An Idiopathic Leukonychia Totalis and Leukonychia Partialis Case Report and Review of the Literature

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Established Facts

- Leukonychia can be caused by nail plate or nail bed abnormalities. Cases of idiopathic leukonychia totalis and leukonychia partialis are very rare.

Novel Insights

- We report a case of idiopathic leukonychia totalis and leukonychia partialis as an isolated finding in an otherwise healthy patient.

Key Words

Idiopathic leukonychia totalis · Leukonychia partialis · Nail · Nail cosmetics · Nail disorder · Nail pathology

Abstract

Leukonychia totalis and leukonychia partialis are rare nail findings characterized by complete or partial whitening of the nail plate. Leukonychia totalis and leukonychia partialis are usually inherited or associated with systemic disease. Here, we report the case of a 25-year-old man with idiopathic acquired leukonychia totalis and leukonychia partialis and review the literature on this topic.

Introduction

The term 'leukonychia' describes a white discoloration of the nail. Leukonychia may be caused by nail plate abnormalities (true leukonychia and pseudoleukonychia) or by nail bed abnormalities (apparent leukonychia; table 1) [1]. True leukonychia can be a benign isolated finding or may rarely be associated with other disorders. True leukonychia has been classified into four main types depending on the morphology of the white discoloration: leukonychia totalis, leukonychia partialis, leukonychia striata, and leukonychia punctata. Leukonychia totalis, where the nail is completely white, and leukonychia part-
talis, where the nail retains only a distal transverse band of normal coloration, have been proposed to represent different manifestations of the same genetic defect [2]. Here, we report the case of a 25-year-old man with concurrent idiopathic leukonychia totalis and leukonychia partialis.

Case Report

A 25-year-old African American man with a past medical history of sickle cell trait presented for an evaluation of the whitening of his fingernails that had been ongoing for 1 year. The patient reported previous treatment by two outside dermatologists for possible onychomycosis with 6 months of oral terbinafine and 3 months of itraconazole, without a noticeable change in his nails. He reported no other medication use and denied any history of trauma to his nails. The patient denied a family history of leukonychia or other dermatologic disorders. The patient, who was right-handed, reported working in information technology and spending the majority of his time at work on the computer.

On examination, the patient had complete whitening of the right 4th and 5th nails and of the left 4th nail as well as partial whitening of the right 3rd and left 2nd, 3rd, and 5th fingernails (fig. 1). The nail surface was smooth without any pits, crumbling, or nail thickening. The toenails appeared normal in color, and there was no evidence of tinea manuum or pedis. The remainder of the physical examination was unremarkable.

The histological examination of clippings from his white nails revealed parakeratosis (fig. 2). Periodic acid-Schiff stain was negative for hyphal elements. The patient’s laboratory workup was unremarkable and included a normal complete blood count with differential and negative arsenic and lead levels.

Discussion

Leukonychia describes a whitening of the nail and is one of the most common discoloration conditions of the nail. True leukonychia results from abnormal keratinization of the distal matrix that causes parakeratosis in the ventral nail plate, and this produces porcelain white nails [3, 4]. Retained keratohyalin granules in the nail plate cells reflect light and prevent the visualization of the underlying vascular nail bed. True leukonychia is distinguished from apparent leukonychia by the finding that nail bed-related leukonychia will disappear when pressure is applied to the nail plate. Psoriasis is a common cause of true leukonychia and results in punctate white areas of the nail plate from foci of parakeratosis. Another possible cause of true leukonychia is proximal subungual onychomycosis, where fungi in the ventral nail plate prevent light reflection. Proximal subungual

<table>
<thead>
<tr>
<th>Table 1. Classification of leukonychia</th>
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</thead>
<tbody>
<tr>
<td>True leukonychia</td>
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<tr>
<td>True hereditary leukonychia</td>
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<tr>
<td>Inherited syndromes (sebaceous cysts and renal calculi, Bart-Pumphrey syndrome, acanthosis nigricans, pili torti, severe keratosis pilaris, duodenal ulcers and gallstones, and LEOPARD syndrome)</td>
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<td>Acquired via chemotherapeutic exposure</td>
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<td>Acquired via heavy metal poisoning (Mees’ lines)</td>
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<tr>
<td>Acquired in association with severe acute illness (psoriasis, cardiac insufficiency, myocardial infarction, acute and chronic renal failure, pleural empyema, systemic infections, sickle cell anemia, systemic lupus erythematosus, and pellagra)</td>
</tr>
<tr>
<td>Trauma</td>
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<tr>
<td>Alopecia areata</td>
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<tr>
<td>Proximal subungual onychomycosis</td>
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<tr>
<td>Idiopathic</td>
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<tr>
<td>Apparent leukonychia</td>
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<tr>
<td>Half-and-half nails in chronic renal failure, Behcet’s disease, Kawasaki disease, liver disease, Crohn’s disease, zinc deficiency, and pellagra</td>
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<tr>
<td>Terry’s nails in liver disease, adult-onset diabetes, and congestive heart failure</td>
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<tr>
<td>Muehrcke’s lines in hypoalbuminemia, chemotherapy, and heart transplant</td>
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<tr>
<td>Pseudoleukonychia</td>
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<td>White superficial onychomycosis</td>
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<td>Keratin degranulation due to nail polish</td>
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LEOPARD syndrome: lentigines, ECG conduction abnormalities, ocular hypertelorism, pulmonic stenosis, abnormal genitalia, growth retardation, and sensorineural deafness.
**Fig. 1.** Total (leukonychia totalis) and partial (leukonychia partialis) whitening of 7 fingernails.

**Fig. 2.** Histopathology of the nail plate with parakeratosis.

**Table 2.** Reported cases of idiopathic leukonychia totalis and leukonychia partialis

<table>
<thead>
<tr>
<th>Authors</th>
<th>Demographics</th>
<th>Examination findings</th>
<th>Disease course</th>
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<tbody>
<tr>
<td>Arsiwala</td>
<td>35-year-old man, South Asian</td>
<td>LT of all 10 fingernails and of both great toenails</td>
<td>Slowly progressive, began at age 12</td>
</tr>
<tr>
<td>Bakry et al.</td>
<td>12-year-old boy, Caucasian</td>
<td>LT of all 10 fingernails</td>
<td>Gradually progressive, began at age 4; histopathology showing parakeratosis, hypergranulosis, and large keratohyalin granules</td>
</tr>
<tr>
<td>Bongiorno and Aricò</td>
<td>32-year-old man, Caucasian</td>
<td>All 20 nails involved</td>
<td>Began at age 23, gradually progressive from LP to LT</td>
</tr>
<tr>
<td>Claudel et al.</td>
<td>12-year-old boy</td>
<td>LT of 7 fingernails</td>
<td>Began 9 months earlier; histopathology notable for globular collection of large immature keratohyalin granules</td>
</tr>
<tr>
<td>Dlova and Tosti</td>
<td>20-year-old man, African American</td>
<td>LT of all 10 fingernails</td>
<td>Histopathology notable for marked parakeratosis within the nail plate</td>
</tr>
<tr>
<td>Eller and Anderson</td>
<td>15-year-old boy</td>
<td>LT of all 10 fingernails</td>
<td>LT and LP since birth with a severity that varied over time</td>
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<tr>
<td>Kim et al.</td>
<td>19-year-old man, Caucasian</td>
<td>LT and LP of all 10 toenails</td>
<td>Rapidly progressive, began 1 month prior; histopathology notable for sparse parakeratosis with globular collection of keratohyalin granules</td>
</tr>
<tr>
<td>Park et al.</td>
<td>26-year-old man, Caucasian</td>
<td>LP and LT of all fingernails except for the left thumb</td>
<td>Began at age 13, progressed slowly from LP to LP and LT, with histopathology showing a globular collection of large, immature keratohyalin granules</td>
</tr>
<tr>
<td>Stewart et al.</td>
<td>23-year-old man</td>
<td>LP and LT of all 10 fingernails and second toenails bilaterally</td>
<td>Began at age 17, slowly progressive from LP to LP and LT</td>
</tr>
</tbody>
</table>

LT = Leukonychia totalis; LP = leukonychia partialis.
onychomycosis is one of the rarest forms of onychomycosis and is a cause of true leukonychia in fingernails and toenails of immunocompromised patients. Proxi-
mal subungual onychomycosis is most commonly due to an infection from *Trichophyton rubrum*; however, *T. megnitii, T. schoenleinii, T. tonsurans, T. mentagro-
phytes, and Epidermophyton floccosum* and molds (part-
icularly *Fusarium* sp.) [5] have also been implicated in the disease [6, 7]. Fungal infection originates in the proximal nail fold and subsequently invades the ventral nail plate and travels distally. Proximal subungual onychomycosis can be diagnosed with periodic acid-Schiff staining of a nail punch biopsy.

In pseudoleukonychia, the white color is associated with superficial nail scaling, and this is seen in keratin de-
granulation due to nail polish and in white superficial onychomycosis. White superficial onychomycosis exclu-
sively affects toenails. Both dermatophytes and non-der-
atophytes can cause white superficial onychomycosis, including *T. rubrum, Cephalosporium* and *Aspergillus* sp., and *Fusarium oxysporum* [8]. Fungal invasion of the su-
perficial plate produces punctate areas of leukonychia that can eventually coalesce to involve the entire surface of the nail plate. Non-dermatophytes, which are an un-
common cause of white superficial onychomycosis, can invade into the depths of the nail plate as seen in ‘deep’ white superficial onychomycosis [9]. Because the most super-
ficial aspect of the nail plate is always affected, the nail plate texture is often rough and crumbles easily. A potassium hydroxide examination of nail plate scrapings is used to diagnose white superficial onychomycosis.

Leukonychia totalis and leukonychia partialis can be inherited as an isolated finding [2, 10–15], as part of a syndrome [16–22], or can be acquired. Numerous inher-
ited syndromes involving skin or skin appendage abnor-
malities can be associated with leukonychia. An autoso-
mal dominant pattern of inheritance has been reported in patients with concurrent sebaceous cysts and renal cal-
culi [16] as well as in Bart-Pumphrey syndrome, which is characterized by knuckle pads and deafness [22, 23]. True hereditary leukonychia has also been associated with ac-
anthosis nigricans [21], pili torti [19, 20], severe keratosis pilaris [18, 19], duodenal ulcers and gallstones [17], and LEOPARD syndrome [24].

Idiopathic acquired true leukonychia totalis and leuk-
onychia partialis is a very rare condition and is not associ-
ated with other systemic findings. Ten cases have previ-
ously been reported, making this the 11th case [3, 4, 25–
30]. Surprisingly, all reported cases have been young men or boys (table 2). Patients were aged 12–32 years (mean 21.0) at the time of diagnosis, with reported symptoms first manifesting from the time of birth to age 26 (mean 14.7). Patients came from varied ethnic backgrounds. In addition to fingernail involvement, several patients also had affected toenails, and 1 patient only had toenail in-
volve [3, 4, 26, 27, 31].

It has been proposed that leukonychia partialis is a sub-
type of leukonychia totalis, and that both are different manifesta-
tions of the same underlying genetic defect [2]. Support-
ing evidence can be seen in individuals who may have concurrent leukonychia totalis and leukonychia partialis on different digits, as seen in our patient. Addi-
tionally, the majority of reported cases of idiopathic leuk-
onychia progressed over time from leukonychia partialis into leukonychia partialis and leukonychia totalis. Leuk-
onychia partialis is characterized by intermittent periods of normal and abnormal nail matrix keratiniza-

In leukonychia totalis, the nail plate is noted to be th-
inner, we report a case of idiopathic true leukonych-
ia totalis and leukonychia partialis in a young man with-
out any other associated symptoms. This is the 11th re-
ported case of idiopathic true leukonychia, which is a very rare disease with a chronic course. Leukonychia is a more serious cosmetic problem for patients with darker skin tones and more visually apparent in these patients.

**References**

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