

Congenital Onychodysplasia of the Index Fingers with Anomaly of the Great Toe

Sir,

Congenital onychodysplasia of the index fingers is characterized by various forms of nail dysplasia of the index fingers. The changes are present at birth. Radiological examination of the fingers shows hypoplasia, and narrowing and bifurcation of the affected distal phalanx. In addition to the index fingers, the flanked middle fingers are sometimes involved. We present two cases of congenital onychodysplasia of the index fingers with toe nail deformity. The association of bony polydactyly of the toe with congenital onychodysplasia of the index fingers has not been reported previously.

CASE REPORTS

Case 1.

A 60-year-old woman consulted us with colour change of the toe nails. On examination, the left great toe nail was thickened, whitish-yellow coloured and dystrophic. The left thumb, middle and ring finger nails showed similar changes. The nails were cut and the specimens showed positive for fungus. These changes of the toe and finger nails were due to tinea unguium. Moreover, the left great toe and the nail were broad with a longitudinal groove in the centre of the nail plate (Fig. 1). A rudimentary nail on the ulnar aspect and small indentation on the radial site were present on the right index finger. A small nail was present on the ulnar aspect on the left index finger. The nail of the left middle finger was narrower than the right middle finger nail. She therefore had a deformity of both index and left middle fingers and the left great toe. There was no family history of a similar nature, nor was there any other apparent deformity. Lateral X-ray view revealed a Y-shaped bifurcation of the distal phalanx of the index fingers and a narrowing and spinal projection to the nail site of the distal phalanx of the left middle finger. The left great toe nail showed two small distal phalanges connected with the parietal phalanx (Fig. 2). The anomaly of the fingers is congenital onychodysplasia of the index fingers. The great toe was polydactyly.

Case 2

A 81-year-old man consulted us with an onychodysplasia on his fingers. The onychodysplasia was congenital. His right index finger nail revealed an onychia except for a small indentation (Fig. 3). The left index and the right middle finger nail showed a micronychchia on the ulnar aspect (Fig. 3). The left great toe was short and the toe nail was one-half the normal size and defective. The other finger nails and toe nails appeared normal. Lateral X-ray view revealed a Y-shaped bifur-



Fig. 1. Case 1. Polydactyly of the left great toe. The nail plate was splitting and layering. Its surface is irregular. She suffered from Tinea unguium.

cation of the distal phalanx of both index fingers and hypoplasia, narrowing at the distal third of the distal phalanx of the right middle finger.

COMMENTS

With our present knowledge, the criteria for congenital onychodysplasia of the index fingers are (1): (i) congenital; (ii) unilateral or bilateral index finger involvement; (iii) variability in nail appearance; (iv) possible hereditary involvement; (v) frequent association of bone anomalies. The fifth criterion "frequent associated anomalies" should be revised to reflect changes other than the hypoplasia, narrowing and bifurcation of the affected distal phalanx. The associated anomalies are not rare and the most frequent is the anomaly of the fingers. Reported findings are brachymesophalangy (1–5), syndactyly of the fingers (2–5), ear anomalies (3, 4, 6), great toe anomalies (6), incontinentia pigmenti and syndactyly of the left IV–V and right II–III toes (4), Poland's syndrome (syndactyly of the fingers with absence of the ipsilateral pectoralis major muscle) (4). In family cases reported by Millman & Strier (7) broad hands with shortened digits, tissue thickening of terminal digit, and clinomicrodactyly were noted. In our cases, anomalies of the toes are associated. An association of polydactyly of the great toe has not been reported previously. As associated toe anomaly, Haneke & Kienlein-Kletschka (6) reported one case with a short abnormal bilateral great toe with small toe nail, which was associated with ear anomaly; and onychodysplasia of bilateral thumbs with polyonychia type was found. Neumann et al. (8) reported bilateral micronychchia and hemionychogryposis of great toes with onychodysplasia of all fingers, including the thumbs. Toe nail deformity and polydactyly of the toe as in our cases should be added as associated anomalies.

A genetic abnormality or environmental disturbance in the fetal period is regarded as the cause of the disease. Ischaemia of the affected digits may induce irreversible change in part or whole of the nail buds, resulting in various forms of nail anomaly (1, 4). As a cause of ischaemia, an abnormal grip of the thumbs in utero has been opposed (9). Certain environmental



Fig. 2. Case 1. Two small distal phalanges connected with the parietal phalanx of the left great toe on X-ray.



Fig. 3. Case 2. Anonychia of the right index finger and micronychia of the middle finger.

factors, such as drugs, are suggested as the aetiology (10). Family examples suggest that some cases occur on a hereditary basis, autosomal dominant with variable expressivity (3, 7). Miura & Nakamura (3) suggested that impediments to the membranous ossification centre of the distal phalanx could lead to a dysplastic crescent-shaped cap with nail anomalies. The critical period of syndactyly and brachymesophalangy, the most commonly associated anomalies of congenital onychodysplasia of the index fingers, is earlier than the 10th week of the foetal period which is a time of the development of the primary nail fold. Coexistence of incontinentia pigmenti

or Poland's syndrome supports the genetic occurrence of this nail change. Our findings of coexistence of the anomalies of fingers and toes in congenital onychodysplasia of the index fingers suggest that it is the genetic factor concerning the development of fingers and toes that causes this disease.

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Accepted May 6, 1998.

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