HEREDITARY LEUCONYCHIA TOTALIS

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Leuconychia totalis is a rare and curious disorder which is met with infrequently. In contrast, leuconychia in the form of spots and bands is exceedingly common, especially in young women. Popular names for these white spots have included: "wishes," "gift or fortune spots," "lies," etc. In Bavaria it is held that a person will live as many years as there are spots on the nails.

It was Unna (1) who formulated the three divisions of the conditions: 1) leuconychia punctata—white spots; 2) leuconychia striata—white bands or striae; and 3) leuconychia totalis—involving the entire nail. Weber (2) added a fourth type similar to leuconychia totalis, but incomplete.

Leuconychia possesses a multitude of apparently synonymous names. It has been variously referred to as albugo, achromia unguium, leuchonychia, (and leukonychia), flores unguium, canities unguium, and leucopathia unguium. Unna originally designated all forms due to trauma or artificial causes as leucopathia unguium, in contradistinction to leuconychia, which term he reserved for spontaneous or idiopathic forms. This fine distinction has not been adhered to, and the two terms are used synonymously by most observers.

Leuconychia punctata or flores unguium is the common so-called white spots. Singer (3), in an observation of a group of 100 unselected patients, found that 62 per cent showed leuconychia punctata or striata of the finger nails. In direct contrast to this Eller and Anderson (4), in a comprehensive review of the literature in 1928, stated that only approximately 50 cases of leuconychia totalis had been reported, of which only seven had been reported in the American literature. Since 1928 only about five additional instances of total leuconychia have been reported (5–8) making a total of 55.

CAUSES

The causes of leuconychia of any of the three types are many. As early as 1792 Reil (9) described white spots and bands in the nails accompanying general illnesses. Beau (10), in 1846, elaborated upon these changes, particularly as they occur with febrile diseases. To date an impressive list of causes of acquired leuconychia may be found among the reported instances of the disorder. This list includes the following: 1) trauma, especially that produced by a cuticle knife, but also by occupation (butcher); 2) febrile illnesses and/or malnutrition, including typhoid fever, measles, relapsing fever, amebic dysentery, and ulcerative colitis; 3) rickets; 4) chronic arthritis and endocarditis; 5) anemia; 6) drugs, such as emetine, sulfanomides, ovarian hormone and arsenic; 7) neuritis; 8)

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Read before the Eleventh Annual Meeting of the Society for Investigative Dermatology, San Francisco, June 25, 1950.

frostbite; 9) "trophic" disturbances, such as Raynaud's disease, trophoneuroses; 10) endocrine disturbances, appearing during menstruation. Whatever the cause, it is apparently an intermittent one with the punctate and striate forms, a continuous one with the total form.

Heredity—leuconychia striata

Most of the reported cases have been of the acquired type and only a few instances of an hereditary transmission could be found. Undoubtedly leuconychia straiata and punctata may be inherited much more commonly than would be implied by the small number of recorded cases. Sibley (11) observed a woman with leuconychia striata whose maternal grandmother, daughter and "several" other members of the family had a similar disorder. DuBois (12) reported the instance of a girl, aged 22, who had a striate leuconychia of the hands and feet whose mother presented the same lesions.

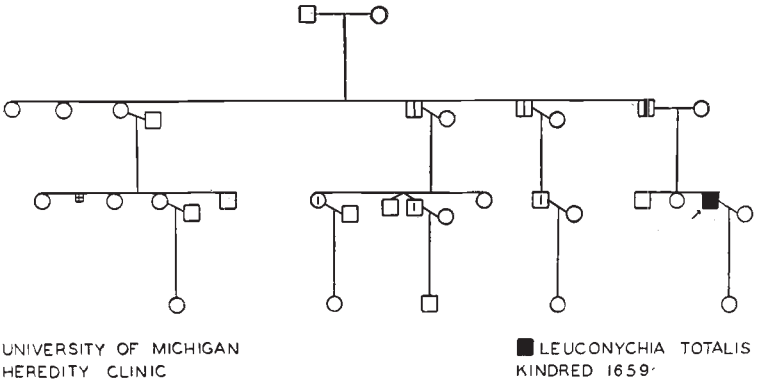

FIG. 1. Pedigree of a family with leuconychia totalis

Heredity—leuconychia totalis

Only three instances of hereditary leuconychia totalis could be found in the literature. In 1913 Gutman (13) observed a 30 year old man with leuconychia totalis whose father also had the condition. Fox and Pisko (14) reported the condition in four individuals in three generations, a father, son, daughter, and grandson. These patients were partially negro and the condition was especially prominent in the grandson who had the darkest skin. The largest reported family was that of Bauer (15) in which 19 individuals had leuconychia totalis, 17 of whom also had multiple sebaceous cyst formation. There were 10 affected males to 9 females and the condition appeared to be a simple dominant.

Pedigree

Figure 1 illustrates the pedigree of a family which the authors have drawn up from the study of an individual with leuconychia totalis. The propositus (designated by the arrow) is a 25 year old white male of Italian descent. He presented porcelain-white nails of both hands and both feet which he had had since shortly

after birth. There were no other abnormalities of any kind, including the hair and teeth. The nails were otherwise entirely normal in shape, surface, and thickness. His father and his father's two brothers were reliably reported by the propositus to have the same condition of the nails. Unfortunately it was impossible

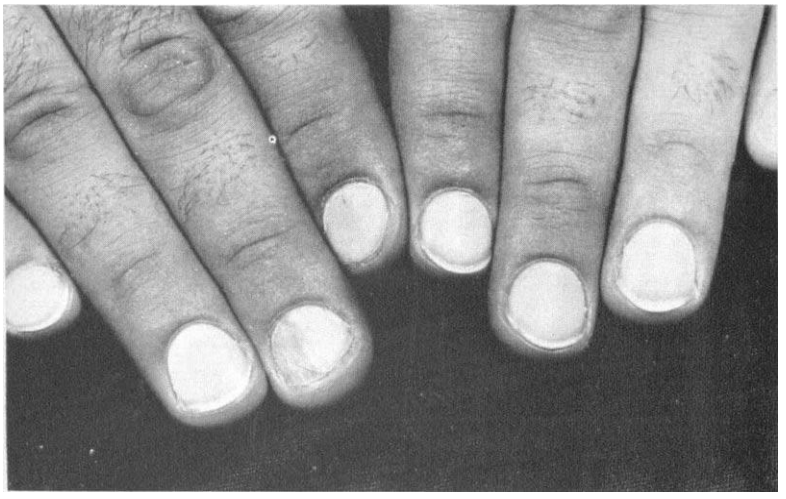

FIG. 2. Fingernails

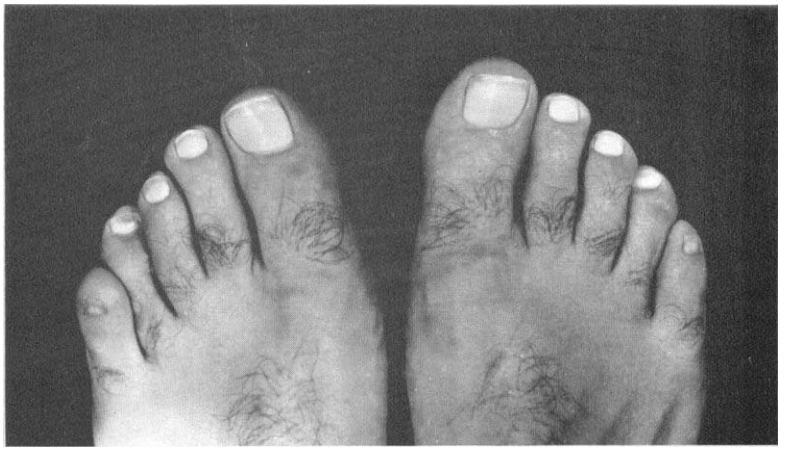

FIG. 3. Toenails

to examine the father and uncles because they reside in Italy. Figure 2 shows the fingernails and Figure 3 the toenails. There no pink nails.

ASSOCIATED FINDINGS

Usually there are no other associated findings either dermatologic or otherwise. The following are the few associated findings which have been reported. In Unna's case some hair was also white, so-called "leucotrichia." Abraham (16)

reported the occurrence of a bandlike area of frontal alopecia in a 28 year old woman who had leuconychia totalis. Darier and LeSourd (17) had a patient who had an associated total alopecia. In Betterman's (18) case, a 23 year old woman, there was an associated extensive vitiligo of the neck, abdomen, back, forehead and hand.

COMMENT

Several explanations have been set forth to explain the production of leuconychia. It was formerly held that the whiteness was due to imbibition of air. Singer (19) stated that the presence of air spaces between layers of insoluble keratin was due to the technic of softening the nail in strong nitric acid with the kerato-hyaline being easily dissolved. Woolf (20) believed that the leuconychia was due to capillary damage and subsequent malnutrition. Singer (19) in 1931 stated that all causes of leuconychia "cause either a local or a general increase in the metabolic rate."

He further stated that the increase in metabolic rate "causes an increase in the peremeability of the cells, and the cells as they are forced away from the root of the nail receive food and oxygen and remove their waste products for a longer time than normally. In this way keratinization is delayed, for the earlier condition of kerato-hyaline granules persists." Josephson and Lerner (21) also believed that it was a generalized disorder which was responsible for the condition. They reported that leuconychia in the form of spots and bands as it occurs with catarrhal conditions of the upper respiratory tract is symptomatic of a constitutional metabolic disorder (a diet high in carbohydrate) which responds to high doses of ammonium chloride and to other substances which effect a shift of the acid-base balance in the direction of acidosis.

In the hereditary cases of leuconychia totalis there has been no mention made of any other findings which would indicate either an increased metabolic rate or a change in the acid-base balance. Admittedly no laboratory findings were done to determine specifically the presence of such abnormalities. In both the acquired and hereditary cases of leuconychia totalis the nails have been normal in all other respects. There was no change in the texture, thickness nor shape of the nails. Eller and Anderson (4) found that in their case, a 15 year old boy, the nails were thicker than usual. In addition there were no symptoms. The nails are best described as having the appearance and color of the normal lunula throughout the length of the nail. The whiteness has been variously described as bluish, chalky, ivory, milky, and porcelain white. Porcelain white would seem to be the best descriptive term in the case reported in this paper.

If nail trimmings from leuconychia totalis nails or from the white portion of leuconychia striata nails are embedded, sectioned, and stained with ordinary H. and E. strain, it is found that the cells in the middle one-third are larger than normal and that both cells and nuclei take only acid stains. These cells retain their nuclei. Heidingsfeld (22), Becker (23) and others have all found essentially the same histologic features. Becker (23) states that "the process is evidently one of abnormal keratinization." Histologic sections stained with H. and E. from the case reported in this paper showed the same features as recorded by Becker.

Except in the family reported by Bauer there are no abnormalities of any of the other accessory skin structures. In the family reported by Bauer all but two had multiple sebaceous cyst formations. Cockayne (24) was of the opinion that this was "probably an example of linkage between the two defects, the genes for both being in he same chromosome. Crossing over would account for the two with only the nails abnormal." In the family reported in the pedigree in this paper only the propositus‡ was examined and he had leuconychia totalis without any other physical abnormalities. His father and two paternal uncles are reliably reported by the propositus to have leuconychia totalis. The disorder would appear to be inherited in a simple dominant fashion as it has been in the other recorded pedigrees in the literature. The occurrence in four males in one family may be coincidence or may be due to a sex limitation. It is amazing that a gene can be so specialized as to affect only the cells which elaborate nails and not affect any other structures. In contrast to the acquired forms of leuconychia, where on very rare occasions serious disease may be the cause of the disorder, hereditary leuconychia totalis seems to be a purely "cosmetic" defect and of academic interest only.

TREATMENT

Becker (23) treated one thumbnail of a patient with leuconychia striata by means of roentgen rays, with apparent aggravation of the condition. The nails are of cosmetic interest only in this condition and can be easily painted with an alcoholic solution of eosin or nail polish to cover the whiteness.

SUMMARY

1. The probable existence of a family of four individuals with leuconychia totalis is reported; however, it was possible to examine only one, the propositus.

2. A review of the classifications and the causes of leuconychia is presented with the theories as to mechanism of production of the phenomenon.

3. Hereditary leuconychia totalis would appear to be a medical curiosity rather than any serious disorder.

REFERENCES

1. UNNA, P. G.: Histopathology of the Diseases of the Skin. English ed.: Macmillan and Co., New York. 1896. 1049–51.
2. WEBER, F. P.: Some Pathological Conditions of the Nails. International Clinics, Lippincott, Philadelphia, 28th series, **1:** 108–30.
3. SINGER, P. L.: Leukonychia—Its normal occurrence and causation. Arch. Dermat. & Syph., **24:** 113, (July) 1931.
4. ELLER, J. J., AND ANDERSON, U. P.: Leuconychia totalis—Clinical report with a review of the literature. Med. J. & Rec., **127:** 318, (March) 1928.
5. COSTA, O. G.: Congenital Total Leuconychia: Case. An. Brasil. de dermat. e sif. **22:** 147, (September) 1947.
6. AOSIMA, S.: Total Leukonychia: Case. Higu-to-Hitunyo (Abstr. Sect.) **8:** 4, (February) 1940.
7. STUBENBORD, J. G., AND STUBENBORD, W. D.: Leukonychia totalis: Case. Arch. Dermat. & Syph., **32:** 761, (November) 1935.

‡ Individual through whom information concerning the rest of the family is obtained.

8. Mukai, T.: Leukonychia. Acta. Dermat., **16:** 389, 1930.
9. & 10. Quoted by Sibley, K. W.: Discoloration Unguium: 1.) Leucopathia Unguium; 2.) Ungues Flavi. Brit. J. Dermat., **23:** 281, (August) 1911.
11. Sibley, K. W.: Leuconychia striata. Brit. J. Dermat., **34:** 238, (July) 1922.
12. DuBois, C.: Un cas de leuconychie semilunaire congenitale. Schweiz. Med. Wchnschr., **9:** 422, 1928.
13. Gutman, P.: Berlin. Klin. Wochenschr., (Verein der Aerzte Wiesbadens), **50:** 1459, (August) 1913.
14. Fox, H., and Pisko, E.: Leuconychia in three generations. J. Cut. Dis., **35:** 559, 1917.
15. Bauer, A. W.: Beitrage zur Klinischen Konstitutionspathologie V. Heredofamiliare Leukonychie und Multiple Atherombildung der Kopfhaut. Anat. u. Konstitutionslehre, Bd. 5, 44–58, 1920.
16. Pernet, Mr.: Dermatological Society of Great Britain and Ireland. Brit. J. Dermat., **12** 100, (March) 1900.
17. Darier, and LeSourd: Pelade decalvante avec des lesions des les angles. Ann. de Dermat. et de Syph., **9:** 1009, (November) 1898.
18. Betterman, S.: Leuconychia Totalis. Dermat. Zeitschr., Berlin. **13:** 461, 1906.
19. Singer, P. L.: Leukonychia—Its normal occurrence and causation. Arch. Dermat. & Syph., **24:** 113 (July) 1931.
20. Woolf, M. S.: Leukonychia striata. Arch. Dermat. & Syph., **12:** 520, (October) 1925.
21. Josephson, E. M., and Lerner, C.: Leukonychia and "colds" secondary to disturbances of metabolism. Arch. Dermat. & Syph., **29:** 703, (May) 1934.
22. Heidingsfeld, M. L.: Leucopathia unguium. J. Cut. Dis., **18:** 490, (November) 1900.
23. Becker, S. W.: Leukonychia striata. Report of a congenital case. Arch. Dermat. & Syph., **21:** 957, (June) 1930.
24. Cockayne, E. A.: Inherited Abnormalities of the Skin and Its Appendages. Oxford University Press, London. 1933.

DISCUSSION

Dr. George C. Andrews: I also have a patient with this condition and when I get home I shall have him investigated along these lines.

Dr. W. T. Kruse: We had hoped to contact the physician in Italy and have the other three individuals examined to see if they also had the same disease. Certainly from what we can find in the literature this is a very interesting case, and hitherto unknown.