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.

REFERENCES

1. Köhler LD, Möhrenschlager M, Ring J. Congenital leukonychia totalis in two brothers. Acta Derm Venereol (Stockh) 1998; 78: 156 – 157.
2. Bauer AW. Beiträge zur klinischen Konstitutionspathologie. V. Heredofamiliäre Leukonychie und multiple Atherombilderung der Kopfhaut. Z Menschl Vererb Konstitutionslehre 1920; 5: 47 – 58.
3. Galadari I, Mohsen S. Leukonychia totalis associated with keratosis pilaris and hyperhidrosis. Int J Dermatol 1993; 32: 524 – 525.
4. Giustina TA, Woo TY, Campbell JP, Ellis CN. Association of pili torti and leukonychia. Cutis 1985; 35: 533 – 534.
5. Friedel J, Heid E, Grosshans E. Le syndrome "FLOTCH". Survenue Familiale d'une LeucOnychie totale, de kystes Trichilemmaux et d'une dystrophie Ciliaire a Heredite autosomique dominante. Ann Dermatol Venereol 1986; 113: 549 – 553.
6. Bart RS, Pumphrey RE. Knuckle pads, leukonychia and deafness: a dominant inherited syndrome. N Engl J Med 1967; 276: 202 – 207.
7. Crosby EF, Vidurrizzaga RH. Knuckle pads, leukonychia, deafness and keratosis palmoplantaris: report of a family. Johns Hopkins Med J 1976; 139: 90 – 92.
8. Bushkell LL, Gorlin RJ. Leukonychia totalis, multiple sebaceous cysts and renal calculi. A syndrome. Arch Dermatol 1975; 111: 899 – 901.
9. Crosti C, Sala F, Bertani E, Gasparini G, Menni S. Leuconychie totale et dysplasie ectodermique. Observation de deux cas. Ann Dermatol Venereol 1983; 110: 617 – 622.

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Response to the Letter by Professor Grosshans

Sir,

Professor Grosshans's letter raises interesting points about the nature of congenital leukonychia totalis as presented in our case (1). We gave the patients a thorough examination: there was no sign or symptom of any associated ectodermal dysplasias in either brother. Some authors have found evidence of leukonychia totalis – associated disorders inconstantly affecting hair shafts, hair sheats, eye lashes, and stratum corneum of palms and soles (2 – 4). The hypothesis of congenital familial leukonychia totalis as one main symptom of a more complex ectodermal dysplasia in keratinizing structures is a promising idea which must be further substantiated by analysing the genetic background too.

REFERENCES

1. Köhler LD, Möhrenschlager M, Ring J. Congenital leukonychia totalis in two brothers. Acta Derm Venereol (Stockh) 1998; 78: 156 – 157.
2. Giustina TA, Woo TY, Campbell JP, Ellis CN. Asssociation of pili torti and leukonychia. Cutis 1985; 35: 533 – 534.
3. Friedel J, Heid E, Grosshans E. Le syndrome "FLOTCH". Survenue familiale d'une leuconychia totale, de kystes trichilemmaux et d'une dystrophic ciliare à hérédité autosomique dominante. Ann Dermatol Venereol 1986; 113: 549 – 553.
4. Crosby EF, Vidurrizaga RH. Knuckle pads, leukonychia, deafness and keratosis palmoplantaris: report of a family. Johns Hopkins Med H 1976; Suppl 189: 90 – 92.

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