Annals of Nuclear Medicine Vol. 20, No. 8, 565–568, 2006

Tc-99m nanocolloid scintigraphic imaging of intracranial meningeal extramedullary hematopoiesis in a patient with idiopathic myelofibrosis

Halil KAYA* and Sevim Sureyya CERCI**

*Department of Nuclear Medicine, Dicle University, School of Medicine, Diyarbakir, Turkey
**Department of Nuclear Medicine, Suleyman Demirel University, School of Medicine, Isparta, Turkey

Meningeal extramedullary hematopoiesis (EMH) is a rare finding in idiopathic myelofibrosis. Intracranial EMH is typically asymptomatic and sites are usually found by chance. Diagnosis of EMH is difficult, based on clinical circumstances and the use of different diagnostic imaging modalities, such as CT, MRI or radionuclide imaging. We present a case with intracranial medullary hematopoiesis due to idiopathic myelofibrosis diagnosed with Tc-99m nanocolloid scintigraphy. Cranium SPECT images that were performed with Tc-99m nanocolloid showed increased radiotracer uptake in the bilateral parietal, bilateral frontal and left occipital bones and especially in falx cerebri of sinus sagittalis superior. In Tc-99m MDP bone scintigraphy, increased osteoblastic activity in the left frontal and parietal bones, in shoulders, knee and ankle joints, and in both metatarsal bones were seen. After gadodiamid injection, T1 weighted MRI showed diffuse contrast increased in the meningeal areas surrounding the brain. A biopsy of the mass revealed extramedullary hematopoiesis composed of erythroblasts, mature and immature myeloid cells, and megakaryocytes. It was deduced that these described foci of EMH.

Key words: meningeal extramedullary hematopoiesis, idiopathic myelofibrosis, Tc-99m nanocolloid scintigraphy

INTRODUCTION

EXTRAMEDULLARY HEMATOPOIESIS (EMH) originates from multipotential cells in any tissue and typically involves the liver, spleen, and lymph nodes. Other common sites include the intrathoracic cavity, paraspinal regions, kidney, adrenal glands and thyroid.[1–5] However, meningeal EMH has rarely been reported in cases with idiopathic myelofibrosis.[6–9] Intracranial EMH most frequently involves the cranial dura and falx, followed by the cerebral parenchyma, optic nerve sheath and the diploic space of the skull.[7] Intracranial EMH is typically asymptomatic and sites are usually found by chance. On the other hand extramedullary hematopoiesis in the intracranial or intraspinal epidural space can lead to serious neurologic complications including subdural hemorrhage, delirium, increased intracranial pressure, papilledema, coma, motor and sensory impairment, and limb paralysis.[9–13]

Diagnosis of EMH, based on clinical circumstances and the use of different diagnostic imaging modalities, such as CT, MRI or radionuclide imaging, is difficult.

Radionuclide scintigraphy can be performed for detect or confirm sites of extramedullary hematopoiesis with different colloids, Fe-52, In-111 and Tc-99m labeled monoclonal antibodies.[14–21] Because of the smaller size of the particles (almost 95% of particles are least than 0.08 μm in size with a mean size of 0.03 μm) more nanocolloid localizes in the bone marrow (~15%) relative to other colloids (2–5%).[22]

We present a case with intracranial medullary hematopoiesis due to idiopathic myelofibrosis diagnosed with Tc-99m nanocolloid scintigraphy.

CASE REPORT

An 18-year-old man with severe headache, dyspnea, fatigue, and anemia was admitted to the hematology clinic for investigation and treatment. There was no history of

Received November 21, 2005, revision accepted July 25, 2006.

For reprint contact: Sevim Sureyya Cerci, M.D., Modernevler 3103 sok. No:16, Isparta, TURKEY.
E-mail: sureyyacerci@hotmail.com

trauma or primary malignancy. He was diagnosed as having myelofibrosis when he was 12 years old. On clinical examination, hepatomegaly was found. Splenectomy had been performed for thrombocytopenia and painful splenomegaly two months earlier. Bone marrow aspiration biopsy revealed fibrosis and focal hypercellular marrow and megakaryocytic hyperplasia. MRI showed diffuse signal enhancement surrounding the brain (Fig. 1). Bone marrow of whole body and cranium SPECT images were acquired 45 minutes after the injection of 370 MBq Tc-99m nanocolloid intravenously. Cross sections of transaxial, sagittal and coronal images of the brain were obtained during the SPECT study. Increased radiotracer uptake was observed in the bilateral parietal, bilateral frontal and left occipital bones and especially in falx cerebri of sinus sagittalis superior (Figs. 2 and 3). In planar images, the liver was enlarged and radiotracer uptake was increased. In addition, increased radiotracer involvement was observed in the femur and tibia (Fig. 4). On the other hand whole body bone scintigraphy was performed using 740 MBq technetium-99m methylenediphosphonate (Tc-99m MDP) too. Bone scintigraphy showed increasing osteoblastic activity in the left frontal and parietal bones, in shoulders, knee and ankle joints, and in both metatarsal bones (Fig. 5). A biopsy of the mass revealed extramedullary hematopoiesis composed of erythroblasts, mature and immature myeloid cells, and megakaryocytes. It was deduced that these described foci of EMH.

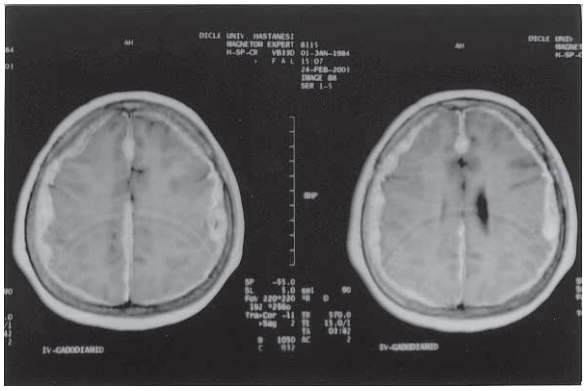

Fig. 1 After gadodiamid injection, T1 weighted MRI showed diffuse contrast increased in the meningeal areas surrounding the brain.

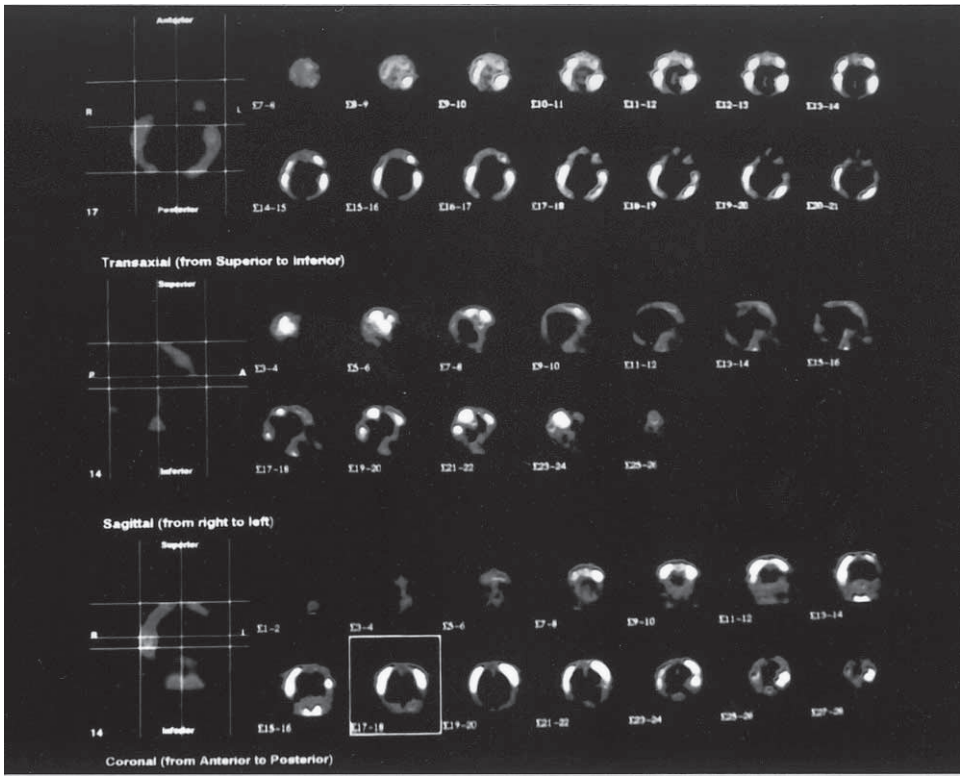

Fig. 2 Cross sections of transaxial, sagittal, and coronal images of the brain were obtained during the SPECT study with Tc-99m nanocolloid. There was increased radiotracer uptake in bilateral parietal, bilateral frontal and left occipital bones and especially in falx cerebri of sinus sagittalis superior.

DISCUSSION

There are two main groups in EMH. The first group shows paraosseous foci that may result from herniation of medullary tissue from the underlying bone and is seen in hemolytic disorders such as thalassemia and sickle cell anemia, where the marrow has tremendous activity. The second group shows extraosseous soft tissue foci, which may arise from multipotential stem cells. This happens when the marrow activity is ineffective, as in idiopathic myelofibrosis or, rarely, with toxic or tumoral marrow destruction.[6]

Lund and Aldridge published the first description of the cranial imaging findings in EMH.[6] Landolfi et al. reported the occurrence of meningeal masses causing exophthalmos and fever in a patient with myelofibrosis secondary to polycythemia vera.[8] Also Urman et al. confirmed the presence of a meningeal intracranial mass due to EMH with radionuclide imaging using Tc-99m nanocolloid.[9]

Although diagnostic radiologic methods such as computed tomography and magnetic resonance imaging can be suggestive of the presence of extramedullary hematopoiesis, neither of them can confirm the nature of hematopoietic tissue.[21] Effective diagnosis involves a specific, reliable, whole-body and low-cost method of screening.

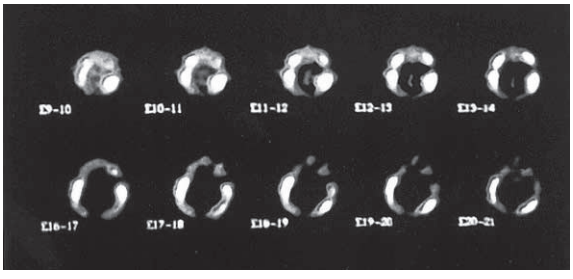

Fig. 3 Increased radiotracer uptake in transaxial section.

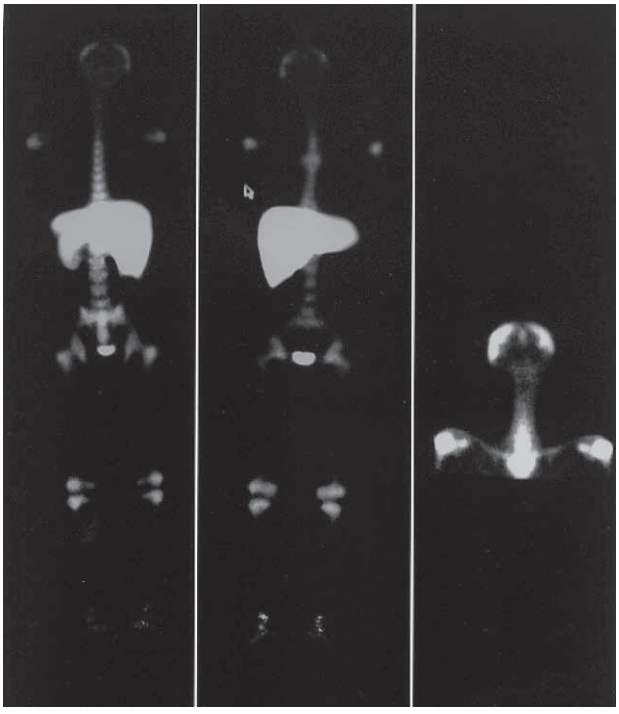

Fig. 4 In planar images, the liver was enlarged and radiotracer uptake was increased. In addition, increased radiotracer involvement was observed in the femur and tibia.

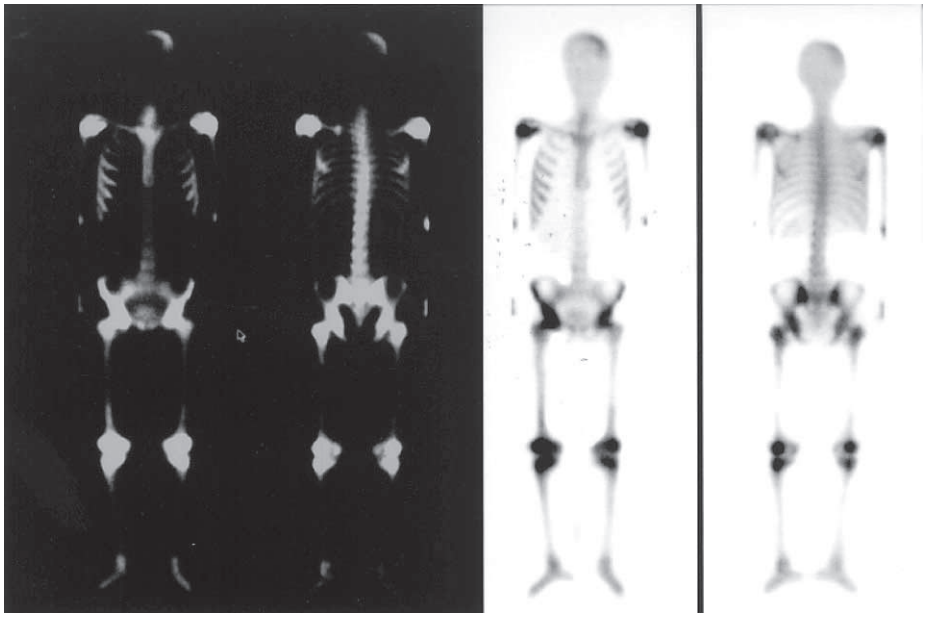

Fig. 5 Tc-99m MDP bone scintigraphy showed increasing osteoblastic activity in the left frontal and parietal bones and in shoulders, knee and ankle joints, and in both metatarsal bones.

Tc-99m labeled colloids allow visualization of bone marrow elements wherever they might arise. In the case of suspected EMH, only scintigraphic methods can detect and confirm the nature of hematopoietic tissue. However, biopsy is needed to provide cytologic or histologic confirmation.[23]

Radionuclide scintigraphy has been used in a number of patients to detect or confirm sites of extramedullary hematopoiesis imaging with colloids, Fe-52, In-111, Tc-99m labeled monoclonal antibodies.[14–21]

Identifying EMH using Tc-99m labeled colloids, especially sulfur colloids, combines several advantages of the different approaches to scintigraphic detection listed above. Tc-99m labeled colloids are reliable whole body radionuclides, are readily available, and provide a low cost method for detecting hematopoiesis. However, this kind of scintigraphy has disadvantages, caused by the distributional properties of the substance. The reticuloendothelial rather than erythropoietic cells trap the colloids .[21] Since a full bone marrow complement of erythroid, reticuloendothelial and megakaryocytic cells is present in the areas of EMH both reticuloendothelial system and erythropoietic agents can be used.[9]

In our patient the diagnosis of EMH was based on localization of Tc-99m nanocolloid in intracranial masses, the nature of which was suggested with MRI.

In MDP bone scintigraphy increased uptake of radiotracer in extramedullary hematopoiesis may be confused with osteoblastic activity.

We reported a very rare case with extramedullary meningeal hematopoiesis and emphasized that nanocolloid scintigraphy has some advantages for the detection of medullary and extramedullary hematopoiesis compared to MRI and computerized tomography imaging.

REFERENCES

1. Cermik TF, Turgut B, Gultekin A, Cakır B, Berkarda S. Diffuse uptake of Tc-99m tin colloid in extramedullary hematopoiesis of the lungs. *Clin Nucl Med* 2003; 28: 410–412.
2. Bartlett RP, Greipp PR, Tefferi A, Cupps RE, Mullan BP, Trastek VF. Extramedullary hematopoiesis manifesting as a symptomatic pleural effusion. *Mayo Clin Proc* 1995; 12: 1161–1164.
3. Schnuelle P, Waldherr R, Lehmann KJ, Woenckhaus J, Back W, Niemir Z, et al. Idiopathic myelofibrosis with extramedullary hematopoiesis in the kidneys. *Clin Nephrol* 1999; 4: 256–262.
4. Wat NM, Tse KK, Chan FL, Lam KS. Adrenal extramedullary hematopoiesis diagnosis by a non-invasive method. *Br J Hematol* 1998; 100: 725–727.
5. Lazzi S, Als C, Mazzucchelli L, Kraft R, Kappeler A, Laissue J. Extensive extramedullary hematopoiesis in a thyroid nodule. *Mod Pathol* 1996; 9: 1062–1065.
6. Lund RE, Aldridge NH. Computed tomography of intracranial extramedullary hematopoiesis. *J Comput Assist Tomogr* 1984; 4: 788–790.
7. Ohtsubo M, Hayashi K, Fukushima T, Chiyoda S, Takahara O. Case report: intracranial extra-medullary haematopoiesis in postpolycythemic myelofibrosis. *Br J Radiol* 1994; 795: 299–302.
8. Landolfi R, Colosimo C Jr, De Candia E, Castellana MA, De Cristofaro R, Trodella L, et al. Meningeal hematopoiesis causing exophthalmos and hemiparesis in myelofibrosis: effect of radiotherapy. *Cancer* 1988; 11: 2346–2349.
9. Urman M, O'Sullivan RA, Nugent RA, Lentle BC. Intracranial extramedullary hematopoiesis. CT and bone marrow scan findings. *Clin Nucl Med* 1991; 6: 431–434.
10. Rizzo L, Greco Crasto S, Tola E, Sardo P, Rubino A. Extramedullary hematopoiesis: unusual meningeal and paranasal sinuses presentation in Paget. *La Radiol Med* 2003; 105: 376–381.
11. Chourmouzi D, Pistevou-Gompaki K, Plataniotis G, Skaragas G, Papadopoulos L, Drevelegas A. MRI findings of extramedullary haemopoiesis. *Eur Radiol* 2001; 11: 1803–1806.
12. Rowe DW, Barest G. Neuroradiology case of the day. Spinal cord compression caused by extramedullary hematopoiesis. *Am J Roentgenol* 1999; 173: 806, 810–812.
13. Lall C, Payne K. A patient with anemia and paraspinal chest mass. *Chest* 2003; 124: 732–734.
14. Parker LA, Vincent LM, Mauro MA, Perry JR. Extramedullary hematopoiesis: demonstration by transmission and emission computed tomography. *Clin Nucl Med* 1986; 11: 1–3.
15. Bronn JL, Paqualet JR, Tetalman MR. Intrathoracic extramedullary hematopoiesis. Appearance of ^{99m}Tc sulfur colloid marrow scan. *Am J Roentgenol* 1980; 134: 1254.
16. Stebner FC, Bishop CR. Bone marrow scan and radioiron uptake of an intrathoracic mass. *Clin Nucl Med* 1982; 7: 86–87.
17. Wosick WF, Bedmutha S, Small B. Radionuclide bone marrow scanning and computerized tomography in the diagnosis of extramedullary hematopoiesis presenting as an intrathoracic paravertebral mass. *Clin Nucl Med* 1984; 9: 419.
18. Borgies P, Ferrant A, Leners N. Diagnosis of heterotopic bone marrow in the mediastinum using Fe-52 and positron emission tomography. *Eur J Nucl Med* 1989; 15: 761–763.
19. Adams BK, Jacops P, Byrne MJ. Fe-52 imaging of extramedullary hematopoiesis in a patient with beta thalassemia. *Clin Nucl Med* 1995; 20: 619–622.
20. Lilien DL, Berger HG, Anderson DP, Benett LR. ^{111}In chloride: a new agent for bone marrow imaging. *J Nucl Med* 1973; 14: 184–186.
21. Moellers MO, Bader JB, Aexander C, Samnick S, Kirsch CM. Localization of extramedullary hematopoiesis with Tc-99m-labeled monoclonal antibodies (BW 250/183). *Clin Nucl Med* 2002; 27: 354–357.
22. Saha GB. Characteristics of Specific Radiopharmaceuticals. In: *Fundamentals of Nuclear Pharmacy*—4th ed. New York; Springer-Verlag, 1998: 112–147.
23. Loh CK, Alcorta C, McElhinney AJ. Extramedullary hematopoiesis simulating posterior mediastinal tumors. *Ann Thorac Surg* 1996; 3: 1003–1005.