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A 29-year-old male presented with thickening of skin of scalp, forehead and face along with clubbing of the fingers. He was diagnosed to have primary pachydermoperiostosis and is being reported in view of rarity of this entity.

Key words : Pachydermoperiostosis

Pachydermoperiostosis is a clinical syndrome characterized by thickening of skin of scalp forehead, face, hands and feet. The skin of the scalp, is thrown into folds and is referred to as cutis verticis gyrata. Thickening of the phalanges with finger clubbing is a usual accompaniment. Pachydermoperiostosis occurs in two forms.[1]

i. Primary pachydermoperiostosis (Touraine-Solente Gole' syndrome) which is a developmental disorder restricted predominantly to males and occurs usually in an autosomal dominant fashion.[2,3]

ii Secondary pachydermoperiostosis which occurs commonly in men to seventh decade. The osseous changes tend to develop rapidly and are often associated with bronchogenic carcinoma pleuralmesothelioma, lung abscess, bronchiectasis, gut malignancy or carcinoma of thymus. Very rarely, congenital cyanotic heart disease may be a predisposing factor for this entity.[1,4]

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Case Report

A 29-year-old male, presented with history of thickening of skin of the scalp, forehead, face, hands and feet since the age of 12. Initially, these skin changes were progressive for about a decade and then remained stationary. There was no history suggestive of hyperhidrosis or involvement of gut or respiratory system and cardiovascular system. Family history was also unremarkable. Cutaneous examination revealed thickening and folding of skin of scalp, forehead and face (Fig.1). Thickening of skin of hands and feet was also noted. Palmoplantar thickening was quite marked. Besides afore mentioned features, patient also had grade IV clubbing of fingers and toes. Neuropsychiatric evaluation revealed a subnormal intelligence quotient of 65. Rest of the systemic examination was noncontributory.

Plain radiograph of the skull showed thickened soft issue which was seen in vertical columns with a periosteal reaction in pari

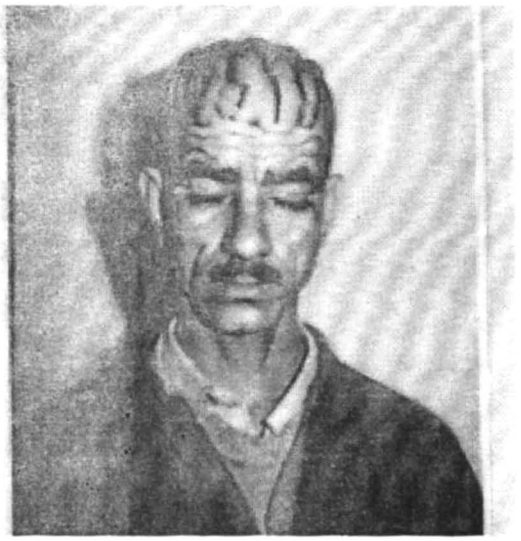

Fig. 1 . Cutis verticis gyrata due to pachydermoperiostosis.

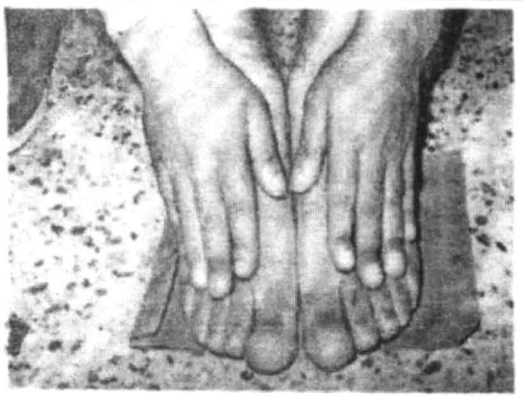

Fig. 2 . Note clubbing and thickening of fingers and toes (acropachy)

etal bones. Radiographs of both forearms showed periosteal reaction medially in lower metaphysis and lower third of diaphysis. Radiographically, similar changes were seen in the tibia and also in the proximal and middle phalanges of left middle and ring finger and very early periosteal reaction in proximal phalanges of middle and ring finger. Chest skiagram was unremarkable. Moreover, clinical and radiological examination of his parents and other four siblings did not add any further information.

Discussion

Primary pachydermoperiostosis, as already mentioned is a rare developmental disorder characterized by cutaneous changes which usually start around puberty, progresses for a few years and then remains static. Such patients may be mentally retarded. The patient under discussion was diagnosed to have primary pachydermoperiostosis in view of his age of onset of symptomatology, unremarkable systemic examination and chest skiagram. In view of his negative family history our patient seems to represent sporadic form of primary pachydermoperiostosis. Mental retardation, a well known accompaniment of primary pachydermoperiostosis was also detected in our patient as was evident form his low intelligence quotient.

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