such as hypochromic anemia, pernicious anemia, erythroblastosis fetalis, and kala-azar, (c) in conditions where control of stem cell differentiation is lost (i.e., myelofibrosis, myelosclerosis,







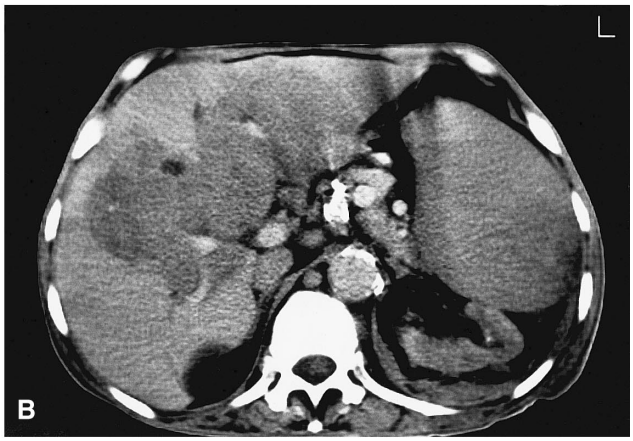


Fig. 1. Hepatic sonogram shows a poorly defined hyperechoic mass in both lobes, which attenuated the sound in some areas.

Fig. 2. Nonenhanced (**A**) and contrast-enhanced (**B**) CT demonstrates a massive, well-defined, lobulated, hypodense intrahepatic mass with periportal location. The lesion displaced and encircled the main branches of the portal vein without signs of vascular invasion. The lesion demonstrated no enhancement when intravenous contrast agent was given. Left posterior pararenal hematoma was seen and was attributed to the previous bone marrow biopsy.

Fig. 3. Immature myeloid cells in US-guided fine-needle aspiration cytology. Diff-Quick stain, 400×.

polycythemia vera), and (d) in nonmyeloid neoplastic diseases including Hodgkin lymphoma, leukemia, and carcinomatosis secondary to a myelophthistic effect on the bone marrow [2].

The most common sites of EMH are liver, spleen, and lymph nodes [1–12]. The paraspinal region is a common location for these hematopoietic deposits in cases of hemolytic anemias [13]. It has been described as rare locations in kidney [6, 14], adrenal gland [5], lung, pleura [4, 15], skin, breast, ovary, thymus, gastrointestinal tract, presacral masses [2], mesenteric and omental masses [8], central nervous system, and dura mater [7], although EMH can develop in any organ of mesenchymal origin [9].

Our patient had a myelofibrosis bone marrow proven by biopsy. The radiographic hallmarks of myelofibrosis are diffuse bone sclerosis (40–50% of cases) and EMH with hepatosplenomegaly. Splenomegaly is the most common manifestation of EMH (80% at initial diagnosis and 97% at follow-up) [10], although some degree of myeloid metaplasia is always present in the liver if the spleen is involved [16]. Homogeneous hepatomegaly is commonly found in patients with myelofibrosis (25% at initial diagnosis and 39% at follow-up) [10], but patterns of focal myeloid metaplasia of the liver have been described in a few case reports [1, 10, 12, 16].

The focal hepatic disease can manifest as solitary or multiple lesions [1, 12]. The US features of EMH are variable. Siniluoto et al. [10] described two distinctly different sonographic appearances of focal intrahepatic myeloid metaplasia: (a) a well-defined homogeneously hypoechoic lesion and (b) a well-defined homogeneously hyperechoic mass encircling the portal vein and its main branches. These lesions can be hypoechoic [1, 10, 12, 16] or hyperechoic [10] masses, frequently inhomogeneous, of variable size (2.5–15 cm), and their borders can be well

defined [1, 10] or poorly defined [12]. The variation in US patterns is probably in proportion to the amount of fat and fibrosis in the mass, so that more fat and fibrosis produces an increase in the echogenicity of the lesions [10]. On CT, lesions of focal intrahepatic EMH are hypodense, frequently heterogeneous, and can show patchy or no enhancement with intravenous contrast agents [12]. In our case, sonography showed hepatosplenomegaly with a large solid hyperechoic mass, with a periportal distribution. The lesion was hypodense on noncontrast CT, with a well-defined edge, and did not show enhancement with intravenous contrast. We thought that these characteristics could be related to a high grade of intralesional fibrosis.

To our knowledge, only one case of EMH intrahepatic has been imaged by magnetic resonance imaging [12]. The lesion showed mild hyperintensity relative to the remaining liver parenchyma on the T2-weighted sequences (it was less than that usually associated with metastasis and markedly less than the signal intensity level found in hemangiomas) and presented heterogeneous enhancement during the bolus administration of Gd-DTPA. No persistent enhancement was observed on subsequent T1 images.

Confirmation of the diagnosis of EMH may be made with nuclear medicine scanning using bone marrow agents, which shows an uptake in the colloid in foci of EMH [1], or with fine needle biopsy [10, 12, 17]. The definitive diagnosis in our case was performed by US-guided FNA biopsy. Cytological examination produced the diagnosis of intrahepatic EMH.

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