# Hereditary leukonychia totalis

Sir,

Leukonychia or white nails have been known from a long time. They are commonly known as gift or fortune spots. Unna classified leukonychia based on distribution of whiteness into the following - leukonychia punctata, leukonychia striata, leukonychia totalis and leukonychia partialis. Others classified it into true leukonychia, where the pathology involves the nail plate and apparent leukonychia, where the pathology is in the subungual tissue. Hereditary total leukonychia is a rare clinical entity.

A 66-year-old male patient presented with itchy and scaly lesions over the body of 2 months' duration. Initial skin lesions were noted on the trunk that gradually extended over other parts of the body. There was no history of photosensitivity, oral ulceration or loss of hair. He was a known diabetic and was on metformin therapy. His eruptions were not attributable to metformin. For the previous 4 months, he was also taking ayurvedic medicines of unknown nature for presbyacusis. On cutaneous examination, he had multiple violaceous scaly papules as well as plaques distributed all over the body. Postinflammatory hyperpigmentation was observed at a few places. There was no abnormality in the oral or genital mucosa. All of his 20 nails showed white discoloration that involved the entire nail plate [Figure 1]. On further enquiry, the patient informed that the nails had

been white ever since he could remember. His maternal grandfather, mother, two of the four siblings and two of his three offsprings have had a similar condition of the nails. However, no one in his family had diabetes.

He was diagnosed to have a lichenoid drug rash with hereditary total leukonychia. He was advised to discontinue the use of ayurvedic medicines and was treated with oral dapsone 100 mg/ day and was reassured regarding his white nails. During the next follow-up, after 4 weeks, he had marked improvement in his lichenoid rash; however, the nails remained white.

Abnormal keratinization of the nail plate is believed to be responsible for development of true leukonychia.[1] Large keratohyaline granules are found in the keratinocytes and the keratohyaline containing cells reflected light, resulting in a white nail that prevents the visualization of the underlying vascular tissue.[2] It is difficult to ascertain, whether our patient had true or apparent leukonychia. Our patient refused a nail biopsy.

There are very few reports of hereditary leukonychia totalis in the literature.[2-9] Familial leukonychia totalis has been found to be inherited in both autosomal dominant[2] and recessive patterns.[5]

Familial leukonychia totalis is also found to be a component of some syndromes– syndrome of leukonychia totalis, multiple sebaceous cysts and renal calculi;[10] leukonychia totalis with knuckle pads and deafness;[11] koilonychia and multiple sebaceous cysts[12] in association with deafness, hyperkeratosis palmaris and plantaris, scalp modifications and dental lesions;[13] duodenal ulcer and gallstones[14] and also as leukonychia totalis associated with hypotrichosis and keratoderma.[15]

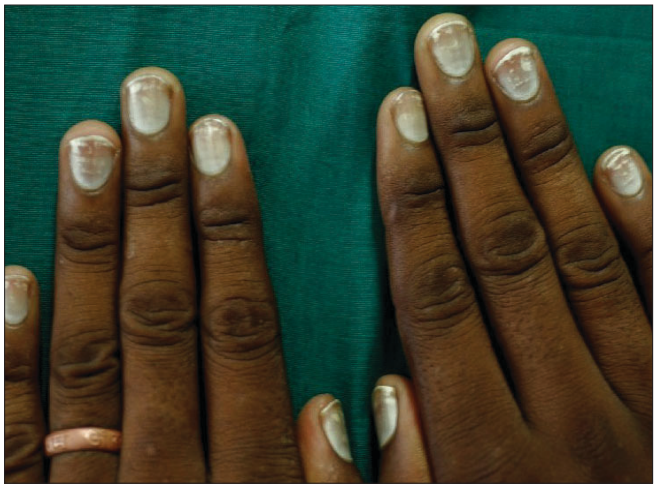

**Figure 1: White discoloration involving the entire nail plate of all finger nails**

Leukonychia totalis has also been associated with severe systemic diseases such as hepatic cirrhosis, chronic renal failure, congestive cardiac failure, diabetes mellitus, chronic hypoalbuminemia and Hodgkin's lymphoma.[16] Although our patient had diabetes mellitus, it does not appear to be the cause of leukonychia in him since he had the discoloration since early childhood and many of his family members were having a similar problem.

It has been proposed that leukonychia partialis may be a phase of leukonychia totalis, both being the expressions of the same genetic defect.[17] Both conditions occurring in different members of the same family[18] and on different digits of an individual have been described.

The importance of this report lies not only in the fact that this condition is rare but also that it may have associated abnormalities. If no association is detected, proper counseling of the patient regarding the benign nature of the condition should suffice.

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