Net Letter

Incomplete pachydermoperiostosis

Sir,

Pachydermoperiostosis or primary hypertrophic osteoarthropathy is an uncommon disorder, with a presumably autosomal dominant inheritance.[1] It is characterized by pachyderma, periostosis, arthralgia and finger clubbing. Variations in the phenotypic expression results in clinical features ranging from mild to severe; and in a given case the skin, soft tissues, and bones may be involved independently or together. We report a rare form of pachydermoperiostosis without cutaneous involvement.

A 30-year-old, unmarried male, born of non consanguineous marriage presented to us with a 15-year history of painless swelling of fingers, wrists, elbows, toes, ankles and knees associated with restriction of movements. Initially he noticed swelling around the ankles, which gradually progressed to involve fingers, toes, wrist and elbows over a period of another ten years. The swelling has not increased since five years. There was no history of arthralgia, fever, weight loss, burning micturition, genital discharge, genital ulceration, bleeding tendency, cough, breathlessness, oral ulceration, hyperhidrosis, recurrent epigastric pain or photosensitivity. Patient denied history of sexual exposure. All the family members were examined. None of them had pachyderma, arthralgia or finger clubbing. General physical examination and systemic examination were un-remarkable. Skeletal examination revealed "spade like" hands and feet and column like forearms and legs [Figure 1]. All the finger and toe nails were clubbed. Detailed cutaneous examination did not show any abnormality including cutis verticis gyrata, seborrhea, eyelid ptosis, hyperhidrosis of palms and soles or thickening of skin. Based on the history and clinical examination we considered the differential diagnosis of pachydermoperiostosis, acromegaly, hypothyroidism and secondary hypertrophic osteoarthropathy.

On investigation, fasting and post-prandial blood sugar levels, serum sodium, potassium, calcium, phosphate and alkaline phosphatase were normal. Growth hormone, T3, T4 and TSH were within normal limits. Both VDRL and TPHA tests were negative. Skin biopsy from 2 different sites did not reveal any abnormality. X-ray of long bones, metatarsals, metacarpals and phalanges showed periostosis, subperiosteal ossification and soft tissue swelling at the distal ends [Figure 2]. X-ray skull, lateral view showed a normal sella turcica. X-ray chest was normal.

Hypertrophic osteoarthropathy is divided into primary and secondary forms. Pachydermoperiostosis (PDP), the primary form, accounts for 5% of all cases

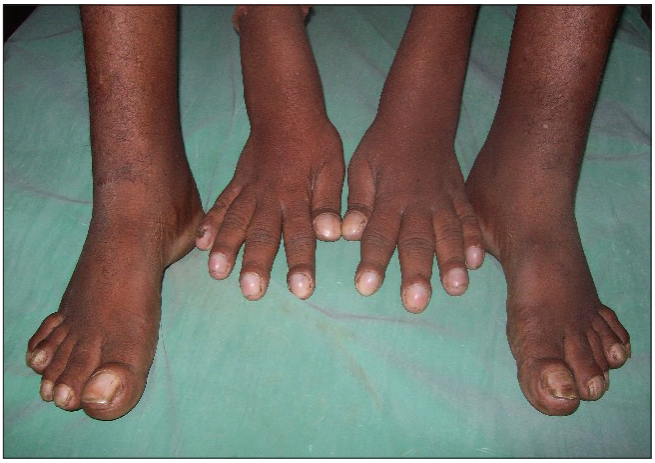

Figure 1: Column like forearms and legs

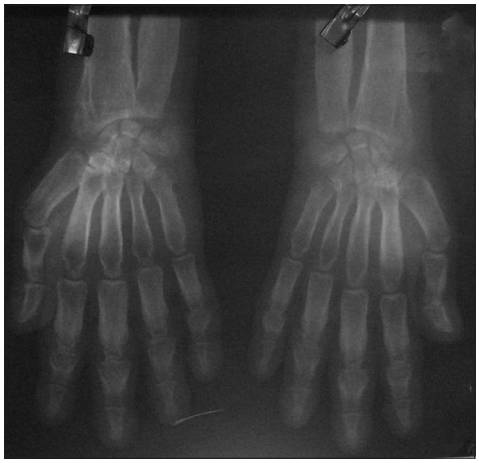

Figure 2: Periostosis of metacarpals

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of hypertrophic osteoarthropathy.[2] It usually begins soon after puberty. Skin and bone changes become progressively more severe for 5-10 years and then remain unchanged throughout life.

Three forms of pachydermoperiostosis or primary hypertrophic osteoarthropathy have been described. They are: (a) Complete form with pachydermia and periostosis, (b) Incomplete form with evidence of bone abnormalities but lacking pachydermia, and (c) Forme fruste with prominent pachydermia and minimal-to-absent skeletal changes.[3] Secondary hypertrophic osteoarthropathy is associated with underlying cardiopulmonary diseases and malignancies. The disease progresses more rapidly than primary hypertrophic osteoarthropathy.[4] It occurs predominantly in men aged 30-70 years. Thus the primary and secondary forms of pachydermoperiostosis must be differentiated by the age of onset, the rate of progression and the presence of a pulmonary lesion. [5] Our case had an incomplete form of PDP with evidence of bone abnormality in the form of cylindrical appearance of upper and lower limbs as well as periostosis on X-ray in absence of skin changes and underlying malignancy. In one series, four cases of milder forms of PDP were reported. However there was absence of skin changes and periostosis in all the four cases.[6] Another study reported familial incomplete form of PDP.[7]

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REFERENCES

1. Burton JL, Rook A. Genetics in Dermatology. In: Rook A, Wilkinson DS, Ebling FJC, editors. Textbook of Dermatology. 4th ed. Bombay: Oxford University Press; 1987. p. 105-66.
2. Lowenthal MN, Tombak A, Lowenthal A. Secondary hypertrophic osteoarthropathy (HOA) mimicking primary HOA (pachydermoperiostitis or Touraine-Solente-Golé) syndrome. Isr Med Assoc J 2004;6:64.
3. Touraine A, Solente G, Gole L. Un syndrome osteodermopathique: la pachydermie plicaturee avec pachyperiostose ds extremites. Presse Med 1935;43:1820-4.
4. Vogl A, Blumenfeld S, Gutner LB. Diagnostic significance of pulmonary hypertrophic osteoarthropathy. Am J Med 1955;18:51-65.
5. Harper JI, Trembath RC. Genetics and Genodermatoses. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. Rook's Textbook of Dermatology.7th ed. London: Blackwell Science; 2004. p.12.72-3.
6. Girisha KM, Mandal K, Phadke SR. Milder form of pachydermoperiostosis: a report of four cases. Clin Dysmorphol 2009;18:85-9.
7. Sethuraman G, Malhotra AK, Khaitan BK, Sharma VK, Kumar R, Makharia GK, *et al*. Familial pachydermoperiostosis in association with protein-losing enteropathy. Clin Exp Dermatol 2006;31:531-4.