CONTRACTURE OF PROXIMAL INTERPHALANGEAL JOINTS OF THE FINGERS ASSOCIATED WITH PES VALGUS*

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In children flexion contracture of the interphalangeal joints occurs in camptodactyly, Leri’s pleonosteosis, and in arthrogryposis. In the first two conditions the deformity usually makes its appearance in the first 2 years of life but occasionally may be present at birth. In arthrogryposis the deformity is present from birth.

Camptodactyly and Leri’s pleonosteosis are hereditary disorders. Both conditions are the result of the activity of an autosomal gene of variable penetrance and of variable expression.

The patient reported in this paper has some features common both to camptodactyly and to Leri’s pleonosteosis. In addition the patient shows some features which are unique.

CASE REPORT

The patient was an 11-year-old Bantu female. Her father is a member of the Hlubi tribe, while her mother is a Xhosa. She was referred to the Princess Alice Orthopaedic Hospital in Cape Town for assessment of her deformities. Her mother stated that the hand deformities had been present since birth but the foot deformities had become more prominent when the child began to walk at the age of 18 months.

On examination the child was well nourished. She was 148 cm. tall. The abnormalities were confined to the elbows, hands and feet. The skin on the extensor aspect of both elbows was thickened. The skin of the hands was thickened on the palmar aspect, the flexion creases being accentuated and each hand being hollowed out into a cup-like shape. The thenar muscles were poorly developed (Figs. 1 and 2). The patient had 90° flexion contractures of the proximal interphalangeal joints of all 4 fingers in each hand. No active or passive movement was possible at these joints. The distal interphalangeal joints of the fingers and the interphalangeal joint of each thumb were capable of full active movement and of passive hyperextension. The deformities of the hands had not affected function adversely. The patient was able to feed herself, attend to her personal hygiene, to write and to sew.

Examination of the heel revealed a bilateral valgus of the hindfoot (Fig. 3). There was bilateral pes planus and metatarsus abductus. The plantar skin was thickened and the skin creases accentuated (Fig. 4). The child normally walks barefoot. A comparison with the patient’s siblings and cousins, who also walk barefoot, showed that her skin was much thicker and the plantar creases were more prominent. The patient could walk, run and jump without difficulty.

X-rays of the patient’s hands confirmed the flexion contractures. Osteoporosis of the carpus, metacarpals and phalanges was present. There was also a constriction of the neck of the proximal phalanx of the little finger on each side (Fig. 5).

The following laboratory results were obtained: haemoglobin level 12.8 G/100 ml.; blood smear was normal. There was no abnormality on urinalysis or chromosome studies. Calcium excretion in the urine was 23 mg./24 hours.
Fig. 4. Soles of the feet. Note the thickened plantar skin and accentuated plantar creases.

Fig. 5. X-ray of the hands. Note the constriction of the proximal phalanx of the little finger and osteoporosis of the carpal bones.

and phosphate excretion 789 mg./24 hours. Serum albumin was 4.3 G/100 ml., serum globulin 3.9 G/100 ml., and alkaline phosphatase 25 KA units.

A careful examination of 21 family members on the paternal side and of 11 members on the maternal side revealed no clinical abnormality.

DISCUSSION

This patient shows some unique features in addition to features common to camptodactyly and Leri's pleonosteosis.

The most common finding in camptodactyly is a unilateral flexion contracture of the proximal interphalangeal joint of the little finger. Less often the ring, middle and index fingers are involved in descending order of frequency. Spear and Smith and Kaplan attribute this flexion contracture to a shortening of the tendon of flexor digitorum superficialis. It is rare for this flexion contracture to exceed 90°. A mild flexion contracture of the distal interphalangeal joints of the fingers may also be present. McKusick states that if more than one finger is involved there is a striking lack of symmetry on the two sides. In addition to being flexed at the proximal interphalangeal joint the little finger may be rotated at the metacarpophalangeal joint. In that event the palmar surface faces laterally towards the ring finger. Palmar creases are poorly developed and even absent. In their series of cases Currarino and Waldman found patients with club-feet and hammer-toes. A family in Cape Town, studied by Gordon, was also found to have members with severe club-foot deformities, which have proved resistant to treatment. In camptodactyly X-ray of the involved finger shows a constriction of the neck of the proximal phalanx and a volar tilt of the neck of the same phalanx. Subluxation of the proximal interphalangeal joint may also be present.

In 1921, André Leri, a Paris orthopaedic surgeon, described a condition now known as Leri's pleonosteosis. His patients had short stature, mongoloid facies, short spade-like hands, flexion contractures of the proximal and distal interphalangeal joints of the fingers and toes, broad thumbs, and great toes in valgus position. Leri’s patients also had a limitation of movement of the joints of the upper and lower limbs, genu valgum and cubitus valgus. X-ray showed osteoporosis, large epiphyses and thick metaphyses of the bones of the hands. Since Leri’s paper other features have been added. Rukavina et al. described hollow palms, accentuated palmar creases, and a thickening of the skin of the forearms and palms of hands.

Watson-Jones reported a case with bilateral carpal tunnel syndrome and bilateral Morton’s metatarsalgia. He described the histological changes in biopsy specimens taken from the flexor retinaculum of the wrist and from the soft tissues of the foot of his patient. He found greatly increased dense collagen tissue which in parts contained fibrocartilage. Elastic fibres were absent. Mucin was present and mast cells were seen in the specimen from the foot. Leri thought that the condition was due to an excessive and perhaps precocious ossification of epiphyses. Watson-Jones proposed that the deformities were due to capsular contractures and in general the deformity was in the connective tissues. He thought that the bone thickening was due to periosetal traction on the metaphyseal side.

Our case shows some features of both of the above conditions. It also has other features which appear unique. The cutaneous features resemble to some extent those of Leri’s pleonosteosis: the palmar skin was thickened, skin creases were exaggerated and the hand was cupped. Osteoporosis of the bones of the hand was also seen. However, the thickening of the plantar skin, the exaggeration of the plantar skin folds and the poor development of the thenar muscles appear to be features unique to the case reported.

The symmetrical involvement of both hands is a feature of Leri’s pleonosteosis rather than of camptodactyly where asymmetry is the rule. The narrowed neck of the proximal phalanx of the little finger is common both to camptodactyly and to our case. There was, however, no volar tilt of the head or subluxation of the proximal interphalangeal joint of the little finger in the X-ray of the case reported.

Club-foot is a well-known feature of camptodactyly. We are not aware of pes plano-valgus having been described in association with the above condition.

SUMMARY

The main features of camptodactyly and of Leri’s pleonosteosis are reviewed. A case showing unique features and features
common to both Leri's pleonosteosis and camptodactyly is


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