Chapter 6
Endonyx Onychomycosis

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Key Feature Box

- Endonyx onychomycosis is a fungal infection of the nails caused by *Trichophyton soudanense*, *Trichophyton violaceum*, and *Trichophyton rubrum*.
- Patients with endonyx onychomycosis present with milky-white discoloration of the nail plate without hyperkeratosis or onycholysis.
- Histopathology will demonstrate an abundance of fungal filaments confined to the nail plate without nail bed inflammation or hyphae.
- Treatment typically consists of oral and topical antifungal therapies; however, recalcitrant infections may necessitate combination therapy, chemical and/or surgical plate avulsion, or photodynamic therapy.

Introduction

Endonyx onychomycosis (EO) is an exceedingly rare pattern of fungal infection of the nail plate associated with the endothrix dermatophytes *Trichophyton soudanense*, *Trichophyton violaceum*, and *Trichophyton rubrum* [1–4]. Currently, there is only one report in the literature of endonyx onychomycosis caused by *Trichophyton tonsurans* [5]. In endonyx, fungal hyphae infect and directly invade the superficial and deep portions of the nail plate [4]. This unique pattern of nail invasion is conceivably related to these organisms' high affinity for hard keratins [4].

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Epidemiology

Endonyx onychomycosis appears to affect both men and women equally and is found among all age groups and ethnicities [1, 2, 6]. The exact incidence of this nail infection remains unknown; however, it is likely to be more prevalent in areas where *Trichophyton* species is endemic, such as in central and west Africa [2].

Clinical Features

Endonyx onychomycosis is clinically characterized a milky-white discoloration of the nail plate. The nail plate surface is normal and the nail has a normal thickness [3, 4]. The nail plate will be firmly attached to the nail bed [4]. The absence of hyperkeratosis and onycholysis separates endonyx from other entities, such as distal subungual onychomycosis [4]. Table 6.1 illustrates clinical features commonly present and absent in endonyx onychomycosis.

Diagnostic Clues

Endonyx onychomycosis should be distinguished from proximal subungual onychomycosis as both present with white nail discoloration without onycholysis or subungual hyperkeratosis. History and clinical examination can help in differential diagnosis. In EO invasion occurs from distal margin, and nail discoloration starts distally and progresses proximally (Fig. 6.1) in contrast with proximal subungual onychomycosis (PSO) that originates proximally and progresses distally.

Diagnosis of endonyx onychomycosis should be confirmed by direct microscopic examination and fungal cultures. Periodic acid-Schiff (PAS) stain will demonstrate tunnels of fungal elements arranged either longitudinally or transversely within the entire thickness of the nail plate [4, 6]. Importantly, there are no signs of fungal invasion or inflammation in the nail bed [4]. Culture of the nail clippings will expose the causative organism [6].

Table 6.1 Present and absent clinical features in endonyx onychomycosis

Present features	Absent features
Normal nail plate surface	Superficial desquamation
Normal nail plate thickness	Hyperkeratosis
Nail plate firmly attached to the nail bed	Onycholysis
Milky-white nail plate discoloration	Subungual changes
Nail plate fungal hyphae	Periungual inflammation

Fig. 6.1 Endonyx onychomycosis of the fingernail. The nail plate shows distal leukonychia in the absence of subungual hyperkeratosis (Courtesy of Chinmanat Tangjaturonrusamee MD)

Summary for the Clinician

Endonyx is a rare form of onychomycosis that affects all ages and both genders. Clinicians should be aware of the presentation of this rare entity, as certain forms may be resistant to standard antifungal therapy [6]. The presence of milky-white discoloration of the nail plate in the absence of hyperkeratosis and onycholysis suggests endonyx pattern [4]. PAS staining for fungal elements confined to the nail plate may help validate the diagnosis [4]. Consider treatment with standard topical and oral antifungals first, followed by combination therapy, chemical and/or surgical plate avulsion, or photodynamic therapy if the infection is difficult to treat.

Clinical Pearls

- Endonyx is an unusual variant of onychomycosis that affects men and women of all ages, races, and ethnicities.
- Patients present with opaque-white discoloration of the nail plate without subungual changes.
- Superficial nail plate involvement, hyperkeratosis, onycholysis, or nail bed inflammation should prompt consideration of an alternative diagnosis.
- Direct microscopic examination with PAS staining that demonstrates plenty of fungal hyphae in the nail plate, but none in the nail bed, is pathognomonic of endonyx.

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