

Scintigraphic diagnosis of intrathoracic extramedullary hematopoiesis in a patient with β -thalassemia

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We report the scintigraphic diagnosis of intrathoracic extramedullary hematopoiesis (EMH) in a patient with β -thalassemia. A patient had a mass in the right thoracic paravertebral region on radiography and CT. Bone marrow imaging of the thorax by means of both Tc-99m nanocolloid and phytate demonstrated uptake of the tracer in the mediastinal mass, establishing the diagnosis of EMH.

Key words: extramedullary hematopoiesis, bone marrow imaging, β -thalassemia

INTRODUCTION

EXTRAMEDULLARY HEMATOPOIESIS (EMH) refers to the production of blood cells outside the skeleton and is a compensatory mechanism for bone marrow dysfunction. It usually occurs in chronic hemolytic anemias, myelophthisic anemias and myeloproliferative disorders.¹ The most common involvement sites are the spleen, liver and lymph nodes. In patients with intrathoracic mass lesions and suspected EMH, marrow scintigraphy may be helpful in the differential diagnosis. Uptake of marrow agent in the mass site indicates EMH.^{2,3} We report a patient with β -thalassemia, who has a mediastinal mass documented as EMH by means of both Tc-99m nanocolloid and phytate marrow scintigraphy.

CASE REPORT

A 55-year-man with β thalassemia was referred to our hospital because of dyspnea and tachycardia. At physical examination, anemic face, hepatomegaly and skeletal deformities were found. He had a past history of splenectomy for which he had been treated 14 years ago. The hemoglobin level was 7.73 g/dl, the hematocrit was 23.4% and mean corpuscular volume was 75.9 μm^3 . The

white blood cell count and platelet count were normal. Biochemical abnormal laboratory data included glutamate oxaloacetate transaminase: 222 U/L, glutamate pyruvate transaminase 97 U/L, lactic dehydrogenase: 1,006 U/L and gamma-glutamyl transferase: 82 U/L, all high values. Serum ferritin was in excess of 1,500 ng/ml.

Chest radiography showed a right paravertebral mass (Fig. 1). Computed tomography of the thorax revealed a mass in the posterior mediastinum in the same location as in chest radiography (Fig. 2). A diagnosis of neurogenic tumor was suggested by the radiologists.

Since intrathoracic EMH was considered in the differential diagnosis, bone marrow scintigraphy was performed. Because the use of small colloids (particle size ≤ 80 nm) causes greater marrow uptake than larger particle sized conventional colloids such as phytate,⁴ in bone marrow imaging we routinely use Tc-99m nanocolloids which have a mean diameter of about 40 nm. Therefore the first radionuclide study was performed with 370 MBq of Tc-99m nanocolloid. Anterior and posterior chest and lower extremity images were acquired. We also performed phytate scintigraphy in order to compare the quality of the images in nanocolloid and phytate scintigraphy. This study was performed after the administration of 300 MBq of Tc-99m phytate. Nanocolloid scintigraphy demonstrated right paravertebral activity corresponding to the mass seen on CT and peripheral expansion of the bone marrow (Fig. 3A, and Fig. 4). Phytate scintigraphy also showed right paravertebral activity (Fig. 3B). But the lesion intensity was less than that with Tc-99m nanocolloid.

We concluded that these abnormal activities repre-

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Fig. 1 PA chest radiography shows a mediastinal mass on the right paravertebral region.

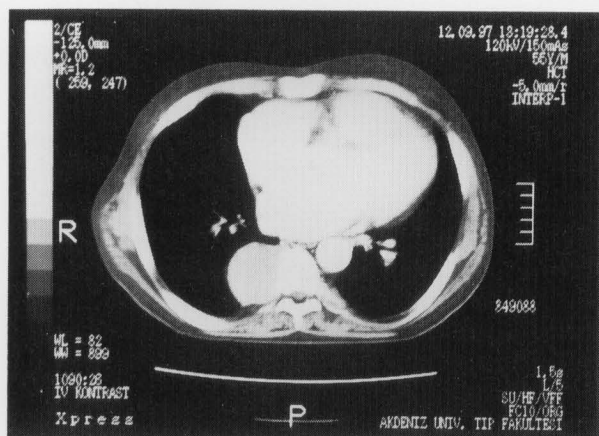
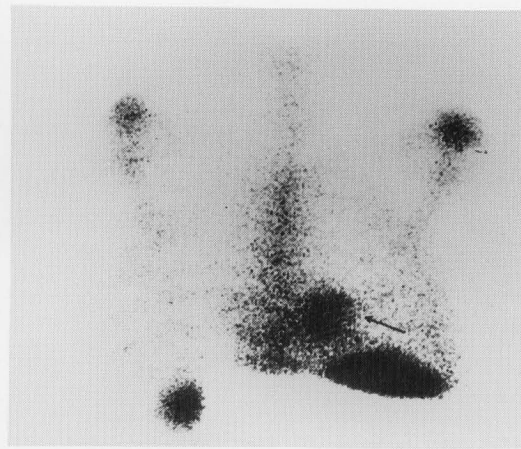


Fig. 2 CT shows a mediastinal mass on the posterior mediastinum at the same location with chest radiography.

sented EMH due to β -thalassemia. Because the patient refused further examination, histologic confirmation could not be performed.

DISCUSSION

Development of normal marrow outside of the bone marrow is defined as EMH and usually occurs in chronic hemolytic anemias, myelophthisic anemias and myeloproliferative disorders. The spleen, liver and lymph nodes are the most commonly involved sites. Less frequently involved sites include the kidney, adrenal glands, breast, spinal cord, intrathoracic cavity, pleura, pericardium, and intracranial cavity.³ Intrathoracic EMH is most often visualized on the chest X-ray or the chest CT scan as a single or multiple paravertebral mass lesions.⁵ While a paravertebral mass may be EMH, other disorders of the posterior mediastinum such as neurogenic tumor, lymphoma, and primary and metastatic malignancy must be considered. The differential diagnosis of these masses is



A



B

Fig. 3 A: Posterior image of the chest with Tc-99m nanocolloid and B: Posterior image of the chest with Tc-99m phytate demonstrate intense uptake of the radioisotope by the right paravertebral mass comparable to the uptake by the liver. It is obviously seen that the intensity of the mass in nanocolloid image is markedly higher than that of phytate image.

important in management of the treatment. Differential diagnosis can be made with clinical findings and radiologic examinations. Patients who have low density hematopoietic masses on CT^{6,7} and a high signal on both T1- and T2-weighted MRI⁸ appear to have mediastinal tumors consisting of bone marrow. Histologic diagnosis requires transthoracic biopsy or thoracic surgery. Since the extraskelatal marrow is highly vascular and these procedures can cause hemorrhagia, noninvasive procedures are necessary in order to explain the nature of these tissues.⁵

Bone marrow scintigraphy has been used for evaluating the bone marrow involvement in inflammatory, hematological and circulatory disorders and metastases.⁹ In addition, this imaging method has been used in the investigation of mass lesions that may represent EMH.

Radio-iron and analogous are the most specific agents for erythroblastic marrow imaging. Both Fe-52 and Fe-59 have been used to demonstrate EMH.^{2,10} Fe-52 is of

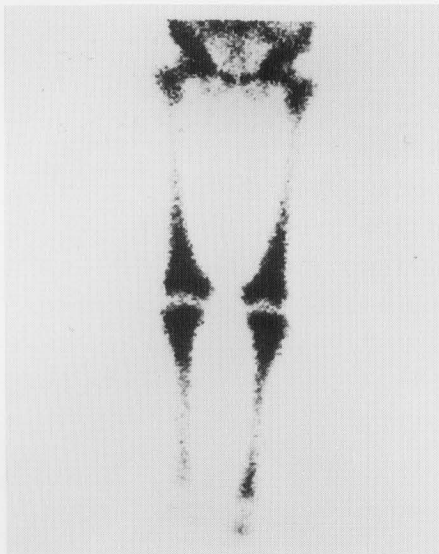


Fig. 4 Anterior lower extremities image with Tc-99m nanocolloid demonstrates peripheral bone expansion.

limited availability. Fe-59 emits high-energy photons and for adequate imaging special equipment is necessary.^{11,12}

Indium-111 chloride has been proposed as an erythroblastic marrow agent, but the mechanism of uptake of In-111 chloride is different from that of the iron-isotopes.¹¹ Although it binds to transferrin similarly to iron, only 4% of the injected activity appears in the circulating erythrocytes within 8–10 days as to 80% for iron. In-111 chloride may also be taken up by the reticuloendothelial cells after binding to transferrin, but in patients with saturated iron-binding capacities, In-111 may not bind to transferrin.⁹

The introduction of Tc-99m colloids has made reticuloendothelial imaging feasible; the radiation dose is acceptable and the images are of good quality. Colloids are removed from the blood by cells of the reticuloendothelial system. In normal adults about 10% of the injected activity will be distributed in the reticuloendothelial cells of the marrow and the remainder will be taken up by the liver (80%) and the spleen (10%). Reticuloendothelial activity generally corresponds to erythroblastic activity. In patients with suspected peripheral expansion of the marrow as well as EMH, radiocolloids appear to be agents of choice in demonstrating the extent of marrow expansion and the presence of EMH.^{5,11}

In our patient we detected EMH by using both Tc-99m nanocolloid and Tc-99m phytate. We observed that the lesion intensity was much higher in the nanocolloid image than in the phytate image. It was thought that this differ-

ence in the intensity of lesion was due to the smaller size of the nanocolloids than that of the phytate.

Since full bone marrow complement is present in EMH sites, both RES and erythropoietic agents are useful in diagnosing these lesions.¹² Because of availability, imaging characteristics, dosimeter and cost, we believe that Tc-99m colloids currently are the best choice for routine marrow imaging as well as for assessing possible EMH.

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