A Characteristic but Rare Skeletal Scintigraphy Findings in Sickle Cell Disease: A Case Report

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Abstract

Sickle cell disease is associated with recurrent vaso-occlusive episodes leading to a wide spectrum of skeletal complications. We report the case of a 39-year-old male with sickle cell disease who underwent bone scintigraphy for evaluation of right hip pain following bilateral hip arthroplasty. The study demonstrates characteristic skeletal and splenic scintigraphy findings in sickle cell disease and emphasizes the diagnostic value of bone scintigraphy in identifying skeletal manifestations of the disease.

Keywords: sickle cell disease, bone scan, splenic uptake.

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Introduction

Sickle cell disease is a hereditary hemoglobinopathy characterized by recurrent vaso-occlusive episodes leading to multisystem involvement. Skeletal complications are among the most common clinical manifestations in these patients. Although they do not directly contribute to mortality, they are responsible for significant morbidity in patients with sickle cell disease. These complications include bone marrow infarctions, avascular necrosis, osteomyelitis, orbital compression, marrow hyperplasia causing osteopenia and growth disturbances, and pathological fractures (1). Bone scintigraphy using technetium-99m methylene diphosphonate (99mTc-MDP) is a highly sensitive modality for detecting alterations in bone metabolism and is frequently employed in the evaluation of musculoskeletal pain in these patients.

This case highlights rare but characteristic findings on bone scintigraphy in sickle cell disease, emphasizing its role in diagnosis and clinical management.

Case presentation

A 39-year-old male with known sickle cell disease presented with new-onset right hip pain. His medical history was significant for avascular necrosis of both femoral heads, for which he had undergone bilateral total hip replacement 15 years earlier. There was no history of recent trauma or infection. The laboratory parameters including leukocyte counts, C reactive protein (CRP) and ESR were within normal limits. The patient was referred for radionuclide bone scintigraphy to evaluate the suspected loosening of the hip prosthesis.

Three-phase 99mTc-MDP bone scintigraphy was performed following intravenous administration of the radiotracer. Flow and blood-pool images of the pelvis and hips showed no evidence of hyperemia or increased soft-tissue activity. Delayed whole-body planar images demonstrated a focal area of increased tracer uptake in the right acetabular region (Fig. 1A). Hybrid single-photon emission computed tomography/computed tomography (SPECT/CT) was subsequently performed, which localized the increased activity to a cortical linear defect involving the right acetabulum, consistent with a fracture line adjacent to the prosthesis (Fig 1C & D).

Planar images revealed focal radiotracer accumulation in the left upper quadrant of the abdomen (Fig. 1B). Focused SPECT/CT

localized this uptake to a small, atrophic spleen with dense calcifications (Fig. 1E & F). In addition, the whole-body images demonstrated area of increased tracer uptake involving the left proximal tibia bone. CT changes showed multiple patchy lucent area with diffuse skeletal sclerosis, likely secondary to longstanding marrow hyperplasia and repeated vaso-occlusive events associated with sickle cell disease (Fig. 1G).

Discussion

Bone involvement is one of the most frequent manifestations of sickle cell disease and represents a major cause of both acute and chronic morbidity. Bone scintigraphy using technetium-99m methylene diphosphonate (99mTc-MDP) is a sensitive modality for detecting skeletal involvement, particularly in patients presenting with bone pain and related complications.

Extraskeletal uptake of 99mTc-MDP is uncommon and has been described in sickle cell disease and other hemoglobinopathies in a limited number of case reports, most commonly involving the spleen and less frequently the kidneys (2-3). In patients with sickle cell disease repeated splenic infarctions leads to progressive fibrosis, dystrophic calcification, and eventual autosplenectomy. In addition, recurrent transfusions may result in hemosiderosis and iron deposition. These processes promote retention of bone-seeking radiopharmaceuticals, resulting in extraskeletal tracer accumulation on bone scintigraphy (4). Recognition of this characteristic finding is important, as splenic uptake may otherwise be misinterpreted as arising from adjacent renal, adrenal, pancreatic, or osseous structures. Hybrid SPECT/CT is particularly valuable in accurately localizing tracer uptake and confirming its extraskeletal origin. Bone scintigraphy is also commonly employed for evaluating periprosthetic pain and for detecting complications such as loosening, infection, mechanical overloading, and occult fractures. The addition of flow and blood-pool images improves diagnostic confidence in the assessment of osteomyelitis and prosthetic joint infection.

The appearance of bone infarction on scintigraphy varies with the temporal stage of ischemia. Acute infarcts may demonstrate reduced or photopenic tracer uptake due to compromised perfusion, whereas subacute and chronic infarcts often show increased uptake related to revascularization and repa-

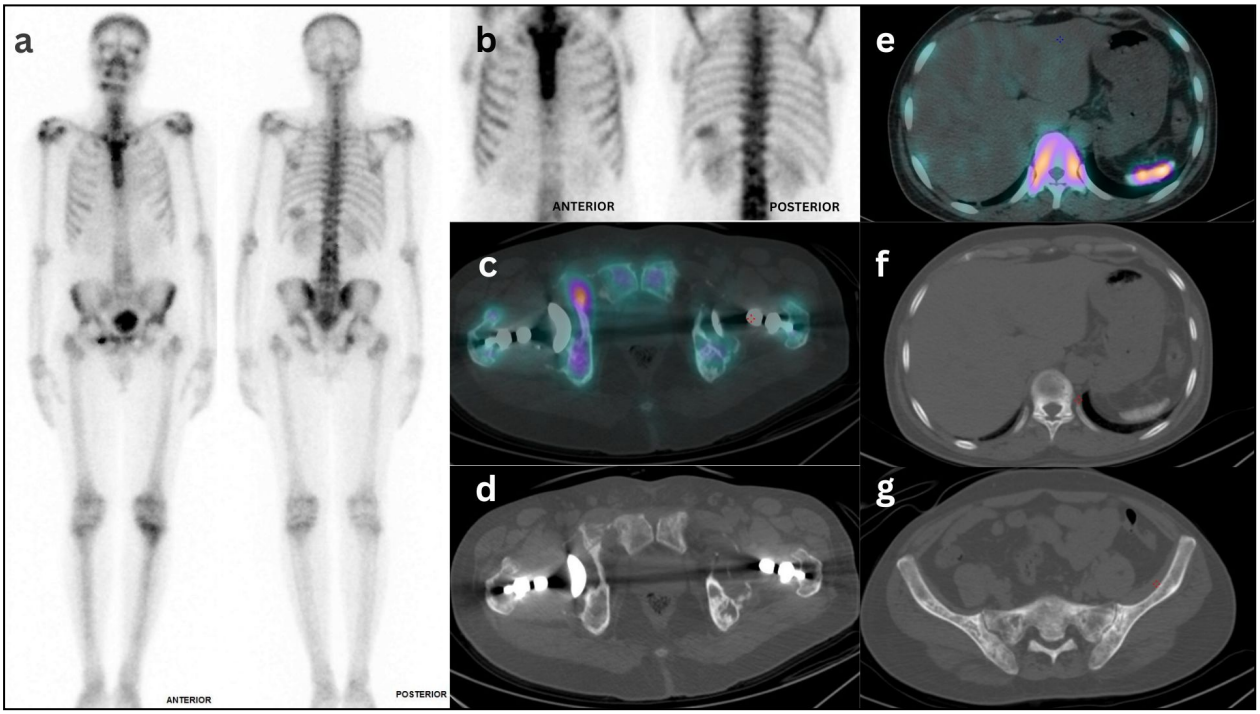

Figure 1: Maximum-intensity-projection (MIP) images from ^{99m}Tc -MDP bone scintigraphy showing focal tracer uptake in the right acetabular region and an additional focus in the left upper quadrant (a-b). Limited-field SPECT/CT localizes the right-sided uptake to a cortical fracture line of the acetabulum (c-d) and demonstrates tracer accumulation within a small, atrophic and calcified spleen (e-f). Additional skeletal changes of sickle cell disease, including patchy lucent areas and diffuse sclerosis due to repeated infarctions are also noted (g).

rative osteoblastic activity. Magnetic resonance imaging (MRI) is also sensitive modality and commonly employed for evaluating bone marrow infarction and avascular necrosis in sickle cell disease, particularly for assessing marrow signal changes, epiphyseal involvement, and associated soft-tissue complications (5).

This case highlights the importance of recognizing characteristic skeletal scintigraphic patterns in sickle cell disease and underscores the continued relevance of 99mTc-MDP bone scintigraphy in clinical practice (6).

Conclusion

Sickle cell disease is associated with a wide spectrum of skeletal complications resulting from recurrent vaso-occlusive events. Incidental splenic uptake on 99mTc-MDP bone scintigraphy, although uncommon, is a recognized finding in patients with sickle cell disease and typically reflects chronic splenic infarction and autosplenectomy. Awareness of these characteristic patterns allows accurate interpretation and improves diagnostic confidence. Bone scintigraphy remains a valuable tool in the comprehensive evaluation of skeletal complications in patients with sickle cell disease.

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Declarations

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