J Korean Med Sci 2000; 15: 460-2
ISSN 1011-8934

CT Findings of Extramedullary Hematopoiesis in the Thorax, Liver and Kidneys in a Patient with Idiopathic Myelofibrosis

Extramedullary hematopoiesis occurring in multiple organs such as thorax, liver and both kidneys is an unusual condition. We report the CT findings of this condition with review of literature. The lesions consisted of intrathoracic paravertebral masses, focal intrahepatic mass and masses of both pelvocalyceal systems on CT scans.

Key Words: *Thoracic Neoplasms; Liver Neoplasms, CT; Kidney Neoplasms, CT*

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Received: 8 November 1999
Accepted: 30 December 1999

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INTRODUCTION

Extramedullary hematopoiesis (EMH) is a rare but well-recognized consequence of ineffectual formation of red blood cells (1). It is thought to be a physiologic compensatory mechanism for disturbed medullary hematopoiesis often accompanying congenital hemoglobinopathies or acquired marrow replacement disorder, such as leukemia, lymphoma, carcinoma and myelofibrosis. Most often found in the liver and spleen, EMH has also been reported in kidney, adrenal gland, lymph node, lung, pleura, skin, breasts, dura mater, ovary, thymus, gastrointestinal tract and central nervous system (2, 3). We report a case of EMH involving the intrathoracic mediastinum, liver and both kidneys on CT scan in a patient with idiopathic myelofibrosis.

CASE REPORT

A 59-year-old woman with a 16-year history of idiopathic myelofibrosis was admitted with recent onset of general malaise and fever. Hepatomegaly was noted on physical examination. Laboratory values included a hemoglobin level of 8.3 g/dL, hematocrit of 24.5%, platelet count of $179 \times 10^3/\mu L$ and leukocyte count of $39,900/\mu L$.

Abdominal ultrasonography showed diffuse hepatomegaly and a large, ill-defined, inhomogeneous hypoechoic mass in the right lobe of the liver and echogenic mass replacing most of pelvocalyceal system of both kidneys. For further evaluation, unenhanced and contrast enhanced computed tomography (CT) was performed with image acquisition after delays of 20 sec and 60 sec. The lesions consisted of intrathoracic paravertebral masses, focal intrahepatic mass and masses of both pelvocalyceal systems of the kidneys. On enhanced CT scans, intrathoracic masses appeared as bilateral smoothly marginated, lobulated, mild enhancing and $6 \times 2.5 \times 2$ cm-sized paravertebral masses without mass effect or bony erosion of vertebra or rib (Fig. 1). Unenhanced CT scans showed diffuse hepatomegaly and $10 \times 8.5 \times 5.5$ cm-sized

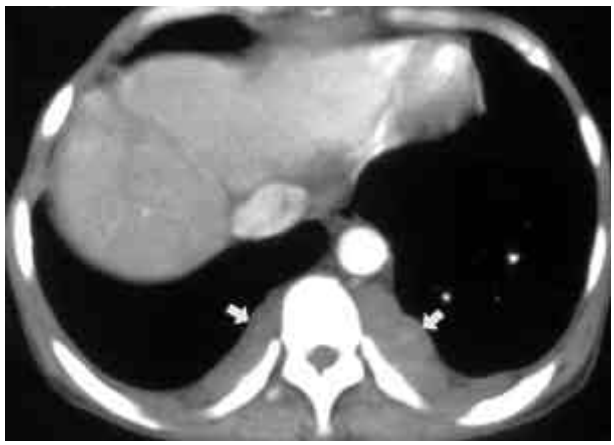

Fig. 1. Enhanced CT scan shows bilateral paravertebral masses (arrows). The masses appear with smooth margin, homogeneous attenuation and mild enhancement. No evidence for vertebral body erosion or mass effect against the adjacent aorta is noted.

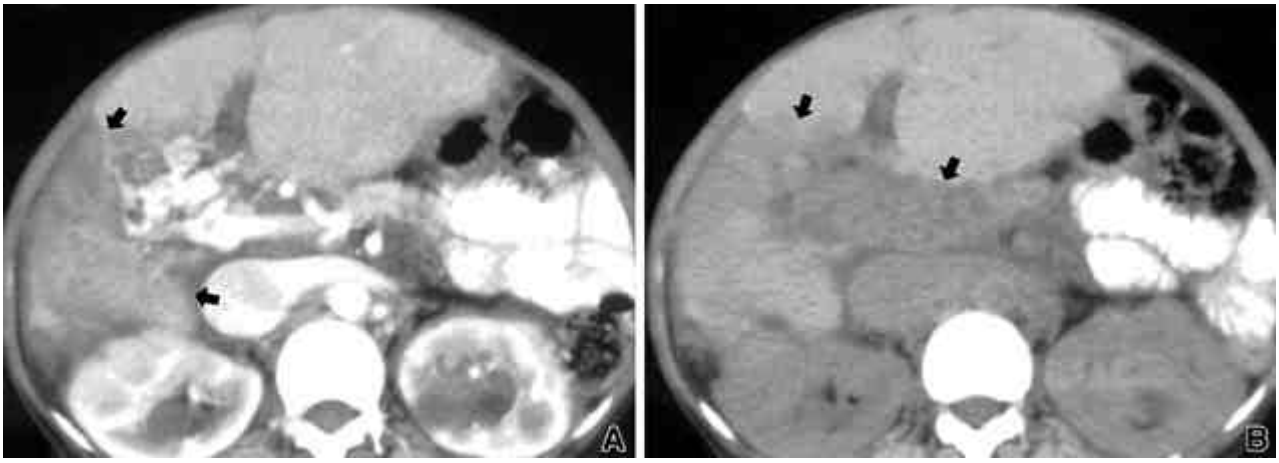

Fig. 2. A: Unenhanced CT scan of liver shows large, ill-defined low-attenuation lesion with focal surface retraction in right lobe of liver (arrows). B: Enhanced CT scans at the same level to A shows patchy enhancement of previous lesion (arrows).

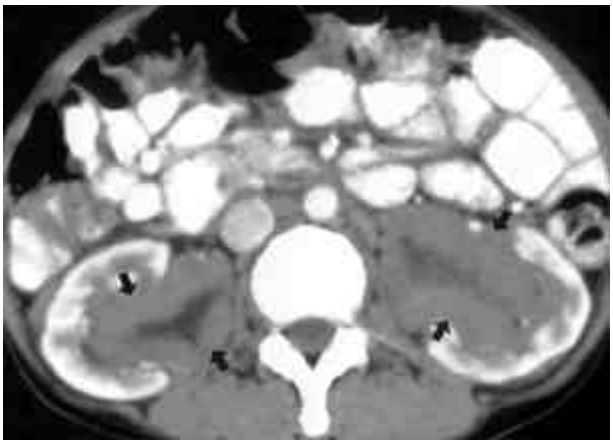

Fig. 3. Enhanced CT scan shows lobulated masses with widening and filling of pelvocalyceal systems with homogeneously mild enhancement (arrows).

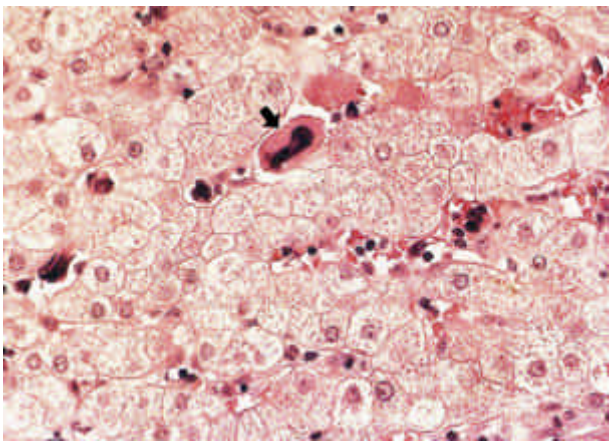

Fig. 4. Photomicrography of biopsy specimen from liver shows megakaryocyte (arrow) inside enlarged sinusoids among hepatocyte cords (H&E, ×200).

ill-defined low-attenuation mass with focal surface retraction in the right lobe of the liver (Fig. 2A). On enhanced CT scans, the lesion showed patchy enhancement (Fig. 2B). The lesions of both kidneys showed lobulated masses with widening and filling of pelvocalyceal systems on unenhanced CT scans. On enhanced CT scans, the masses showed homogeneously mild enhancement (Fig. 3). Ultrasound-guided biopsy (18G Automated Gun, Tsk Labalatory, Japan) of hepatic mass was performed. Histologically, pleomorphic clumps of erythroid and myeloid precursors together with megakaryocytes in the sinusoidal spaces (Fig. 4). Myelod precursors of red blood cell were found on urine analysis. Although thorax masses were not performed histological examination, EMH was diagnosed on imaging finding and clinical information compared to previous report (4). A final diagnosis of extramedullary hematopoiesis of the thorax, liver and both kidneys in association with idiopathic myelofibrosis were made.

DISCUSSION

Idiopathic myelofibrosis is a chronic myeloproliferative disorder characterized by marrow fibrosis, hepatosplenomegaly, the presence of myeloid and erythroid progenitors in the peripheral blood and a variable degree of anemia. EMH is a characteristic feature of the disease.

More recently, the pathogenesis of EMH has been suggested to result from hematogenous spread of multipotential stem cells, with consequent infiltration of organs and tissues. These cells are thought to be of clonal origin, according to the notion that idiopathic myelofibrosis, like other chronic myeloproliferative disorders, arises from clonal proliferation of a single pluripotent

hemopoietic stem cell (5).

Lawson et al. (6) theorized that EMH arises from the extrusion of proliferating marrow through the cortex into a subperiosteal location. This explains its presence in a paravertebral or presacral location. Embryonic rests or totipotential cells are thought to be responsible for EMH in visceral sites such as the liver, spleen, kidney, adrenal gland, lymph node, lung, pleura, skin, breasts, dura mater, ovary, thymus, gastrointestinal tract and central nervous system (2, 3). Microscopically, there is diffuse infiltration of hematopoietic cells, although there may be tumor-like masses of hematopoietic tissue with a variable degree of fibrosis (7, 8).

Intrathoracic EMH showed smoothly marginated, homogeneous, lobulated paravertebral masses, usually multiple and bilateral and caudal to the sixth thoracic vertebra (4), exhibiting coarsening of the trabecular pattern, rib expansion and posterior new bone (4) or no mass effect or bony erosion of vertebra or rib (9). In our case, the mass was well marginated and of homogeneous bilateral with soft-tissue attenuation. No evidence for vertebral body erosion or mass effect against the adjacent aorta was noted.

Only 10 cases of focal intrahepatic EMH have been reported (10). Intrahepatic EMHs were solitary in four cases and multiple in six. On unenhanced CT, the masses were described as hypodense lesions. Enhanced CT, described in two cases, showed patchy enhancement in one (11) and heterogeneous enhancement in the other (10). In the current case, unenhanced CT scans showed poorly defined low-attenuation lesion with focal surface retraction in the right lobe of the liver. Enhanced CT scans showed heterogeneous but mild enhancement of the lesion, similar to those described in the literature (10, 11). Ligumski et al. (12) reported that liver architecture such as focal surface retraction of the present case was grossly distorted because of massive impaction of the sinusoids with hematopoietic cells.

Renal involvement of EMH is usually intraparenchymal and characterized by interstitial focal infiltrates or tumor like-nodules extending into the pelvocalyceal system (13). These findings in some circumstances may be confused with hypernephroma, peripelvic cyst and other kidney neoplasms. Extensive parenchymal replacement or ureteral obstruction in these circumstances usually causes renal failure. In the current case, the masses were well-defined, homogeneously tumor-like nodule in the bilateral pelvocalyceal system. Unenhanced CT scan showed low-attenuation lesions. The masses showed homogeneously mild enhancement on enhanced CT scans. Renal failure was not seen in our case because of partial pelvic obstruction by the masses.

Extramedullary hematopoiesis involving multiple organs is an uncommon condition in patients with a history of myelofibrosis. In the present case, the masses involved multiple organs including the intrathoracic mediastinum, liver and both kidneys in a patient with idiopathic myelofibrosis were considered EMH. Final diagnosis of EMH necessitates a histopathologic examination of the biopsy specimens.

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