Assessment of Treatment Response by Bone SPECT-CT in a Case of Dermatomyositis with Calcinosis Cutis

Abstract

Calcinosis of soft tissue is a rare but known complication of dermatomyositis (DM), mostly associated with juvenile DM and rare in adult DM. Bone scan with Tc-99m Methylene diphosphonate is useful in disease mapping and has high sensitivity to know the extent of calcinosis. However, there is scanty literature available on the utility of bone scan in treatment response evaluation in DM. Rituximab has been found useful in adult DM with calcinosis unresponsive to conventional treatment. We describe an interesting case showing partial response to rituximab on bone scan with single-photon emission tomography-computed tomography.

Keywords: *Cutaneous calcinosis, dermatomyositis, response, rituximab, scintigraphy, single-photon emission tomography-computed tomography, Tc-99 m methylene diphosphonates*

P. Sai Sradha Patro, Subhabrata Patra[1], Rajesh Kumar[2], Kanhaiyalal Agrawal, Girish Kumar Parida

Departments of Nuclear Medicine and [1]General Medicine, All India Institute of Medical Sciences, Bhubaneswar, [2]Department of General Medicine, All India Institute of Medical Sciences, Deogarh, Jharkhand, India

Introduction

Dermatomyositis (DM) is an inflammatory myopathy of autoimmune etiology. Calcinosis of skin, subcutaneous tissue, muscles, tendons, etc., due to tissue injury is a rare but known delayed complication of DM. Dystrophic calcification due to tissue injury secondary to inflammation is the likely cause. It is more common in juvenile onset than adult-onset DM.[1,2] It is commonly visualized at the site of repeated/chronic mechanical stress or trauma. It can be painless or painful with associated movement restriction. Hence, its early diagnosis and treatment is essential. Whole-body bone scan with bone-seeking radiotracers such as Tc-99m methylene diphosphonates (MDP) is more sensitive than planar regional radiographs in mapping the disease extent. Although not studied extensively, bone scan could potentially be useful in evaluating treatment response. Addition of single-photon emission tomography-computed tomography (SPECT-CT) with planar bone scan can add more detailed information regarding the extent of calcinosis in the subcutaneous tissue, muscles, and tendons. This case shows the usefulness of Tc-99m MDP bone scan with SPECT-CT in response evaluation.

Case Report

A 28-year-old male presented with polyarthritis involving large and small joints, with proximal muscle weakness, pain in bilateral lower limbs, and restricted movement around hip joints for 1 year which was gradually progressive. He had a few hyperpigmented lesions on the face and ear. The antinuclear antibody titer was strongly positive, creatine phosphokinase (CPK) level was 196.8 U/L (normal range: 24–195 U/L), and erythrocyte sedimentation rate (ESR) 60 mm/Hr (normal range: 0–20 mm/Hr). Electromyography was consistent with proximal myopathy. He underwent muscle biopsy, and findings were consistent with DM. After confirmation of diagnosis, the patient was treated with methotrexate, steroids, hydroxychloroquine, and sulfasalazine. Even after treatment for about one and a half years, there was no significant response clinically, and the patient developed few hard subcutaneous swellings around the elbows and hip region. X-ray of elbow joint showed subcutaneous calcification suggestive of calcinosis cutis. Further, no clinical response was seen with diltiazem. Tc-99m MDP whole-body bone scan to look for the extent of cutaneous calcification showed multifocal heterogeneous increased tracer

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Address for correspondence:
Dr. Kanhaiyalal Agrawal, Department of Nuclear Medicine, All India Institute of Medical Sciences, Bhubaneswar, Odisha, India.
E-mail: nucmed_kanhaiyalal@aiimsbhubaneswar.edu.in

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uptake in the soft tissues of the left cheek, bilateral arms, elbows, left forearm, posterior thorax, pelvis, bilateral thighs, and left feet [Figure 1a-1b]. The patient was given 3 doses of intravenous 1 g rituximab as an initial dose, again at 1 and 6 months along with methotrexate and hydroxychloroquine. Follow-up whole-body bone scan after approximately 8 months showed regression in the intensity of tracer uptake in a few preexisting lesions with no new lesions [Figure 1c-1d]. Partial response to treatment was further better characterized by baseline and follow-up SPECT-CT of gluteal region [Figure 2]. There was both clinical and biochemical response (ESR-40 mm/Hr and CPK-73U/L) correlating imaging findings.

Discussion

DM is a rare autoimmune disease characterized by inflammation in various tissues including muscles and skin. Calcinosis cutis is dystrophic calcification in the dermis due to tissue injury secondary to inflammation and occur in the setting of normal serum calcium and phosphorus levels. It is more common in juvenile DM (20%–40% cases) than adult-onset DM (around 20%) and is seen late in the course of disease.[1,2] It can be painless or can be painful if associated with complications such as ulceration, infection, and nerve compression. Hence, its diagnosis and treatment are crucial. The mechanism of Tc-99m MDP uptake in bone scintigraphy is mostly due to adsorption into hydroxyapatite crystals in the soft tissues similar to that in bones.[3] Bone scan has higher sensitivity compared to plain radiograph to assess the extent of the extraosseous calcinosis.[3-6] Rituximab has been found useful in unresponsive calcinosis to conventional therapies in connective tissue disorders.[7] Bone scan can be useful in objective monitoring of treatment response but has not been studied extensively in this condition.[8] In this particular case, interval bone scan with SPECT-CT showed partial response after treatment and hence is useful for response evaluation. Since treatment response is slow in DM, interval bone scan should be delayed 2–3 months after treatment to better characterize the response.

Patient consent

The patient consent was obtained.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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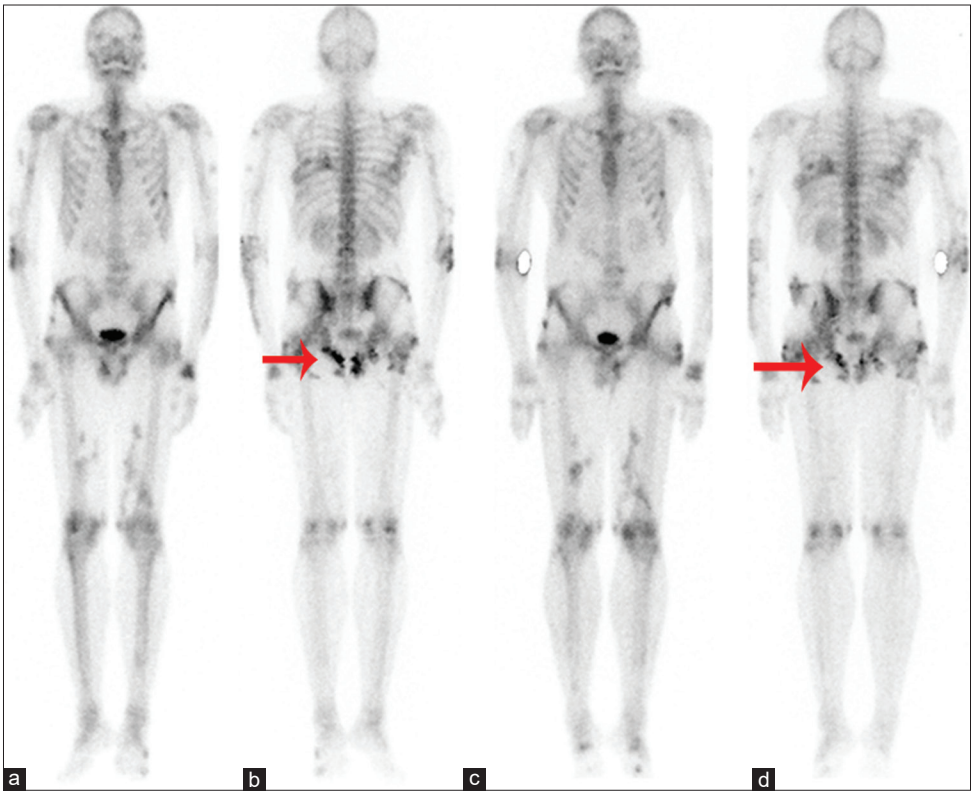

Figure 1: (a and b) Initial whole-body bone scan shows multifocal heterogeneous increased tracer uptake in the soft tissues of the left cheek, bilateral arms, elbows, left forearm, posterior thorax, pelvis, bilateral thighs, and left feet. (c and d) Follow-up scan shows regression in the intensity of tracer uptake in a few preexisting lesions (arrows) and no new lesions

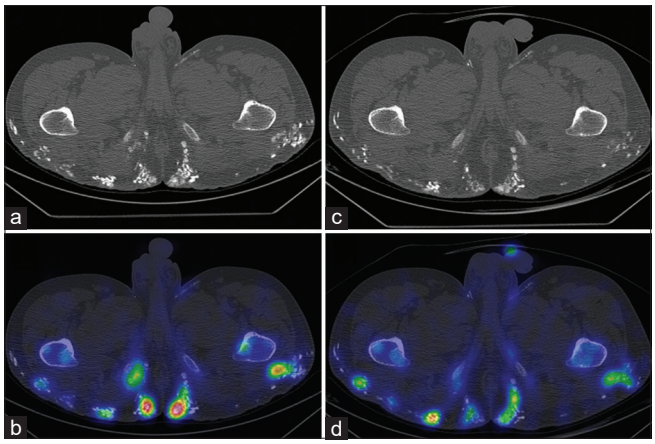

Figure 2: (a and b) Baseline single-photon emission tomography-computed tomography images shows tracer uptake in the multifocal calcifications in the subcutaneous fat of the gluteal region. (c and d) Follow-up single-photon emission tomography-computed tomography images show regression in the intensity of tracer uptake and minimal regression in the extent of calcification suggestive of partial response to treatment

Conflicts of interest

There are no conflicts of interest.

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