CAMPTODACTYLY WITH KNUCKLE PAD

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A 23-year-old male had camptodactyly affecting the little fingers of both hands and the ring finger of left hand. He had associated knuckle pad on one of the affected little fingers. Elder sister of the patient also had camptodactyly.

Key words : Camptodactyly, Knuckle pad, Association.

Hereditary camptodactyly is a rare fibrodysplastic condition characterized by persistent flexion contractures of the proximal interphalangeal joints of little fingers, while the metacarpophalangeal joints and the palmar fascia remain unaffected. Inheritance is by an autosomal dominant trait and the sexes are equally affected. It has been reported in association with Marfan's syndrome, oculo-dento-digital dysplasia, pectus excavatum, scoliosis, ptosis and taurinuria.[1] We report a case of hereditary camptodactyly in a young male who had an associated knuckle pad on the dorsum of one of the affected little fingers.

Case Report

A 23-year-old male college student, born to non-consanguinous parents, developed persistent flexion contractures of the proximal interphalangeal joints of the little fingers of both hands and ring finger of the left hand (Fig. 1) since early childhood, and a firm, circumscribed, skin-coloured, 3 mm raised, circular, plaque-like lesion of 16 mm diameter on the extensor aspect of the proximal interphalangeal joint of the left little finger since 5 years. He denied history of any preceding local trauma. All the other fingers and toes and their joints were normal. There was no thickening or contracture of the palmar fascia. There was no thickening of peripheral nerves and cutaneous sensations in the hands were normal. His elder sister, aged 30 years was found to have flexion contracture of both the little fingers. But there was no plaque-like lesion on the finger. General physical and systemic examination did not reveal any abnormality.

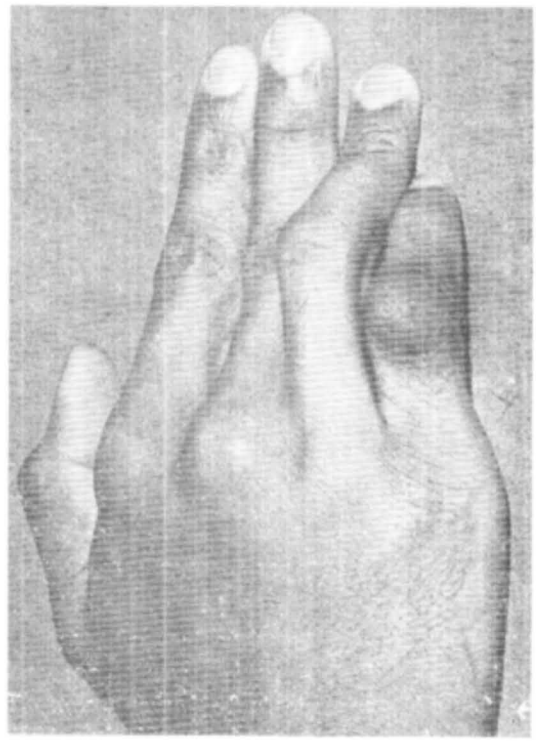

Fig. 1. Camptodactyly affecting the ring finger and the little finger of left hand. Note knuckle pad on the dorsum of little finger.

Results of routine laboratory investigations on blood, urine and stools were normal. X-ray of the hands showed flexion at the interphalangeal joints of the affected fingers. Bone structure and metacarpal index were normal. Histopathological study of the plaque on the dorsum of the

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finger revealed marked hyperkeratosis and acanthosis with thickening and hyperplasia of the dermis.

Comments

Camptodactyly affects mainly the little fingers[2] producing a persistent flexion contracture of the proximal interphalangeal joints. Rarely, other fingers also may be affected.[1] The metacarpophalangeal joints and the palmar fascia which are characteristically affected in Dupuytren's contracture remain spared in camptodactyly. The association of camptodactyly with knuckle pad, another fibrodysplastic condition, in the present case is quite interesting. Knuckle pad has been reported in association with pseudoxanthoma elasticum, leukonychia, deafness, Dupuytren's contracture and palmo plantar keratoderma.[3,4] The association of knuckle pads and camptodactyly also had been mentioned.[5] Two cases of knuckle pads reported recently by Pavithran[6] had no other fibrodysplasias or defects. Camptodactyly must be differentiated from streblodactyly[2,7] which is characterised by flexion deformity at the metacarpophalangeal joint of the thumb and the proximal interphalangeal joints of little fingers. It is seen in females and some fingers may show swan-neck deformity. It is easy to differentiate amptodactyly from claw-hand which is characterized by hyperextension at metacarpophalangeal joints and flexion at interphalangeal joints of the fingers.

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