Dominant Anonychia and Onychodystrophy

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Simple hereditary familial anonychia and onychodystrophy has been reviewed by Vogel and Dorn (1964) and by Maisels (1966). The appearances in the recorded cases range from pure onychodystrophy to the absence of one nail (anonychia policum) or total finger-nail anonychia. The present case report shows the variations observed within one family over three generations.

Present Investigation

Fig. 1 shows the pedigree of this family. I.1 and II.5 are reported to have been affected. Three cases in direct descent could be studied in detail.

Case 1. The grandfather, II.3, born in 1889, suffers from lymphatic leukaemia and diabetes. As can be seen in Fig. 2a, the hands show complete anonychia of the thumb, the forefinger, and the middle finger, with nail-bed vestiges present as small ridges on the outer finger edges. The fourth and fifth fingers show the absence of nails in the central part, and two small atrophied nail lamellae on the sides. In the feet, Fig. 2b, there was total anonychia on the big toe, a rudimentary ridge on the second right toe, thin, narrow nail lamellae on the second and third toes, and hypoplastic nails on the third and fourth toes. The bulging aspect of the terminal phalanges on the first, second, and third toes is noteworthy.

Case 2. The daughter, III.6 born in 1926, is obese. In the hands (Fig. 3a) there was total anonychia on the forefinger and the middle finger of the right hand, and on the left forefinger (with small sunken areas on the sides, corresponding to the nail edge); nail rudiments, nearly punctiform, on both thumbs and hypoplastic nails on the fourth fingers, while the fifth fingers had normal nails. The feet (Fig. 3b) showed total absence of nails and nail beds on the first and second right toes, hypoplastic nails on the other toes, the degree of hypoplasia decreasing towards the ends.

FIG. Pedigree of family.

FIG. 2a.

FIG. 2b.

FIG. 3a.

FIG. 3b.

FIG. 4a.

FIG. 4b.

FIG. 2, 3, and 4 show the hands (a) and feet (b) of Cases 1, 2, and 3 respectively.

Case 3. The grandson IV.1, born in 1945, is healthy. His hands (Fig. 4a) show total absence of nails on both forefingers and middle fingers, with sunken areas at the outer edges of the fingers. Lamellar nail rudiments on both thumbs and on the right fourth finger were present, while the left fourth finger and both fifth fingers had normal nails. The feet (Fig. 4b) showed nearly complete lack of nail beds on the first left and right toes, with small nail rudiments; rudimentary nail lamellae were seen on the other toes.

None of the cases showed any radiological evidence of anomalies in the patellae. The chromosome patterns were normal.

Summary

Uncomplicated onychodystrophy, dominantly transmitted over three or possibly four generations, is recorded. The forefingers, middle fingers, and big toes showed anonychia. The fourth and fifth fingers and toes were mainly normal, and varying degrees of onychodystrophy were seen in the remaining digits.

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