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Large Splenic Mass of Extramedullary Hematopoiesis

The imaging findings of focal splenic, presacral and paraosseous extramedullary hematopoiesis in a 35-year-old man with ß-thalassemia major are described with particular reference to ultrasonography and CT scan. A review of the radiological appearances of this rare condition is presented.

KEYWORDS: Computed Tomography, splenic mass, extramedullary hematopoiesis.

Introduction

Hematopoiesis is the formation and maturation of blood elements. Hematopoiesis normally occurs in the marrow of long bones, the ribs, vertebrae and flat bones of the adult, in contrast to the fetus, in which the principle sites of hematopoiesis are the yolk sac, spleen and the liver. When the primary sites of hematopoiesis in the adult fail, as in myelofibrosis and hemoglobinopathies (especially thalassemia and sickle cell disease), various extramedullary sites take on the role of blood formation.

Extramedullary hematopoiesis (EMH) favors certain sites such as the liver, spleen and the paraspinal regions of the thorax. However, in addition to these common sites of EMH, the process can involve virtually any organ or tissue.[1] Reported sites include abdominal viscera, lymph nodes, adrenal glands, kidneys and intracranial structures.[2-6]

Although foci of EMH may be commonly identified microscopically in the liver and spleen, in such patients, identification of focal lesions of EMH in the liver and spleen on cross sectional imaging is rare.[7-10] The classic imaging finding is that of hepatosplenomegaly. Involvement of these organs is diffuse, but mass-like foci of hematopoiesis is a rare finding.

The differential diagnoses include lymphoma, hematoma, metastasis, hemangioma, and sarcoidosis.[11] But the main differential diagnoses for the isolated splenic lesions include non-Hodgkin lymphoma or EMH. Because of the potential hazards of the splenic biopsy, it is better to establish the diagnosis non-invasively by patient follow up.[11]

In most reported cases of splenic EMH, ultrasonography has demonstrated a well-circumscribed hyperechoic mass, whereas CT scan has displayed a heterogeneous mass, better visualized after administration of intravenous contrast medium.[12]

Case Report

We describe a 35-year-old Iranian man with β-thalassemia major, who required chronic blood transfusion and deferoxamine mesylate (Desferal) administration.

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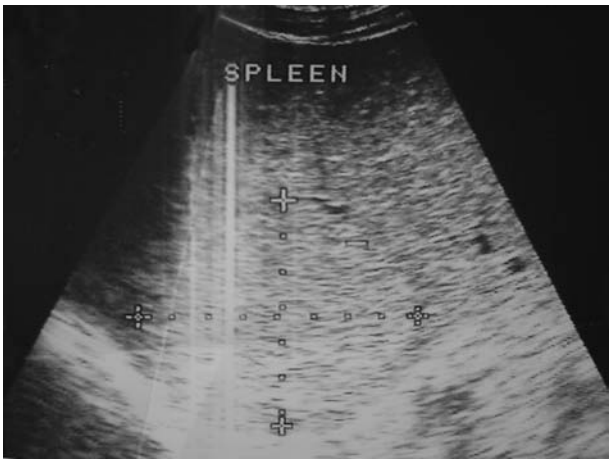

Fig 1. Ultrasonography at splenic level reveal an uniformly hyperechoic, well-marginated oval lesion in spleen.

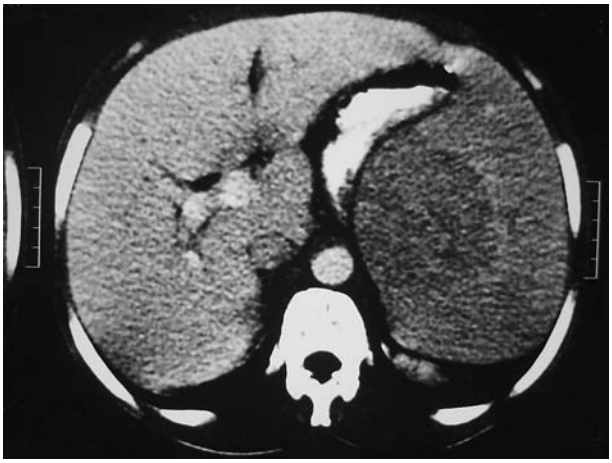

Fig 2. Axial enhanced CT scan at upper abdominal levels reveals huge splenomegaly and an uniformly low-attenuation (compared with spleen parenchyma), well marginated mass that undifferentiated from normal spleen in late phase. No associated paraaortic or pelvic lymphadenopathy.

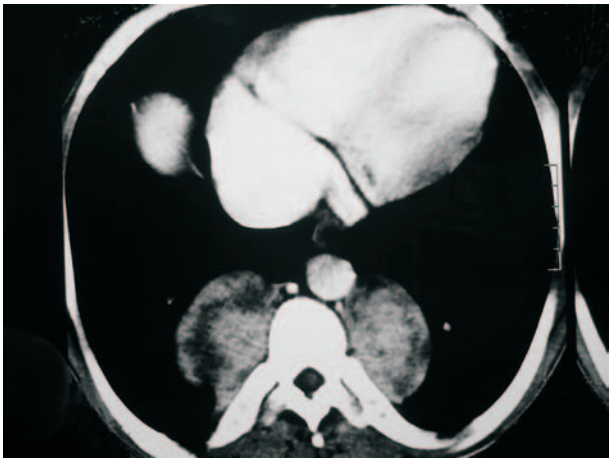

Fig 3. Axial CT scan through chest shows non uniformly enhancing hemopoetic masses with no bony erosion.

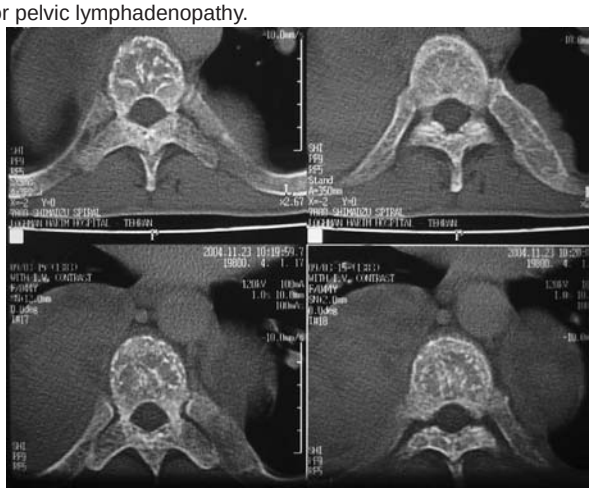

Fig 4. Axial contrast CT scan through chest shows non uniformly enhancing paraspinal masses with generalized bony expansion and coarse trabeculation.

In ultrasonic evaluation of the upper abdomen, a huge splenomegaly with a well-defined hyperechoic mass (80×64 mm) was seen in the spleen (Figure 1) that appeared as a hypodense area on the unenhanced and early-enhanced CT scans, located at the splenic hilum (Figure 2) which on delayed scans became insolence with normal splenic parenchyma. In addition, bilateral heterogeneous paravertebral masses due to paraosseous EMH were seen in the lower thoracic spine (Figure 3). Generalized coarse trabeculation of skeleton and expansion of the ribs adjacent to the paravertebral masses were noted accompanied by increased density of the liver due to blood transfusions (Figure 4). No lymphadenopathy was detected. Expansion and destruction of sacrum by a presacral pelvic mass due to EMH was visualized as well (Figure 5).

Discussion

Scattered reports have noted tiny focal splenic lesions simulating an infectious process, which proved to be EMH in such patients. Rarely, large lesions of EMH in the liver have been noted and reported in the literature .[13]

In a review of literature, only five large foci of proven EMH in spleen have been reported: 1) A 58-year-old man with history of small cell carcinoma of the lung who underwent abdominal sonography and the large splenic mass was proven to be EMH associated with his concurrent β-thalassemia intermedia, using fine needle aspiration of the splenic lesion.[13] 2) Two splenic masses in an enlarged spleen in a 65-year-old woman with anemia.[5] 3) A 57-year-old man with a 2-year history of high grade prostatic adeno-

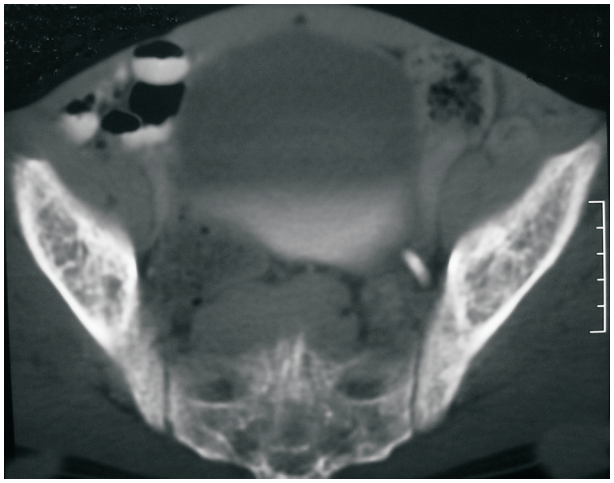

Fig 5. Axial enhanced CTscan through pelvis shows well marginated presacral soft tissue mass with bone expansion and coarse trabeculation.

carcinoma displayed a tumor-like splenic mass that was first assumed to be a metastatic lesion; nevertheless, it was an EMH focus.[6] 4) A 33-year-old man with β-thalassemia major and chronic splenomegaly and two splenic masses.[13] 5) A 65-year-old woman with anemia and two low attenuation lesions within an enlarged spleen.[11]

Although splenic lesions in patients with longstanding thalassemia are extremely rare, the differential diagnosis should include EMH, particularly if there is no other explanation for such lesions after the initial clinical and laboratory examinations.

A Tc-99m sulfur colloid liver-spleen radionuclide scan provides a convenient and non-invasive method to determine whether a solid focal splenic lesion is secondary to EMH in a patient with unexplained anemia, albeit it was not performed in our case.[11]

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