

## Congenital True Leukonychia Totalis Case Report, Beyond The Skin

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## 1. Abstract

Congenital true leukonychia is an infrequent disorder characterized by a white coloration of the nail since birth without organ or syndromic abnormalities. We present the case of a 16 years old boy with isolated white nail coloration with psychological impact, no syndromic or organ alterations were found.

## 2. Introduction

Leukonychia is defined as a nail disorder characterized by the appearance of white macules on the nail plate that generates high psychosocial impact and scholar bullying which turns into a frequent dermatological consultation. Leukonychia is caused by an alteration in the keratinization progress at nail matrix cells that manifest with the white coloration at the nail plate. Leukonychia can present different types of expression according to the size of the lesions as little spots defined as leukonychia punctata, long bands or leukonychia striatum, and a full white coloration or leukonychia totalis. According to the time onset of the lesions, it can be classified as hereditary (HL) or acquired (AL) [1,2].

Leukonychia merits the performance of a thorough physical examination and laboratory studies due to the relation with hepatic, renal, infectious, immunosuppression, trauma, or exposition to drugs, however, it can also be part of a set of organic and dermatological manifestations in syndromic entities, and less frequently it

can be an isolated idiopathic clinical sign [3].

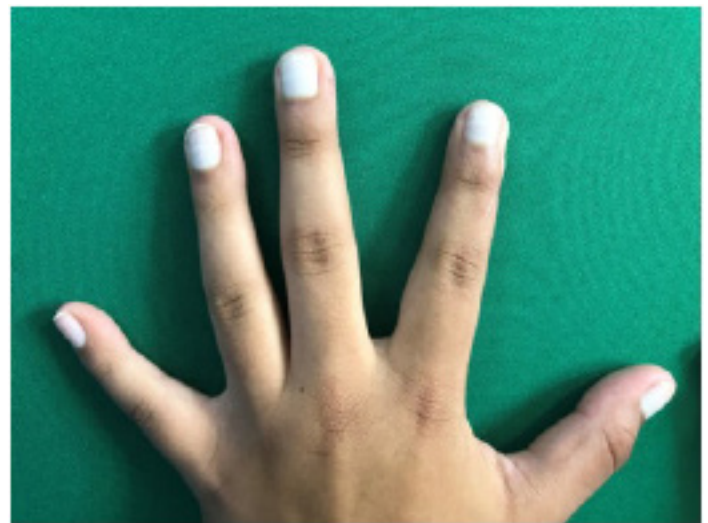
Hereditary leukonychia (HL) is extremely rare. Since 1913 a total of 44 cases have been reported to date, of which 63% (n = 28) do so in an isolation way [4]. In this work we describe a patient with leukonychia totalis since birth, presenting clinically as an isolated entity and without syndromic alteration, then configuring for the diagnosis of leukonychia totalis congenital isolated. Additionally, a review of leukonychia is performed

## 3. Case Description

A 16 years old patient, without clinical record. He consulted due to white coloration of the ten fingernails since birth, without alteration in the coloration of the toenails, had not presented discomfort until his school time when he suffered from bullying. The patient had no family history of nail alteration. The physical examination found a complete leukonychia of all ten fingernails, without other alterations in the nail matrix or nail bed (Figure 1 and 2). No other relative findings in dermatological evaluation. Laboratory tests were requested to rule out secondary causes of leukonychia totalis. Hemoglobin 16 mg/dl, hematocrit 45%, leukocytes 6300, neutrophils 4110, lymphocytes 1460, platelets 365000. ELISA for HIV negative, syphilis test negative, glutamic-pyruvic transaminase 9 u/L, 12 U/L glutamic oxalacetic transaminase, non-reactive HTLV I and II.



**Figure 1:** Leukonychia in the 10 fingers of the hands.

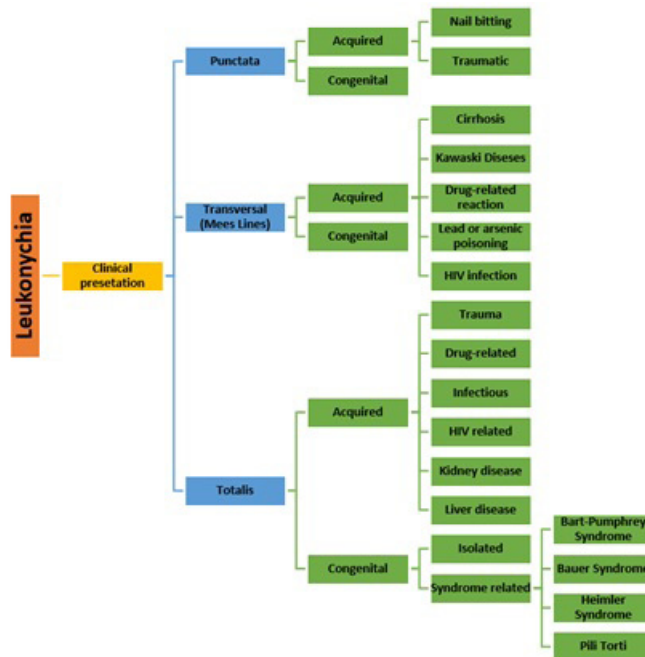


**Figure 2:** Close up photo of the true congenital leukonychia

**4. Discussion**

Alterations of nail coloring are frequent consultations in the dermatologist practice. Leukonychia is part of coloration disorders, and it is described as the appearance of white macules on the nail plate. It is considered that pathophysiologically it is generated by an alteration of keratinization in the cells of the nail matrix with the persistence of parakeratosis and the presence of large granules of keratohyalin. Leukonychia can be classified according to the size of the lesion in leukonychia punctata, striatum, or totalis, or according to its origin in hereditary or acquired (Figure 3) [1,5].

Leukonychia punctata occurs more frequently and is related to trauma and nail bite events. Striated leukonychia or also called Mees lines manifests clinically as lines parallel to the lunula and can occur in hereditary form, or present as a secondary finding of systemic diseases such as cirrhosis, patients with Kawasaki disease, a drug-related reaction, lead or arsenic poisoning, and as a manifestation due to secondary to HIV infection. Leukonychia totalis corresponds to the complete coloration of the nail plate, can be hereditary (HTL), or acquired (ATL).



**Figure 3:** Diagnosis Approach in leukonychia

ATL can be the product of trauma, local or systemic infections, medications, or inflammatory diseases. Although usually, the condition of all nail plates is uncommon, in these cases the search for immunosuppression conditions such as HIV, consumption of immunosuppressants, and liver or kidney disorders that generate an immunological or metabolic compromise in the patient should be performed.

HTL is extremely rare. Since 1913 a total of 44 cases have been reported to date, of which 63% (n = 28) do so in isolation. HL can have an autosomal dominant inheritance pattern and present as part of a syndrome, however, in some patients, no organic or systemic alterations are found and presented in isolation [6,7].

HTL can occur as part of a syndrome with alterations such as hyperparathyroidism, hypoparathyroidism, palmoplantar keratoderma, pili torti, kidney stones, sebaceous cysts, as well as Bart-Pumphrey syndrome (which is characterized by the triad of leukonychia, hyperkeratotic nodules, and deafness). Less frequently, it can be presented as a finding in Bauer, Heimler syndrome, and others [3,5].

In our case, the patient had a leukonychia totalis since birth, with an affection of the ten nail plates of the hands, he did not present neurodevelopmental alteration, deafness, or other clinical signs on physical examination; in the same way, biochemical and infectious studies were carried out in the search of immunodeficiency processes or metabolic alteration which were normal. Finally, given that in our patient the leukonychia followed a totalis pattern since birth and without other organic alterations that suggested a syndromic process, it was considered that the patient was taking a congenital leukonychia totalis. He also was remitted to a psychological evaluation to improve his low self-esteem and management of scholar bullying

## 5. Conclusion

Leukonychia is a clinical entity characterized by the white coloration of the nail plate. It is a frequent disorder in dermatological consultation that has an impact on the population. It should be borne in mind that multiple syndromic or acquired congenital clinical conditions can manifest themselves through this finding to the physical examination, so it is necessary to be attentive to make timely evaluations and diagnoses early. It can also be a manifestation of an organic or infectious status so the studies need to go beyond skin evaluation.

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