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Letters to the Editor

HEREDITARY CAMPTODACTYLY: A CONDITION MIMICKING CLAWHAND DUE TO LEPROSY

Sir,

Hereditary camptodactyly is a rare type of fibromatosis with an autosomal dominant inheritance that affects both sexes equally. It begins in childhood, involving the little fingers (bilateral), producing persistent flexion of the proximal interphalangeal joints with sparing of the metacarpophalangeal joint.[1] Sometimes the other fingers are also affected.[2] The resulting deformity closely simulates clawhand[3] caused by leprosy. Because leprosy is a major cause of clawhand in a leprosy-endemic country like India, identification of this entity is essential to prevent the misdiagnosis of cases of 'Camptodactyly' as clawhand due to leprosy. We have recently seen 2 patients suffering from hereditary camptodactyly who were initially suspected as clawhands because of nerve damage due to leprosy.

Both patients (a 16-year-old girl and a 45-year-old man) presented with flexion deformity of the little finger of both hands that they had suffered from birth. The man also had similar deformities of the ring fingers. In both cases there was no history of hypopigmented patch, sensory deficit or neuralgic pain anywhere on the body, including the fingers. The first patient's father and the second patient's mother suffered from similar deformities. Local doctors first suspected both cases to be 'leprosy', and the girl received anti-leprosy treatment for 6 months, without benefit.

Examination revealed fixed flexion deformity of the proximal interphalangeal joint of the little fingers of both hands in the female patient and the little and ring fingers of both hands in the male patient. All modalities of sensations were preserved. There was no wasting or weakness of the small muscles of the hand. In both cases there was no thickening of the ulnar, median or radial cutaneous nerves, and no thickening of the palmar fascia. Systemic examinations and relevant laboratory investigations produced results that were within normal limits in both cases.

Both patients were diagnosed as suffering from 'Camptodactyly' with an autosomal dominant inheritance. Their real condition was explained to them and they were reassured.

Both the above cases were misdiagnosed/suspected as clawhand due to leprosy. The same observation was made by Pavithran.[3] In ulnar clawhand due to leprosy, the deformity consists of hyperextension at the metacarpophalangeal joints and flexion at the interphalangeal joints of the affected fingers. In addition there is a sensory deficit in the distribution of ulnar nerve and wasting and weakness of the small muscles of the hands supplied by the ulnar nerve. However, the 'bilateral' nature of clawing, the absence of any sensory or motor deficit and a family history of flexion contracture is an aid in diagnosing 'Camptodactyly' and helps to rule out leprosy. Camptodactyly is also to be differentiated from Dupuytren's contracture and Sterbiodactyly.[1] Camptodactyly has been associated with Marfan's syndrome, pectus excavatum, scoliosis, ptosis, Dupuytren's contracture, knuckle pad[4] and taurinuria.[5] However, no such associations were observed in our patients.

The idea of this communication was to emphasize two facts. First, 'Camptodactyly' is probably not as rare as is suggested in the standard textbooks because many cases are being diagnosed as 'leprous clawhand', as happened to our patients. Secondly, in an endemic area for leprosy,

physicians, leprologists and field workers must be made aware of this condition in order to avoid the psychological trauma and 'stigma' caused by incorrectly diagnosing patients as 'leprous clawhand'.

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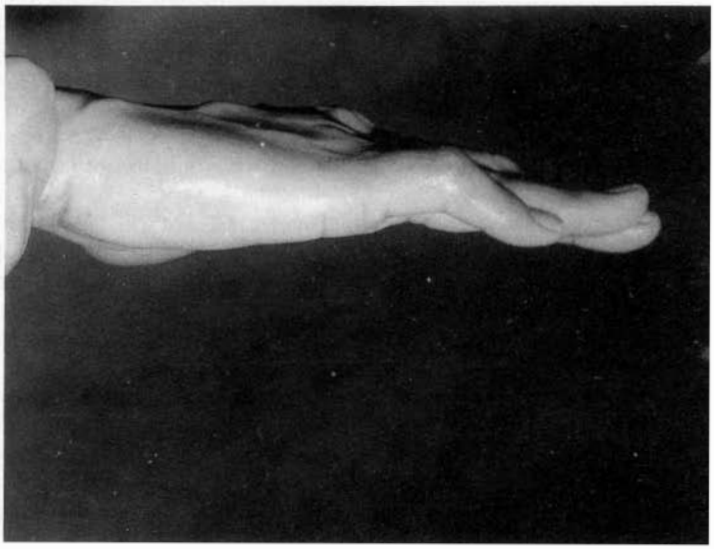

Figure 1. Showing a fixed flexion deformity of the proximal interphalangeal joint of the right hand.