

Familial Leukonychia Totalis

Sir,

The case report of Köhler et al. in a recent issue of this journal (1) deserves some comment. Congenital familial leukonychia totalis is actually a rare condition, but the associated symptoms are not exceptional. In the two brothers whose white nails are described in the Case Report, no other symptoms were observed, but have all the keratinizing adnexal structures been thoroughly examined?

In the family described by Bauer (2), dealing with 31 family members in 3 generations, 17 of the 19 patients with white nails also disclosed pilar cysts. Galadari & Mohsen (3) reported a leukonychia totalis in a man and his sister disclosing a severe keratosis pilaris. In a family indicating a dominant inherited leukonychia, one member of the kindred also had pili torti (4). We observed a family where 11 members in 4 generations had a leukonychia totalis; two of them also had a ciliary dystrophy and one multiple pilar cysts. We proposed to denominate this condition "FLOTCH syndrome" (for Familial LeucOnychia, Trichilemmal cysts, Ciliary dystrophy, autosomal dominantly inherited) (5).

Keratosis palmo-plantaris (6, 7, 9), knuckle pads (6, 7), pilar cysts (8), keratosis pilaris (9), pili torti (9) have also been mentioned in other inherited cases of leukonychia totalis reported in the literature.

I believe that congenital familial leukonychia totalis is usually a dominantly inherited autosomal disease and that the white finger- or toenails are the main symptom of a more complex ectodermal dysplasia involving inconstantly the other keratinizing structures (hair shafts, hair sheaths, eyelashes, stratum corneum of palms and soles) and most probably related to the deficiency of a gene regulating the structure of a hard keratin.

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