Kirner's Syndrome: Displaced Bone – Misplaced Diagnosis

A 11-year-old boy with a painless but gradually progressive deformity of bilateral little fingers noticed for the past 1 year was referred to us by a pediatrician for a rheumatology consultation. A diagnosis of fractured distal phalanges was offered previously, and a surgical correction was advised. There was no significant family history or previous trauma or infection to his finger. Physical examination showed palmar and radial curving of distal phalanges of bilateral little fingers. Nails of the affected fingers were also curved in volar direction (Fig. 1). Radiological findings showed ventro-radial angulations of terminal phalanx relative to middle phalanx with an apparent overgrowth of epiphysis – a tiny bony spur, which projected distally and fitted into a groove in the basal part of the shaft. Physeal plate appeared widened with sharply narrowed and sclerosed diaphysis. (Fig. 2). A diagnosis of Kirner's syndrome was made and parents were counselled accordingly.

Kirner's deformity or dystelephalangy is an uncommon condition that presents in late childhood or early adolescence (8-14 years) with painless, progressive, bilateral radiovolar curving of the terminal phalanges of the little fingers. It is a clinico-radiological diagnosis. Clinically, Kirner's deformity is characterized by a short and stubby terminal phalanx of the little finger, and deviated in a palmar and radial direction, typically described as 'eagle-claw-like', and a dysmorphic nail.

The deformity needs to be differentiated from other similar deformities such as clinodactyly (radial deviation at the distal interphalangeal joint) and camptodactyly (flexion deformity at the proximal interphalangeal PIP joint). Association has been reported with musculoskeletal (genu valgus, pes cavus, myositis ossificans, absence of flexor digitorum superficialis tendon in the little finger) and cardiovascular abnormalities, and with syndromes such as Turner syndrome, Cornelia de Lange syndrome and Silver syndrome. Treatment modalities recommended are observation, splinting and osteotomy. As the deformity usually ceases after physis closure, reassurance may be sufficient. Temporary splinting may be of help in painful cases. Volar osteotomies leaving an intact dorsal periosteal

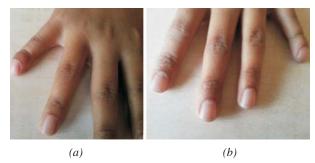


FIG. 1 Symmetrical deformity of tip of both right (a) and left (b) little fingers with shortening of the terminal phalanx, making it stubby and deflected in a palmar-radial direction.



FIG. 2 Radiograph showing ventro-radial angulations of terminal phalanx relative to middle phalanx with an apparent overgrowth of epiphysis.

hinge with K-wire fixation for correction of the deformity has been advocated. Surgery is delayed until physeal closure to prevent recurrence of the deformity.

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