Pachydermoperiostosis: Technetium-99m-Methylene Diphosphonate Scintigraphic Pattern

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We present two cases of pachydermoperiostosis. Both patients presented with joint pains and swelling. Bone scan revealed marked pericortical uptake involving the distal end of long bones. The distribution pattern and differential diagnosis of the scan abnormalities are discussed.

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Primary hypertrophic osteoarthropathy (*1*) or pachydermoperiostosis (Touraine Solente Gole syndrome) is a rare disorder, which usually begins insidiously during puberty and is characterized by digital clubbing, thickening of the skin of the face, scalp and extremities along with sebaceous gland overactivity. The disorder is inherited as an autosomal dominant with variable expression (*2*).

Reported here are two of these rare cases with their scintigraphic distribution patterns seen on routine bone scans.

CASE REPORTS

Case 1

A 28-yr-old black man presented with a 5-yr history of joint pains. Initially, the pain started in the toes and ankles, which were swollen with an associated early morning stiffness usually occurring four to five times per week. Involvement of knee, elbows, wrists and cervical joints followed. He had noticed thickening of his fingers over the past 10 yr. He denied symptoms that would suggest chest or abdominal illness. There was no significant family history.

Pertinent clinical findings included an oily skin, large hands and feet with marked clubbing. His grip was firm, which virtually excluded any significant arthritis in the hands. He had restricted movement of ankles and knees with effusions in both joints. Examination of the other systems was unremarkable.

Investigations revealed erythrocyte sedimentation rate of 10 mm/hr, hemoglobin, leukocyte, and platelet count were normal. Serum calcium, phosphorous, alkaline phosphatase, uric acid, thyroid indices and urine analysis were normal. Tests for rheumatoid and antinuclear factor were normal. Radiographic examination showed irregular periosteal proliferation and cortical thickening of distal ends of tibiae, fibulae (Fig. 1A), ulnae, and radii. X-rays of other areas were normal. Bone scan performed with 20 mCi of ^{99m}Tc-methylene diphosphonate (MDP) showed increased pericortical linear uptake especially along the distal ends of tibiae, fibulae (Fig. 1C), ulnae and radii. Increased uptake was also noted in the terminal phalanges of both feet (Fig. 1D), metacarpophalangeal, and interphalangeal joints and phalanges of both hands.

Case 2

An 18-yr-old Asian man presented with a 3-mo history of pain and swelling of his ankles and knees, initially involving left ankle, followed by the right ankle and subsequently the right knee. He also gave a history of pain in hips, some stiffness of lower back, but denied early morning stiffness. Recently, he also noted some change in the shape of his nails and had developed a skin rash over the dorsum of the fingers of both hands. Other symptoms to suggest chest or abdominal disease were absent. His father had severe psoriasis complicated by arthritis, otherwise family history was unremarkable.

Relevant findings included an oily skin, marked finger and toe clubbing and dystrophic nail changes affecting the fingers and toes. There was a dry eczematous rash over the dorsal surface of the left metacarpophalangeal joint. There was mild bilateral soft-tissue swelling with pain at the wrists. Both knee joints were warm with small effusions and tender to palpation and some tenderness of the left heel was also observed. There was full range of movement of all joints. Other joints were normal. Examination of other systems was unremarkable.

On investigation his erythrocyte sedimentation rate was 68 mm/hr. Hemoglobin, blood count, serum calcium, phosphorous, alkaline phosphate, protein electrophoresis, thyroid indices, uric acid, creatinine and urine analysis were normal. Tests for rheumatoid and antinuclear factor were negative. Skin and nail scrapings revealed Trichophyton rubrum. X-rays of the knee and ankles revealed generalized modeling deformity with widening of the diaphysis and metaphysis of the long bones, with evidence of periosteal reaction on the medial and lateral aspect of the tibia. X-rays of other sites were unremarkable. A routine bone scan revealed prominent uptake along the periosteal surface especially of the distal end of tibia, fibula, ulna and radii (Fig. 2). A needle synovial biopsy of the left knee revealed mild hyperplasia of the synovial cell lining and normal vessels with some adipose tissue.

DISCUSSION

Pachydermoperiostosis (PDP) is an uncommon disease. The onset of PDP coincides with puberty and the clinical

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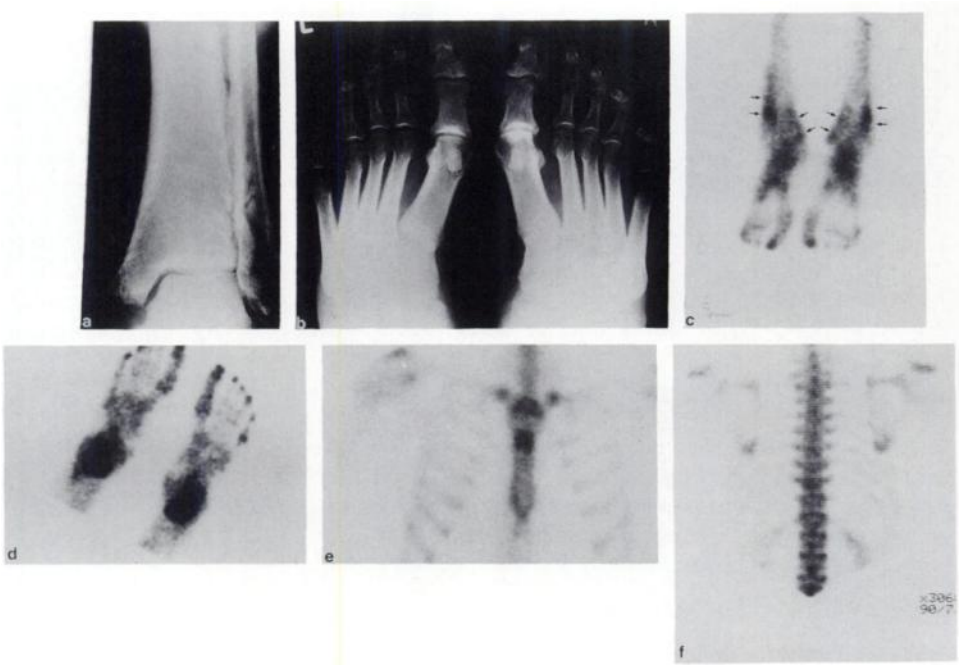

FIGURE 1. Case 1. (A) Radiographs show irregular periosteal new bone formation on tibia and fibula. (B) Distal phalanges appear normal. (C) Anterior view of feet (digital image). Arrows indicate pericortical uptake at the distal end of tibiae and fibulae. (D) Plantar view of feet (digital image). Prominent uptake is seen in the distal end of first metatarsal and phalangeal bones of both feet. In addition uptake is also seen in the distal phalanges. (E) Anterior view of the chest. (F) Posterior view of chest.

findings remain stable after the second decade, apparently unassociated with any morbid consequences (*2,3*). In this study, both cases presented with joint pains and swelling, one early (Case 2) and the other late (Case 1) in the clinical onset of this disease.

Skeletal scintigraphic features of PDP have been mentioned previously (*4–6*). However, a whole-body image was shown only in one case (*4*). The other studies illustrated either a blood-pool image of the hands (*5*) or a rectilinear scanner image of knee joints (*6*). In comparison, the bone uptake seen in this study was much more prominent. Bone scintigraphy in both cases consistently revealed symmetrical increased pericortical linear deposition of the tracer especially along the distal ends of tibia, fibulae, ulna and radii. In addition, increased transverse diameter of the long bones especially at the distal ends was also noted (Fig. 1C; Fig. 2). One of the explanations given for this pericortical uptake was increased blood flow during the active stage of disease and decreased flow during the quiescent stage (*4,5,7*). In this study, both cases were imaged during the active stage of the disease. However, flow could not be assessed, since prior knowledge of the clinical condition was not known. The difference between these two cases was the comparatively high pericortical uptake in long bones in Case 1. It is likely that this also reflects the duration of disease on the uptake pattern, in addition to blood flow.

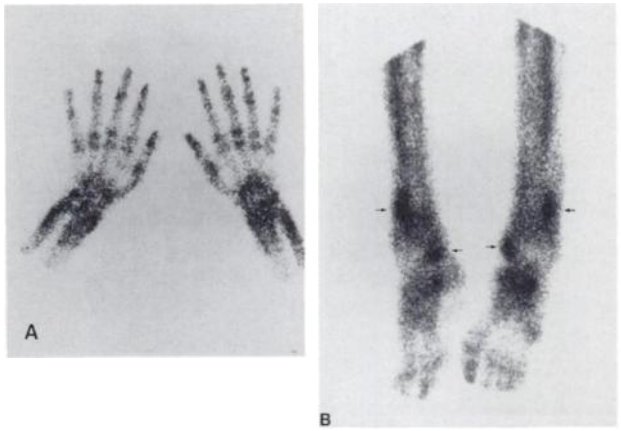

FIGURE 2. Case 2. (A) Palmar views (analogue image). Increased uptake is noted in the phalanges and distal end of radius and ulna. (B) Anterior view of feet arrows Indicate pericortical uptake at the distal end of tibiae and fibulae.

Another interesting feature was the high uptake in the terminal phalanges of both feet in Case 1 compared to Case 2. This was seen despite the fact that marked clubbing was noted in both patients. However, the phalangeal uptake of the hands in each case was similar. This raises the contentious issue of whether uptake in the clubbed extremities was due to vasodilatation in the tips of the digits, excessive growth of cellular tissue in the nail bed (*8*), duration of the disease, or a combination of these factors.

Because of the benign course of PDP, it is important to distinguish it on scintigraphy from diseases such as secondary hypertrophic osteoarthropathy (SHOA) and thyroid acropachy, although, clinically this is not a problem. Comparison of bone scans performed in patients with SHOA showed that uptake was higher in PDP especially at the distal ends of tibiae, fibulae, radii and ulnae. The distribution pattern was similar elsewhere in the axial skeleton. In thyroid acropachy, the uptake was more in the diaphyseal region usually confined to the hands and wrists and rarely involving long bones (*9,10*).

In conclusion, it is felt that in patients presenting with joint pains with symmetrical increased uptake along the

distal end of long bones on bone scintigraphy, the diagnosis of primary pachydermoperiostosis should be considered once other causes of hypertrophic osteoarthropathy have been eliminated.

ACKNOWLEDGMENT

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SELF-STUDY TEST

Skeletal Nuclear Medicine

Questions are taken from the *Nuclear Medicine Self-Study Program I*, published by The Society of Nuclear Medicine

DIRECTIONS

The following items consist of a heading followed by numbered options related to that heading. Select the options that you think are true and those that you think are false. Answers may be found on page 1914.

True statements regarding prostatic carcinoma include which of the following?

1. Osseous metastatic disease is manifested by bone pain in nearly 90% of patients.
2. Elevation of the serum acid phosphatase level indicates metastatic disease to bone.
3. The finding of new areas of increased uptake on serial bone scintigrams has little prognostic importance in patients with known osseous metastases.
4. Radiography and bone scintigraphy are equally sensitive in demonstrating improvement in metastatic disease following therapy.
5. Bone scintigraphy often demonstrates progression of disease in advance of changes in the serum acid phosphatase level.

True statements regarding osteosarcoma include which of the following?

6. The peak incidence is in the second decade of life.
7. It is the most common malignant primary bone tumor.
8. In elderly patients, most osteosarcomas arise either in sites of Paget's disease or after radiation therapy.
9. For radiation-induced osteosarcoma, the average latent interval between irradiation and diagnosis of the tumor is 15 yr.
10. Osseous metastases rarely, if ever, occur before pulmonary metastases.

True statements regarding Ewing's sarcoma include which of the following?

11. Osseous metastases at presentation occur in less than 5% of patients.
12. Most occur in patients between the age of 5 and 20 yr.
13. It most commonly arises in the long bones of the extremities.
14. Pain and swelling are uncommon as presenting symptoms.
15. The diagnosis can be made reliably by radiographic findings alone.

True statements regarding Paget's disease include which of the following?

16. The prevalence varies with geographic location.
17. The tibia is the most frequent site of involvement.
18. A common site of sarcomatous degeneration is the humerus.
19. The most common complication of skull involvement is trigeminal neuralgia.
20. Affected bones are frequently enlarged.
21. The most common tumor complicating Paget's disease is fibrosarcoma.

True statements regarding solitary abnormalities detected by bone scintigraphy in patients with known cancer include which of the following?

22. They should be considered benign when radiographs are normal.
23. The probability of metastasis varies with location of the lesion.
24. Photon-deficient lesions are most often due to metastases.
25. Those in ribs are due to metastasis in more than 50% of cases.
26. They rarely are due to trauma.