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Histiocytic Medullary Reticulosis: Radiologic Diagnosis of Splenic Infarction

—A Case Report—

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A case of histiocytic medullary reticulosis with splenic infaraction from a 23-year-old male is presented. Radiologic findings on selective spleen scintigraphy and abdominal CT are described. Selective spleen scintigraphy showed huge, multilobulated spleen with numerous photon-deficient areas in it and peripherally. Abdominal CT showed large peripheral band-like low density and infiltrative lesion in spleen with accompanying intraabdominal lymphadenopathy. Histological features were consistent with HMR in spleen and liver specimens.

Key Words: *Histiocytic medullary reticulosis, splenic infarct*

INTRODUCTION

Histiocytic medullary reticulosis (HMR) has been widely accepted as a distinct clinical and pathological entity (Friedman, 1965; Warnke et al., 1975). It has often escaped radiologic concern probably bacause of its rarity and unwillingness to perform imaging studies by physician. Radiologic diagnosis of HMR is a hard nut to crack, but radiologists can help understanding clinical presentation and determining precise etiology. We report radiologic findings on selective spleen scintigraphy and abdominal CT in histologically confirmed HMR.

CASE REPORT

A 23-year-old serviceman was admitted with abdominal distension, pain in upper abdomen, fever, and easy fatigability over 1 month. Physical examination on admission revealed acutely-ill looking appearance and hepatosplenomegaly without palpable subcutaneous nodule.

Initial laboratory data included hemoglobin, 15.0/dl; hematocrit, 45%; WBC count, 4,200/mm^3; platelets, 82,000/mm^3; and increased serum alkaline phosphatase and SGOT/GPT. Platelets count was falling to 42,000/mm^3 and WBC count varied between 5,500 and 3,400/mm^3 during the hospital course.

Because of hepatosplenomegaly, radioisotope liver scan was taken which revealed RES dysfunction and vague space-occupying lesions in the enlarged spleen. And selective spleen scintigraphy using 15 mCi of 99mTc-heated autologous RBC was performed which showed multilobulated spleen of 23 cm in length with many photon-deficient areas in it and peripherally (Fig. 1).

Under the impression of splenic infarct induced by hematologic malignancy, abdominal CT was performed which revealed large peripheral band-like low

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density in spleen and infiltrative low density of residual splenic parenchyma with contrast enhancement (Fig. 2A). More caudal CT image revealed interaorticocaval and mesenteric lymphadenopathy, and a few foci of high density in the peripheral low density area which proved to be recent hemorrhage in accordance with the finding of splenectomized specimen (Fig. 2B).

On the 10th day following admission, he suddenly went into shock after aspiration biopsy. Emergency laparotomy revealed splenic rupture with resulting hemoperitoneum. He expired because of irreversible hemorrhage and anoxic brain damage. Splenectomized specimen measuring 20×15×10 cm and 1,750 gm in weight showed multiple infarcted areas with grayish white infiltrative lesion on cut surface. Its friability and severe hemorrhagic necrosis hindered pathologist from taking a good gross photograph. Final pathologic diagnosis of liver and spleen was HMR (Fig. 3).

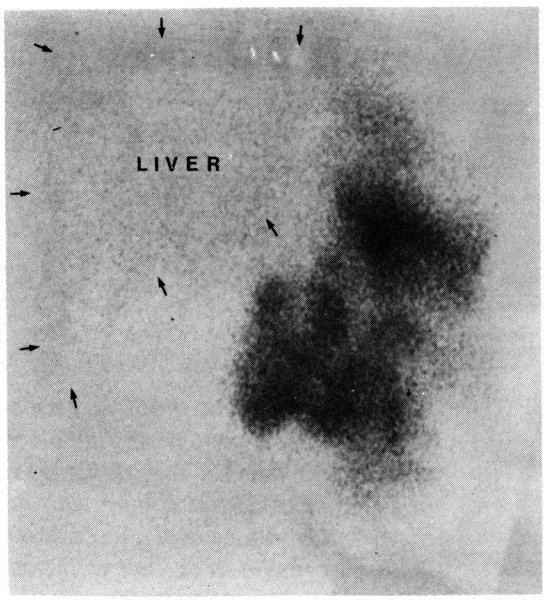

Fig. 1. Anterior view from selective spleen scintigraphy shows a large spleen with multiple defects in the parenchyma and peripheral zone. (Liver is indicated by small arrows.)

DISCUSSION

HMR, also called malignant histiocytosis, is a rare, usually rapidly progressive systemic disease. Clinical presenting symptoms of a typical case of HMR are

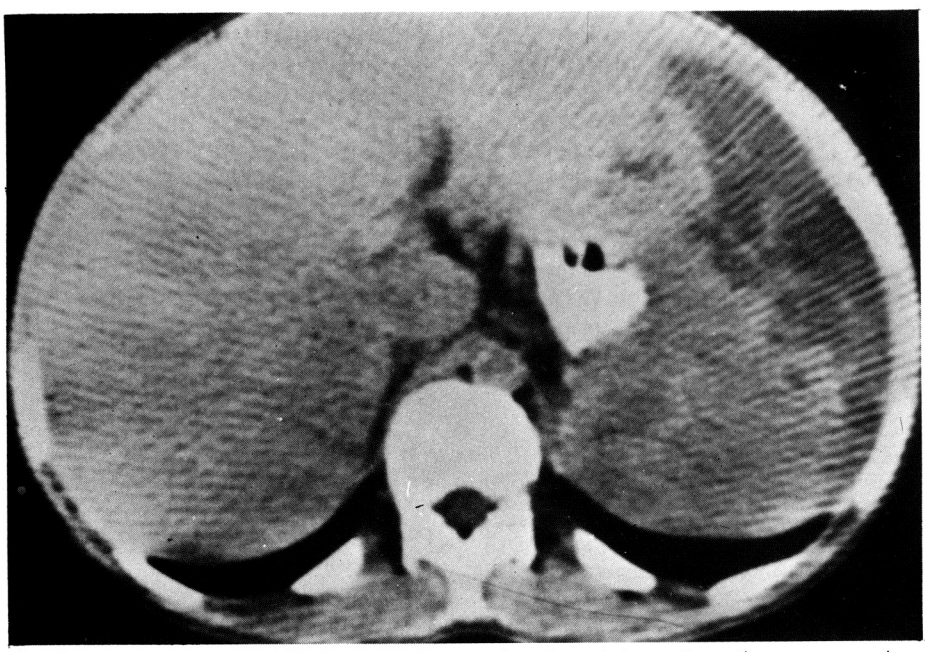

Fig. 2. A, Contrast-enhanced CT shows peripheral wedge-shaped low attenuation areas and intraparenchymal infiltrative lesion in spleen.

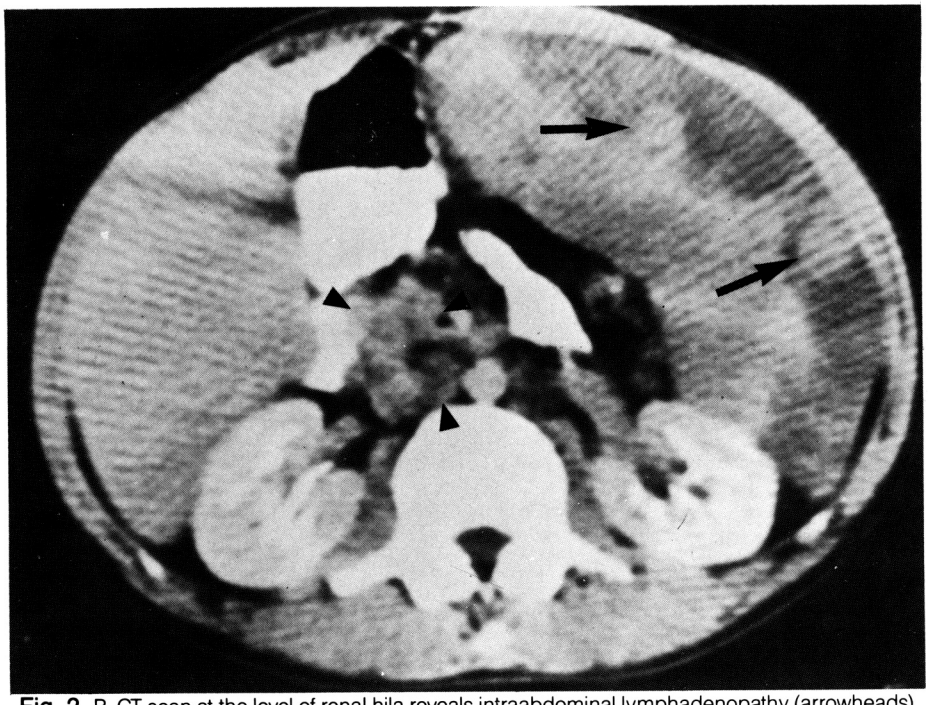

Fig. 2. B, CT scan at the level of renal hila reveals intraabdominal lymphadenopathy (arrowheads) and two foci of high density (arrows) in the band-like splenic infarction.

fever, wasting, lymphadenopathy, hepatosplenomegaly, and progressive pancytopenia. The disease runs a rapid course, usually terminating fatally within six months after the onset of symptoms (Friedman and Steigbigel, 1965). At postmortem examination the most frequent gross findings are splenomegaly and hepatomegaly. In this case the spleen weighed 1,750gm which is heavier than the two surgically removed cases reported by Warnke et al. The typical microscopic finding of HMR is proliferation of histiocytes showing intense erythrophagocytosis in the spleen, liver and bone marrow (Warnke et al., 1975). This case showed progressive leukopenia and thrombocytopenia with unusually normal hematocrit during the hospital course. The splenic cut surface was more similar to leukemia than lymphoma.

We want to emphasize the usefulness of selective spleen scintigraphy and CT in the clinical setting of hematologic disease with hepatosplenomegaly. Bone marrow biopsy and laboratory data can suggest HMR in most cases, but it is mandatory to perform radiologic study to know the extent of lymphadenopathy and the existence of splenic infarction accurately. The spleen may be secondarily involved in many systemic diseases, as well as by a small number of primary processes. It is difficult organ to evaluate clinically. Nuclear scintigraphy, ultrasonography and CT offer a relatively noninvasive and often reliable means of examining the spleen (Freeman and Tonkin, 1976; Front et al., 1984; Costello et al., 1985). Splenic infarcts are comparatively common lesions caused by occlusion of the major splenic artery or any of its branches by systemic emboli. Much less often, infarcts in the spleen are caused by local thromboses, especially in the myeloproliferative syndromes, Hodgkin's lymphoma, leukemia, and pancreatitis (Haaga and Alfidi, 1983). The splenic artery is an end artery supplying blood to the conical areas; therefore, splenic infarction is usually demonstrated as a wedge-shaped area and peripheral in location. The superimposition of multiple infarctions will produce variously shaped lesion (Lin and Donati, 1981). CT images of the splenic infarct as a complication of the hepatic arterial embolization showed multiple low density areas of a wedge or fused-wedge shape in most patients and of a rod or round shape in others (Takayasu et al., 1984). These infarcts

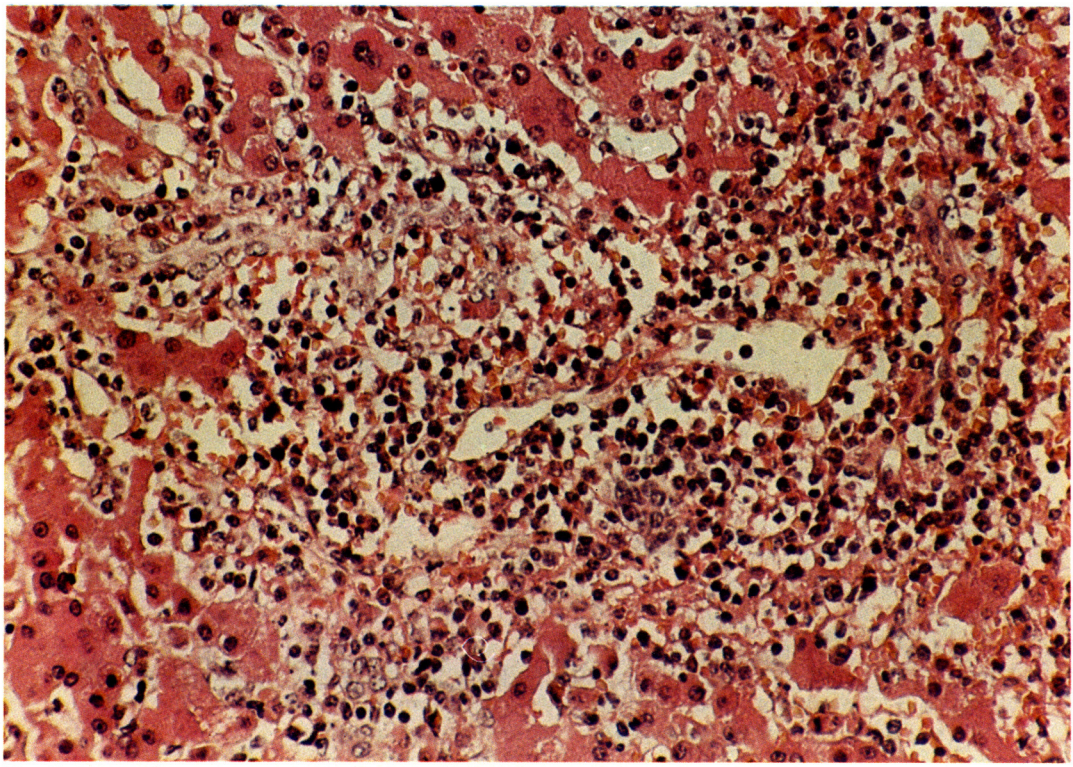

Fig. 3. Photomicrograph of the liver shows typical pattern of involvement included both a portal and sinusoid proliferation of atypical histiocytes and erythrophagocytosis

may be silent clinically or the source of the left upper quadrant pain. Symptomatic splenomegaly owing to infarct and hemorrhage will be the warning sign of spontaneous rupture and catastrophic state. So it is helpful to perform splenic imaging in hematologic diseases which show splenomegaly including abdominal pain.

In summary, HMR can be diagnosed earlier in patients who have clinical findings of fever, hepatosplenomegaly, lymphadenopathy, and progressive pancytopenia with corroborative radiologic evidence of splenic involvement with or without infarction. Awareness of splenic infarction in HMR will help to manage the patient properly.

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