

Idiopathic acquired true leukonychia: a unique entity

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Abstract

Idiopathic acquired true leukonychia is a rare benign acquired disorder that typically affects adults. Diagnosis is made clinically, with investigations required to exclude any underlying cause. The cause of this condition is largely unknown, yet it understandably causes significant anxiety to patients due to concerns about underlying systemic disease. We report a case of idiopathic acquired true leukonychia totalis in a healthy 20-year-old man. Intermittent complete whitening of some of the fingernails was reported over a 7-year period. No underlying medical cause or laboratory abnormality was identified. This case highlights the importance of considering idiopathic acquired true leukonychia as a differential diagnosis of nail whitening. In addition, increased awareness of this unique entity is needed for patient reassurance, as well as avoidance of unnecessary investigations and prolonged systemic treatment.

Keywords: true leukonychia, nails disorders, idiopathic leukonychia, acquired leukonychia, nail disease

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Introduction

Leukonychia, or white nails, is a benign sign in the majority of cases; however, it can be a sign of an underlying disease or a congenital disorder. Leukonychia can occur due to nail bed or nail plate abnormality, and it can be classified according to morphology or pathology (1, 2). One way to classify leukonychia is to divide it into three subtypes: apparent leukonychia, which occurs due to nail bed abnormalities; true leukonychia due to the nail plate or matrix abnormalities; and pseudo-leukonychia secondary to whitening of the superficial nail plate (1). Another classification according to morphology categorizes leukonychia into total, striate (classified as transversal or longitudinal), and punctate (2). The various leukonychia subtypes can coexist on the same digit or on different digits in the same patient (3). When true leukonychia occurs without an underlying cause or genetic association, it is called idiopathic acquired true leukonychia. This is a rare benign and chronic condition, with only a few cases reported to date. In this brief report, we present our own case and provide a summary of the published reports and their characteristics.

Case report

A 22-year-old healthy male reported complete whitening of the fingernails that started at age 15. This discoloration would last for a few months and then resolve spontaneously. However, 6 years later his complaint recurred but did not resolve. The leukonychia started as transverse porcelain-white lines, progressing distally as the nail grew and then coalescing to produce total leukonychia (Fig. 1a). There was no reported history of trauma to the nail unit, manipulation of the nail, or manicure procedures. The patient was otherwise healthy with no history of immunosuppressive states or drug intake. The episodes were not preceded by stressful events or occupational exposure to arsenic or other metals. The patient had previously undergone multiple long courses of oral antifungal agents, including fluconazole and terbinafine, with no impact on the disease's natural progression. Mycology results were negative.

Physical examination revealed true leukonychia involving

most of the fingernails, varying between partial and total involvement. The toenails were completely spared. Dermoscopy revealed deep white leukonychia that did not disappear with pressure. The cuticle was normal and there were no signs of abnormal vessels (Fig. 2). The skin, teeth, and hair were all found to be without abnormalities.

Laboratory tests, including a complete blood count, liver and kidney function tests, thyroid profile, vitamin B12, vitamin D, and serum calcium levels, were all within normal range. Serology for hepatitis and HIV was negative.

After a few months, the patient reported gradual and spontaneous improvement with eventual complete resolution (Fig. 1b), only for symptoms to recur a few months later (Fig. 1c). A diagnosis of acquired true leukonychia totalis was made, and the patient was counseled regarding the benign nature of the disease. No treatment was given, and the patient was advised to avoid nail trauma, follow good nail hygiene, and return for a follow-up visit if any changes or progression occurred.

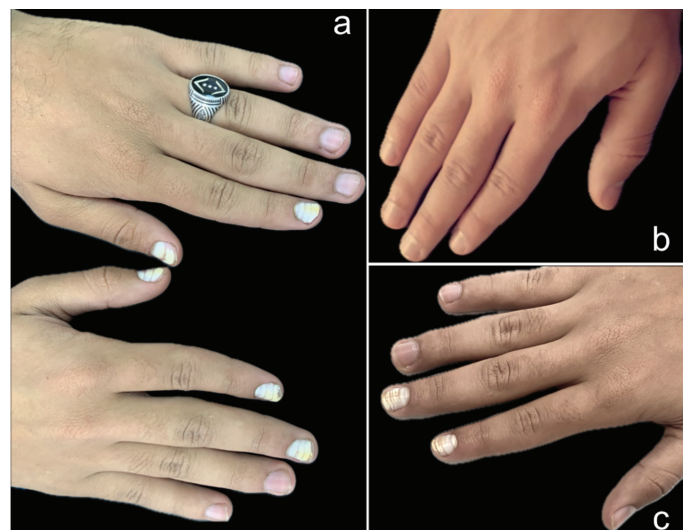


Figure 1 | a) Leukonychia totalis on both indices, thumbs, and right middle fingernails, b) spontaneous resolution of leukonychia with normal appearing fingernails, c) recurrence of true leukonychia on the right middle and index fingers. In addition, there was involvement of the left index and thumb nails (not shown).

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Figure 2 | Dermoscopy showing transverse white leukonychia involving the entire nail; the cuticle is normal, and there are no signs of abnormal vessels.

Discussion

Leukonychia refers to white discoloration of the nail and is considered one of the most common nail dyschromias (4, 5). The disorder may present as true leukonychia or pseudo-leukonychia due to nail plate pathology, or as apparent leukonychia resulting from abnormalities in the nail bed (4). Application of pressure to the nail can help differentiate these two entities because true leukonychia persists with pressure whereas apparent leukonychia disappears (4, 5).

The nail pathology underlying true leukonychia is related to abnormal keratinization within the distal nail matrix resulting in parakeratosis and retention of keratohyalin granules within the ventral nail plate (4, 5). On the other hand, pseudo-leukonychia results from keratin degranulation attributed to nail polish or white superficial onychomycosis and is usually associated with superficial scaling of the nail (4, 5).

Differential diagnoses of true leukonychia are numerous and include trauma, drugs such as hydroxyurea, selenium deficiency, systemic diseases such as anemia and ischemic cardiomyopathy, and white proximal subungual onychomycosis occurring mostly

in, but not limited to, immunosuppressed patients (3, 4).

Proximal subungual onychomycosis can present as either punctate or transverse pseudo-leukonychia, whereas superficial white onychomycosis can present as punctate, transverse, partial, or total pseudo-leukonychia (3). Superficial white onychomycosis is usually limited to toenails and rarely affects fingernails (4). The site of fungal invasion is the surface of the nail plate, and the most common isolated fungal infections are dermatophytes. In immunocompetent patients, the most common causative agent is *Trichophyton mentagrophytes*, while *T. rubrum* is more frequently found in HIV patients. However, non-dermatophyte fungi, such as *Fusarium oxysporum*, *Acremonium* spp., or *Aspergillus* spp., have also been identified. These non-dermatophyte fungi are more commonly seen in healthy individuals (6, 7). On the other hand, proximal subungual onychomycosis can be seen in both fingernails and toenails, and the site of fungal invasion is the stratum corneum of the proximal nail fold. Proximal subungual onychomycosis is usually caused by *T. rubrum*, *Epidermophyton floccosum*, or *T. schoenleinii* (4).

Only 22 cases (8-26) of idiopathic acquired true leukonychia have been described in the literature (Table 1). All reported cases were described in males, who mostly reported the occurrence of leukonychia in the second decade, although there have been reports of occurrence at birth. The mean age of presentation was 21.7 years (range 10–34). In most cases, the leukonychia was progressive and persistent. Eleven out of 22 cases had involvement of all fingernails (9, 13–18, 20, 22, 23), whereas six patients had involvement of all fingernails and toenails (9, 12, 19, 21, 24, 25). Where biopsies were taken, parakeratosis or keratohyalin retention was noted.

We believe that the case described in this report is that of leukonychia totalis. At various points in time there were concurrent partial and total leukonychia, and we believe this is reflective of the evolution of the leukonychia (27). This is supported by other reports describing both partial and total leukonychia in the same patient, as well of the evolution of partial leukonychia to total leukonychia (4, 8, 9, 11, 12, 14, 15, 18, 22, 24, 25).

Conclusions

In summary, idiopathic acquired true leukonychia is a rare benign acquired disorder with only a few cases reported to this date. It can be mistaken for onychomycosis or raise concerns about underlying systemic disease. Hence, increased awareness can help avoid unnecessary investigations and prolonged treatment.

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