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Focal intrahepatic extramedullary hematopoiesis: color Doppler US and CT findings

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Abstract
Focal intrahepatic extramedullary hematopoiesis is an extremely unusual condition that can mimic any other solid mass of the liver. Only seven cases have been reported in the literature. Color Doppler ultrasonographic findings of this rare entity have not been described. We report a case of focal mass in a myeloid metaplastic patient diagnosed as focal extramedullary hematopoiesis. Color Doppler findings of this rare condition are described and compared with computed tomography features.

Key words: Liver—Extramedullary hematopoiesis—Ultrasonography—Computed tomography.

Extramedullary hematopoiesis is a compensatory phenomenon that occurs when erythrocyte production is diminished or destruction is accelerated. It can be seen in a variety of hematologic disorders including primary myelofibrosis.

Extramedullary hematopoiesis is usually microscopic and commonly involves the liver, spleen, and lymph nodes. Rarely, the involvement may be macroscopic, as in our case, and should be included in the differential diagnosis of the masslike lesions.

Case report

A 60-year-old man presented with a history of fatigue, anorexia, and weight loss. Physical examination showed a pale cachectic man. Both the spleen and liver were markedly enlarged on palpation. Laboratory results included a hemoglobin level of 9.3 g/dL, platelet count of 315 $\times$ $10^3/mm^3$, white blood cell count of 6.5 $\times$ $10^3/mm^3$, LDH of 577 U/L (normal = 91–232 U/L), SGOT of 32 U/L (normal = 0–35 U/L), SGPT of 46 U/L (normal = 5–43 U/L), and total bilirubin of 0.8 mg/dL (normal = 0.1–1.2 mg/dL). Erythrocyte sedimentation rate was raised at 30 mm/h. Bone marrow biopsy was consistent with agnogenic myeloid metaplasia (primary myelofibrosis).

Abdominal ultrasound showed diffuse hepatosplenomegaly and a large, fairly well-defined, inhomogeneous solid mass 6.5 cm in diameter in the left lobe of the liver (Fig. 1A). Color Doppler examination showed a left hepatic vein traversing the central portion of the lesion and a left portal vein branch along its posterior edge (Fig. 1B). Neither vascular distortion nor neovascularization of any type was detected. For further evaluation, dynamic computed tomography (CT) was performed. On noncontrast CT, the lesion was seen as a fairly well-marginated lobulated low-attenuation mass of 35–50 Hounsfield units. No enhancement was detected during dynamic scanning, which also displayed the left hepatic vein crossing the central portion of the mass (Fig. 1C). Although sonographic appearance, color Doppler features and CT findings were not pathognomonic the patient's history of the myelofibrosis allowed us to suggest a preliminary diagnosis of extramedullary hematopoiesis, and ultrasound-guided Trucut biopsy was performed. Histology showed immature and mature cells of the myeloid and erythroid series, without any atypical or blastic cells (Fig. 1D), thus confirming the diagnosis of extramedullary hematopoiesis.

Discussion

Agnogenic myeloid metaplasia or idiopathic myelofibrosis is primarily a disorder of middle-aged or older adults [1]. The disease begins insidiously and then follows a chronic course. It is characterized by anemia and bone marrow fibrosis, often with accompanying osteosclerosis and extramedullary hematopoiesis [1, 2]. Extramedullary hematopoiesis is a compensatory mechanism and is uncommon in adults [2–4]. Although etiology and pathogenesis of extramedullary hematopoiesis are unknown, it is believed that the abnormal stimulus can cause proliferation of multipotential stem cells already present in mesenchymal tissues throughout the body [2, 3]. Any

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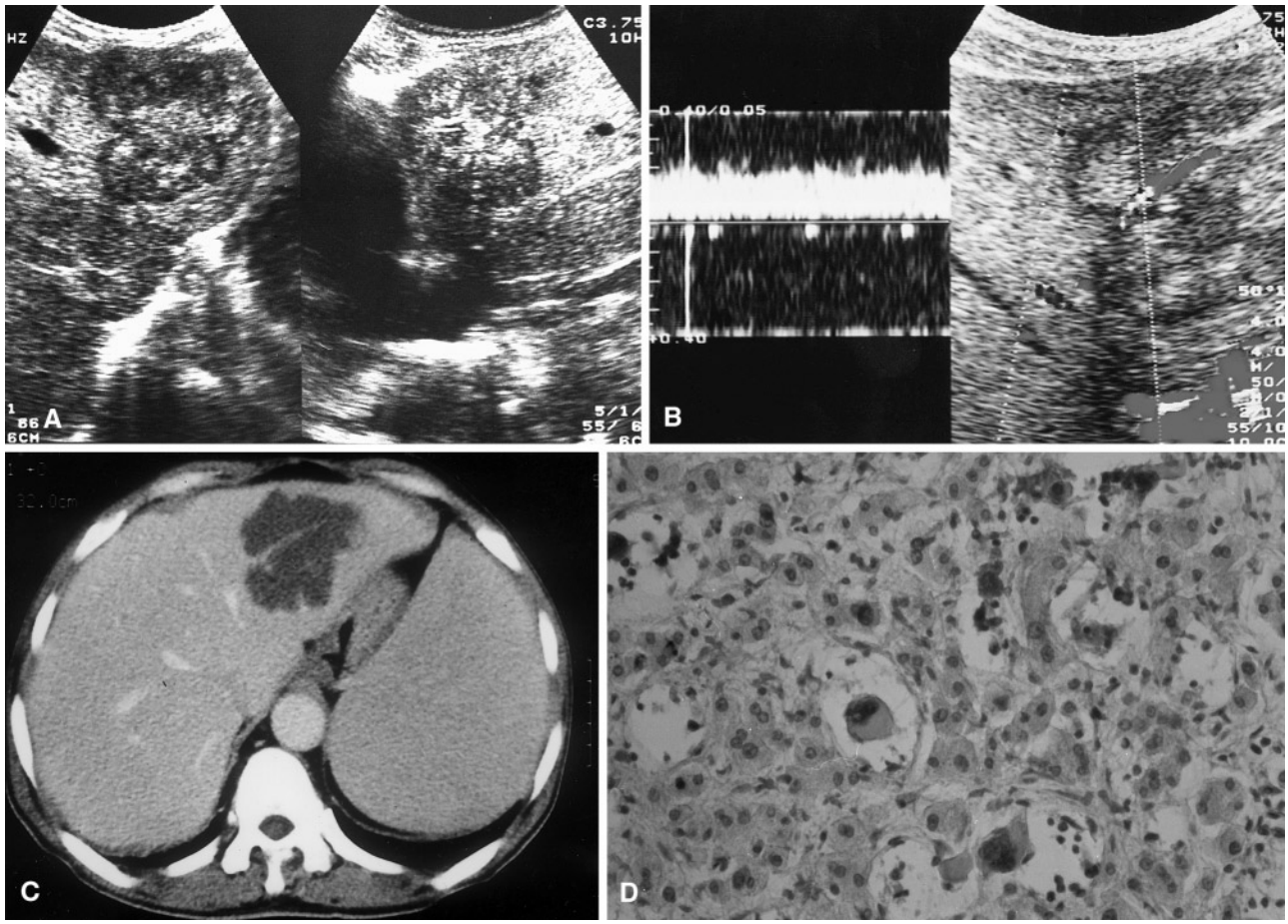

Fig. 1. A Oblique sagittal ultrasonography through the left lobe of the liver shows a hypoechoic heterogeneous solid mass. **B** Color Doppler ultrasonography displays the left hepatic vein crossing the central part of the mass. **C** Dynamic enhanced CT section demonstrates the well-marginated hypodense lesion traversed by a vascular structure. **D** Microscopic specimen shows megakaryocytes and other hematopoietic precursor cells inside enlarged sinusoids among hepatocyte cords. Hematoxylin and eosin. Original magnification, ×200.

organ of mesenchymal origin can theoretically serve as a site of extramedullary hematopoiesis in agnogenic myeloid metaplasia, but spleen and liver are most commonly involved [2, 5, 6]. In a study of 70 cases of myelofibrosis, extramedullary hematopoiesis was almost always seen in the spleen microscopically [7]. Splenomegaly is usually the leading clinical finding, but some degree of myeloid metaplasia is always present in the liver if the spleen is involved [1]. Other less frequent sites of involvement include lymph nodes, adrenal gland, cranial vault, retroperitoneum, breast, spinal canal, renal pelvis, thymus, heart, lung, pleura, skin, dura mater, and epididymis [1, 5, 7, 8]. Only seven cases of focal tumorlike myeloid metaplasia in the liver have been reported, but none included color Doppler features of the lesions.

Ultrasonographically, focal myeloid metaplasia of the liver may have variable patterns, which have been documented in a few cases in the literature. Instead of a heterogeneous, fairly well-defined hypoechoic appearance, as in our case, homogeneous echogenic or hypoechoic heterogeneous echogenic masslike lesions have been described [1, 4–6]. There is no definite explanation based on cytological grounds regarding this variation in sonographic presentation. Whereas many investigators have correlated the echogenicity of the lesions with their fat content, Siniluoto et al. suggested that fibrosis may also account for the increased echogenicity [5]. In our case, paucity of fibrosis in histopathological examination and the hypoechoic inhomogeneous appearance support this idea. Variation in ultrasonographic patterns can cause diagnostic difficulties. In the differential diagnosis, both infiltrative lesions such as leukemia and lymphoma and noninfiltrative processes such as primary and metastatic tumors, hemangioma, focal nodular hyperplasia, and abscess should be taken into consideration. Color Doppler sonography plays an important role in the differentiation of benign lesions from malignant tumors by showing abnormal vascularization, vascular infiltration, and thrombosis in the malignant tumor. In the present case, the lesion encircled a vascular structure, just like another

case defined by Siniluoto et al. [5];, however, vessel distortion and thrombosis were not present. This appearance favors mostly infiltrative lesions and benign processes. Because determination of enhancement pattern and fat content on CT may be diagnostic for certain lesions, dynamic enhanced CT was performed to characterize the lesion further. However, CT offered no additional information over ultrasonography. As with the sonographic findings, CT appearance of the lesion was not specific. In the literature, CT appearances have been described only in two cases: one had a mixed attenuation pattern with patchy contrast enhancement and the other, in which contrast medium was not used because of renal insufficiency, had a hypodense appearance [1, 6].

Because current experience is limited to describing specific radiological features, this extremely rare condition should be considered as part of the differential diagnostic spectrum of hepatic masses, especially in patients with a history of myelofibrosis. Final diagnosis necessitates histopathological examination of the biopsy material, as was done our case.

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