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Periportal Extramedullary Hematopoiesis[1]

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In a bone marrow failure patient, a soft tissue mass lesion in the periportal area is a rare presentation. We present the sonographic and dynamic CT findings of a histologically confirmed case of hepatic periportal extramedullary hematopoiesis.

Index words : Hematopoiesis, Extramedullary
Liver
Tomography, X-Ray Computed

Extramedullary hematopoiesis (EMH) is a type of ectopic hematopoiesis that is a common feature of chronic bone marrow failure. However, EMH is rarely encountered in general radiological practice (1). A mass-like EMH lesion in the liver is an atypical presentation. Furthermore, hepatic periportal EMH is less common than other types of focal hepatic EMH (2).

We present the sonographic and CT findings of a histologically confirmed case of hepatic periportal extramedullary hematopoiesis.

Case Report

A 72-year-old female presented with a history of abdominal distension over the last few months accompanied with the presence of a palpable mass. During the few weeks before admission, the patient had suffered from diarrhea. The patient had no history of any previous surgery or medical illness. A physical examination revealed a large spleen that was subject to palpation in the left abdomen. An initial peripheral blood smear was suspicious for microcytic hypochromic anemia with marked leukoerythroblastosis.

A sonography and dynamic CT were performed as part of a work-up. The sonography revealed hepatosplenomegaly and hypoechoic soft tissue lesions in the periportal area and both renal pelves (Figs. 1A, B). Dynamic CT arterial phase images revealed hypoattenuating soft tissue mass lesions in the periportal area and both renal pelves (Figs. 1C, D). Portal venous phase images showed the presence of minimally enhanced, well demarcated, hypoattenuating soft tissue lesions in the periportal area and both renal pelves (Figs. 1E, F). Also noted were multiple mesenteric enlarged lymph nodes and hepatosplenomegaly. The initial radiological differential diagnosis was lymphoma or leukemia.

Initially, bone marrow aspiration and a biopsy were performed for a pathological work-up. The bone marrow aspiration showed marked diluted samples, while the bone marrow biopsy demonstrated the presence of severe fibrosis, bony formation, and fresh hemorrhage. Based on the possibility of lymphomatous or leukemic involvement, a sonography-guided liver biopsy was performed on the periportal soft tissue lesions. Two lesion cores were taken and were found to demonstrate sinusoidal dilation in areas of predominant megakaryocytes and clumped erythroid precursor presence, as well as in an area where histiocytoid cells were present. This find-

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ing is consistent with an EMH (Fig. 1G). Immunohistochemical staining was performed with anti-LCA (leukocyte common antigen), anti-pan T, anti-pan B, anti-myeloperoxidase, and anti-CD68 antibodies. The megakaryocytes and erythroid precursor cells all showed negative immunostaining with the antibodies (data not shown).

The patient was diagnosed with myelofibrosis with diffuse fibrosis and osteosclerosis based on the bone marrow biopsy, and EMH in the hepatic periportal area based on the sonography-guided liver biopsy and severe anemia. Although we did not perform a histological examination for the soft tissue lesions in both renal pelves, EMH was considered as a possible diagnosis because of the imaging findings and clinical information in comparison to previous reports (1, 3).

Discussion

EMH is the proliferation of hematopoietic cells outside of the bone marrow, which occurs in compensation of insufficient blood cell production. This insufficient production results from either marrow failure (a myeloproliferative disorder or an infiltrative disease) or ineffective circulating of mature blood elements (hemoglobinopathy) (4-6). When the primary sites of hematopoiesis fail in an adult, various extramedullary sites take on the role of blood formation. EMH favors certain sites such as the liver, spleen, and paraspinal regions of the thorax (4). In addition to these common sites for EMH, the process can occur in a wide variety of organs such as the pleura, lung, gastrointestinal tract,

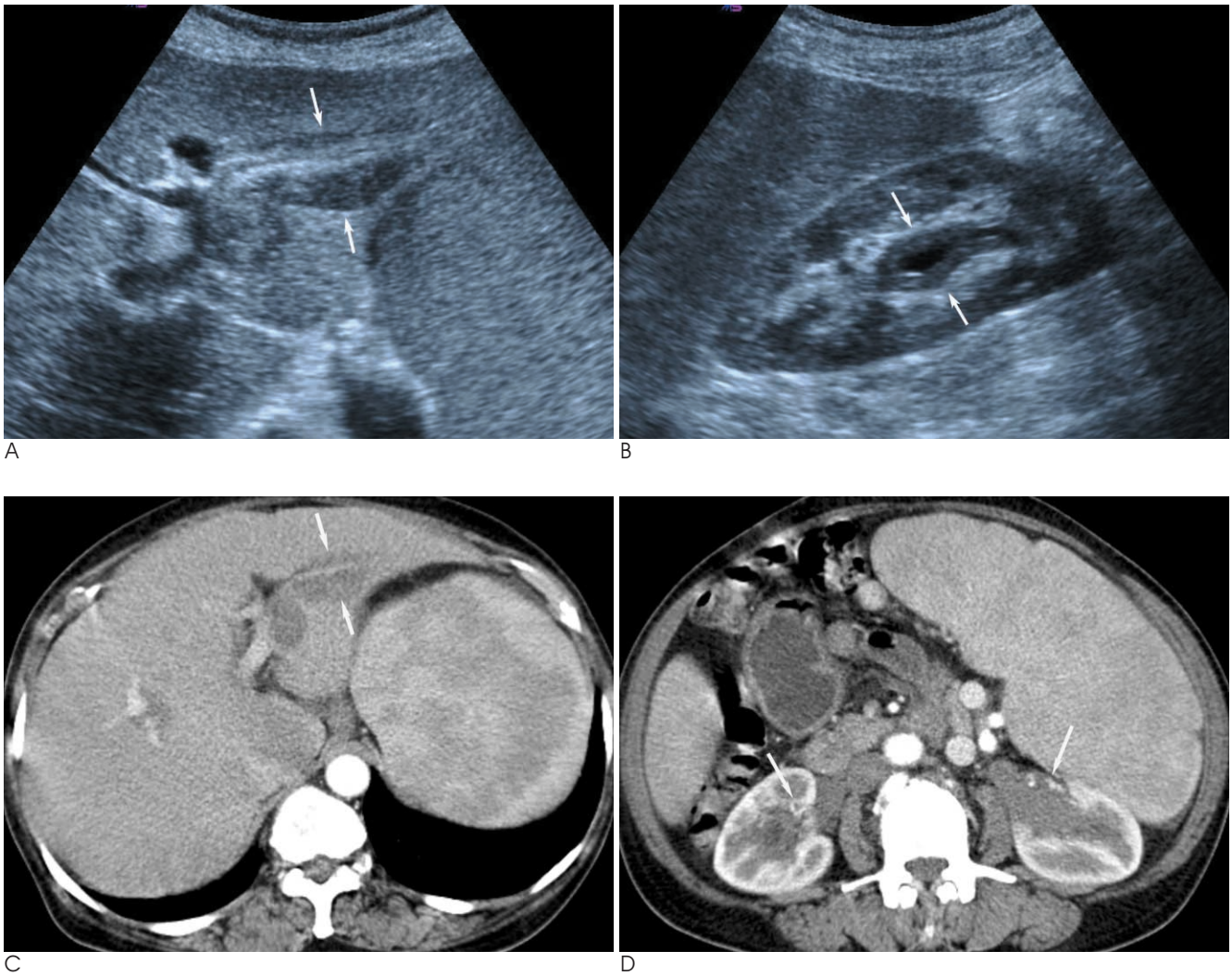
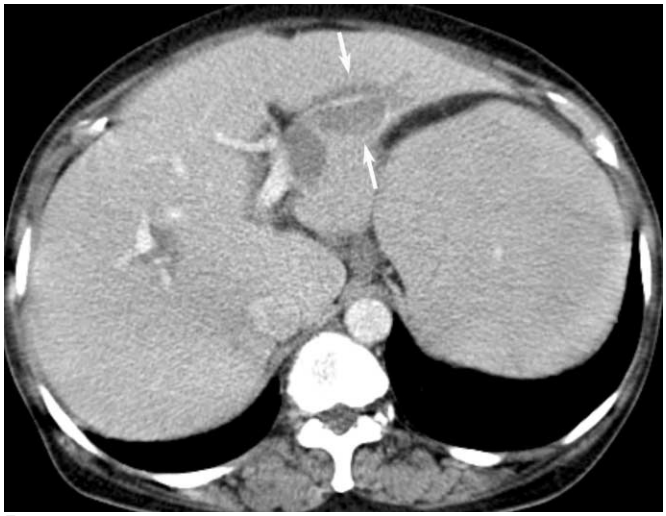


Fig. 1. A 72-year-old female patient presented with a palpable spleen and vague abdominal discomfort.
A, B. Ultrasonography shows the presence of hypoechoic lesions (arrows) in the periportal area and both renal pelves.
C, D. Arterial phase images show hypoattenuating soft tissue mass lesions (arrows) in the periportal area and both renal pelves.



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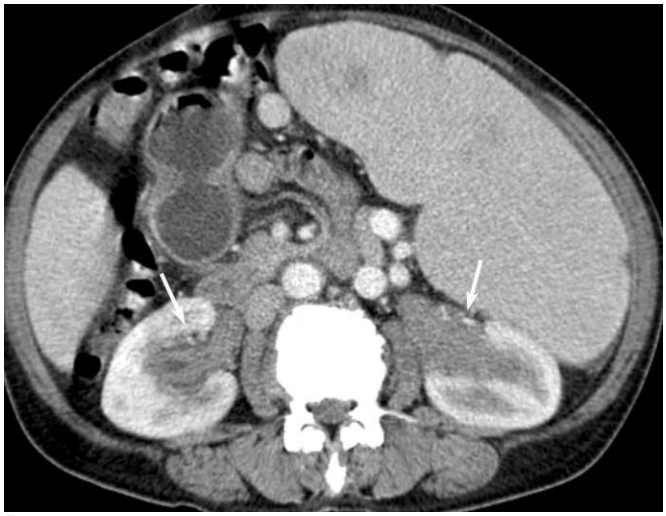

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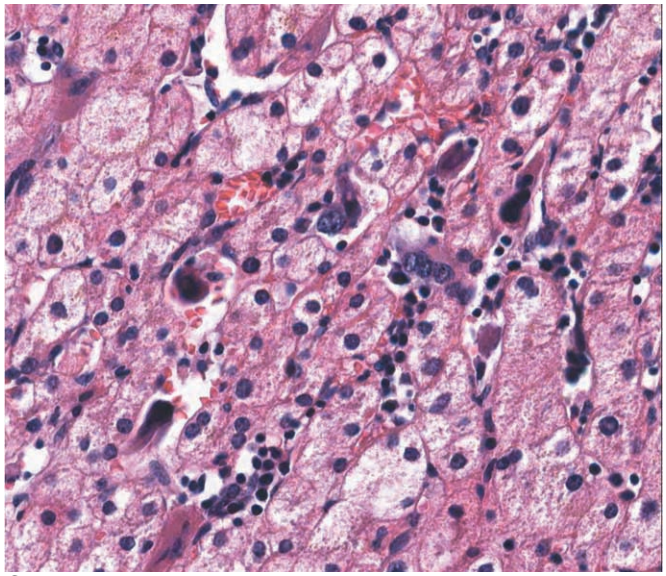

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Fig. 1. E, F. Portal venous phase images demonstrate minimally enhanced, hypoattenuating soft tissue mass lesions (arrows) in the periportal area and both renal pelves as well as marked splenomegaly and enlarged mesenteric lymph nodes.
G. H & E staining shows sinusoidal dilation with predominantly megakaryocytes and clumped erythroid precursors in addition to histiocytoid cells.

breast, skin, brain, kidneys, and adrenal glands (1).

Hepatic involvement of EMH is usually a diffuse, but a mass-like focal hepatic EMH can be confused with a neoplastic process (4). Wong et al. (2) summarized 10 cases of focal hepatic EMH that included nine previously reported cases over the last 20 years and one novel case. Focal hepatic EMH lesions were singular in four cases and were multiple in six cases. All of the cases were found to be located within the liver parenchyma, with the exception of one case with nodular masses around the portal vein. A periportal soft tissue-like lesion is a very rare manifestation of EMH (2, 7).

Imaging findings of focal hepatic EMH are variable. The variability of the imaging appearance of focal hepatic EMH has been hypothesized to be dependent on the relative amounts of normal marrow constituent-fat or hematopoietic cells and fibrosis within an extramedullary hematopoiesis deposit (6). Based on previous reports, focal hepatic EMH depicted on unenhanced CT scans have been described as hypodense lesions. Enhanced CT scans have shown heterogeneous and mild enhancement of the focal lesions (2, 7).

To the best of our knowledge, few reports have described hepatic periportal EMH on dynamic CT. In our case, dynamic CT portal venous phase images showed the presence of well-demarcated minimally enhanced periportal hypoattenuating lesions. During the portal venous phase of dynamic CT, hypovascular tumors remain unenhanced or minimally enhanced and are depicted as hypoattenuating areas with high contrast (8). Thus, hypoattenuating periportal EMH, as seen on portal venous phase images, may be a reflection of the poor

vascularity.

The differential diagnosis of periportal hypoattenuating lesions on CT imaging includes leukemic cell invasion in acute myelogeneous leukemia, hepatic involvement of Schistosomiasis mansoni, or hepatic lymphangitis carcionoma in an undifferentiated carcionoma of the gall bladder. Lymphatic dilatation and perivascular lymph edema in patients that have had blunt trauma, operative injury, malignant abdominal lympadenopathy or congestive hepatomegaly also present periportal hypoattenuating areas on CT images (7).

A biopsy for the soft tissue mass lesion in the renal pelvis was not performed in our case. However, the presence of soft tissue mass lesions in both renal pelves was consistent with previously described reports of EMH (1, 3). Renal involvement can be seen as parenchymal, intrapelvic or perirenal (1). Intrapelvic involvement presents with hypovascular masses in the renal hilum, which encase the pelvocalyceal system. The types of intrapelvic renal involvement are often bilateral, but can also be unilateral. In a bone marrow failure patient, CT scans indicating bilateral homogeneous hypoattenuating infiltration in the renal pelvis should raise the suspicion of EMH as a possible diagnosis (1). The differential diagnoses of a mass encasing the renal pelvis includes transitional cell carcinoma, lymphoma, lipomatosis, hypernephroma, and renal cell carcinoma (3, 9).

Recognition that the imaging findings may be compatible with extramedullary hematopoiesis is important, as a biopsy will exclude the possibility of a neoplasm and subsequent management. When hepatic periportal hypoattenuating lesions on dynamic CT portal venous phase images are detected in a bone marrow failure patient, extramedullary hematopoiesis should be included in the differential diagnoses.

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하 동 호

골수 외 조혈 소견 중 간 문맥 주위의 국소적 연부 조직 소견은 아주 드물게 나타난다. 이에 저자들은 조직학적으로 확진된 간 문맥 주위에 발생한 골수 외 조혈을 경험하였기에 초음파와 컴퓨터 단층촬영 소견을 보고하고자 한다.