Case Report

Tc-99m MDP bone scintigraphy in a case of Touraine-Solente-Gole syndrome

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ABSTRACT

Pachydermoperiostosis is a form of primary hypertrophic osteoarthropathy, also known as Touraine-Solente-Gole syndrome. It is a rare disease. In this report, we present the case of a 29-year-old man with this rare disorder, having significant findings on Tc-99m methylene diphosphonate bone scan.

Keywords: Methylene diphosphonate, pachydermoperiostosis, Tc-99m, Touraine-Solente-Gole syndrome, primary hypertrophic osteoarthropathy

INTRODUCTION

Pachydermoperiostosis (Touraine-Solente-Gole syndrome) is a rare entity characterized by digital clubbing, periostitis and thickening of the skin of face, scalp and extremities with associated hyperhidrosis. The disease has bimodal presentation, one at the age of 1 year and the other at puberty. The disease has male preponderance.[1] It has an insidious clinical course and needs to be differentiated from other causes of hypertrophic periostitis as well as thyroid acropachy by clinical as well as imaging correlation.

CASE REPORT

We present an interesting case of a 29-year-old male, born of a non-consanguineous marriage, who complained of predominantly lower limb joint pain and swelling. There was a history of profuse acne over face and upper chest for last 12 years. Clubbing also developed during this period. The patient was earlier diagnosed to be a case of seronegative rheumatoid arthritis and was subjected to leflunomide and analgesics. There was no history of cough, fever, dyspnea or weight loss. There was no family history of clubbing or arthritis. On examination, grade IV clubbing was present. Painful swelling was present in bilateral knee and ankle joints. The pain got relieved after administration of analgesics. Acne was present all over the face and chest. It was associated with skin thickening on the face. No swelling was seen in the joints of the hand. USG of the knee joint showed the presence of increased synovial fluid and synovial hypertrophy. The chest X-ray was normal. The digital skiagram of both hands AP view showed the presence of shaggy periosteal reaction in the distal radius on both sides, also involving parts of the epiphysis [Figure 1]. Similar shaggy periosteal reaction was noted in the 1st and 5th metacarpal diaphysis on both sides. There was increased thickening of the cortical bone in the region of diaphysis of all metacarpal bones, proximal and middle phalanges [Figure 2]. Similar periosteal reaction involving the tibia and fibula on both sides was noted [Figure 3]. On further investigations, his erythrocyte sedimentation rate (ESR) was 8 mm in 1st hour, and hemoglobin, leukocyte and blood counts were normal. HLA B-27 and rheumatoid factor were negative.

Skeletal scintigraphy was done after the administration of 740 MBq/20 mCi of Tc-99m Methylene diphosphonate. It showed the presence of pericortical linear uptake along the distal ends of tibiae, fibulae, ulnae and radii [Figure 4]. There was increased radiotracer concentration in the terminal phalanges of feet, metacarpophalyngeal, interphalyngeal joints and phalanges of both hands [Figures 5 and 6]. The axial skeleton appeared normal [Figure 7].

DISCUSSION

Pachydermoperiostosis was first described by Nikoleus Friedrich in 1868. It is a rare disorder most commonly affecting adolescent males, accounting for 5% of all the cases of hypertrophic osteoarthropathy. It is a familial autosomal dominant disease, but can be autosomal recessive or idiopathic.[2,3] The disease is epitomized by extensive clubbing of fingers, hyperhidrosis of thickened skin, arthralgia/arthritis and acne.[4] The disease is sometimes self-limiting.

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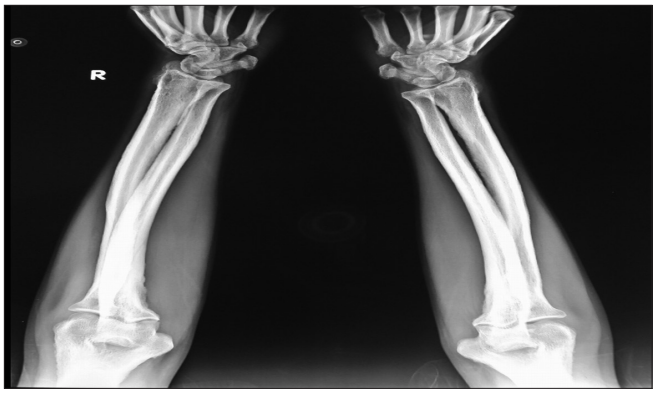

Figure 1: Radiograph of both forearms AP view shows linear periosteal reaction

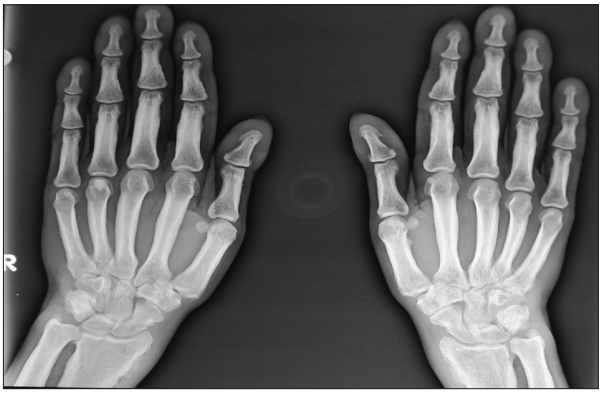

Figure 2: Radiograph of both hands AP view shows shaggy periosteal reaction in the distal radius bilaterally involving the epiphysis. Increased cortical thickness in the region of the diaphysis of the metacarpals, proximal and middle phalanges

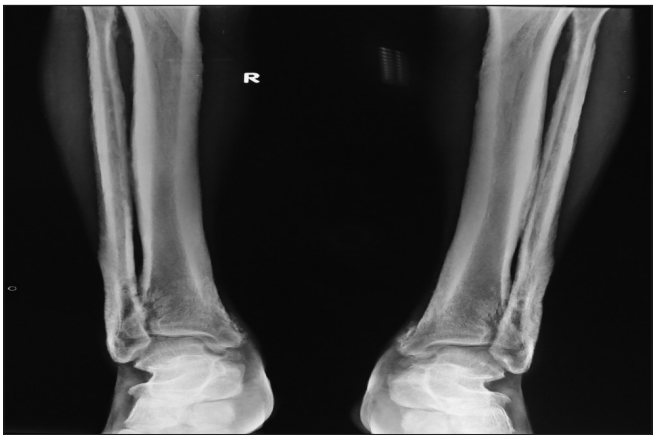

Figure 3: Radiograph of both legs shows periosteal reaction involving tibia and fibula on both sides, extending to involve the epiphysis

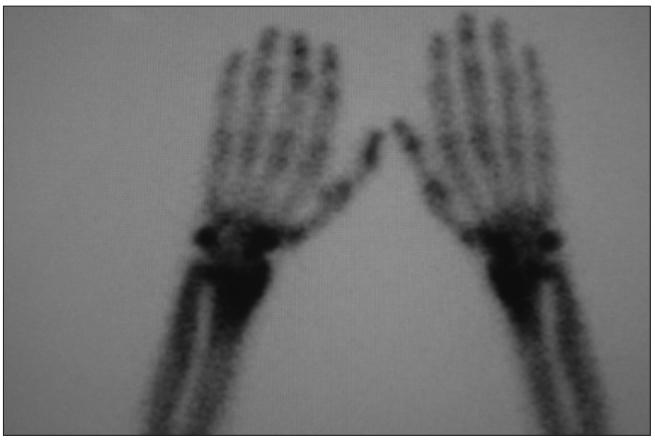

Figure 4: Posterior view of distal forearm and hands shows increased uptake in the phalanges, distal ends of metacarpals, radius and ulna

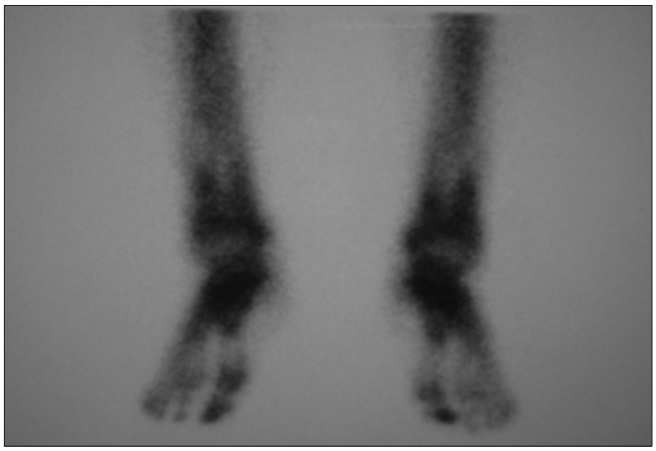

Figure 5: Anterior view of feet and lower limb on a Tc-99m MDP bone scan shows pericortical uptake in the distal ends of tibia and fibula. Prominent uptake in distal ends of 1st metatarsal and phalanges of both feet is also seen

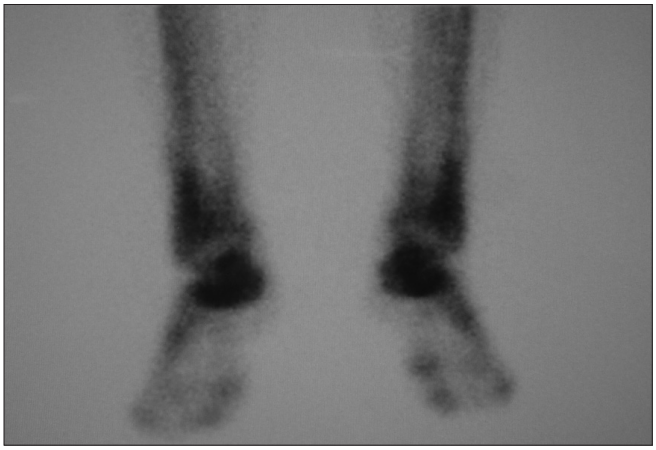

Figure 6: Posterior view of both lower limb and feet showing increased uptake at the distal ends of tibia and femur bilaterally with increased pericortical uptake at distal ends

Tc-99m MDP bone scan is a very sensitive investigation for detecting primary hypertrophic osteoarthropathy. Scintigraphic findings reveal symmetrical increased pericortical linear uptake of the tracer along the distal ends of long bones of upper and lower extremities, namely, radius, ulna, tibia and fibula. These findings are more marked in the skeletal phase on a triphasic study in the individuals with active disease.

Moreover, the uptake may also be seen in the small tubular bones of the hands and feet. This is attributed to the vasodilatation in the tips of fingers, excessive growth of cellular tissue in the clubbed fingers and the duration of disease. A complex interplay of these factors has been suggested for increased activity of the radiotracer in the phalanges.[5]

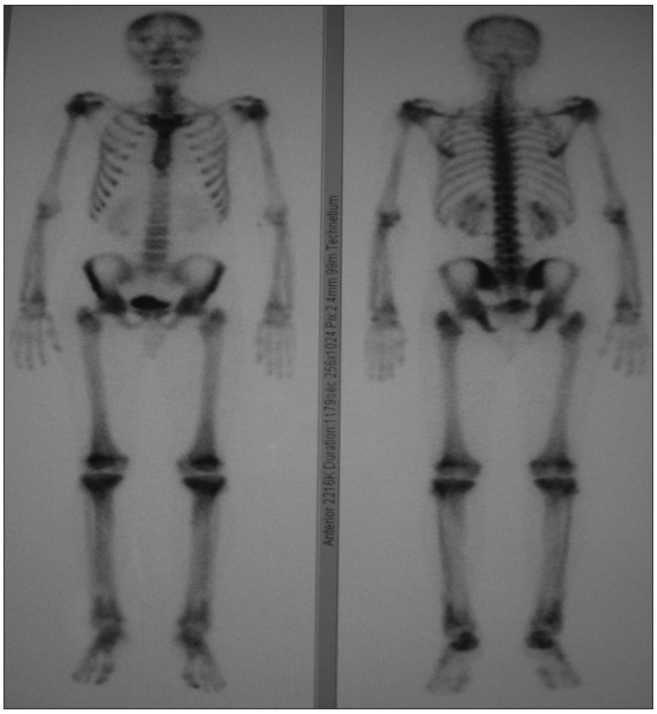

Figure 7: Whole body bone scan with normal chest anterior and posterior view and normal axial skeleton

The disease needs to be differentiated from other causes of hypertrophic osteoarthropathy and thyroid acropachy, given its benign clinical course. Scintigraphically, pachydermoperiostosis demonstrates a higher uptake of Tc-99m MDP, especially at the ends of tibia, fibula, radius and ulna compared to secondary hypertrophic osteoarthropathy. A higher diaphyseal uptake in hands and feet is noted in thyroid acropachy with rare involvement of long bones on bone scan.

CONCLUSION

To conclude, a diagnosis of pachydermoperiostosis must be considered in a patient presenting with joint pains and symmetrical uptake of the tracer along the ends of long bones of legs and forearms on bone scintigraphy, once other causes of hypertrophic osteoarthropathy have been excluded.

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