

Nail Changes in Chilblains Mimicking Lichen Planus

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Abstract

Background: Chilblains or perniosis is characterized by development of pruritic or painful erythematous and violaceous papules, plaques and vesicles over acral areas on exposure to cold conditions. Most of the cases resolve with rewarming and cold protection without any adverse events. Two patients presented to us with chilblains that developed severe nail changes mimicking nail lichen planus due to this inflammatory condition.

Clinical cases: A young adult male and a female presented with chilblains on cold exposure. Severe brittleness and thinning of nails developed with onset of chilblains in winters followed by spontaneous and painless falling off of the nails. Some regrowth of nails occurred during summers with recurrence in winters. Besides onychia, pterygium formation and longitudinal striations were also present. There was no history of smoking, drug exposure, Raynaud's phenomenon or any other features suggestive of connective tissue disease or systemic illness. Anti-nuclear antibodies were negative. Nail lichen planus was considered initially as the clinical differential in both, however, the histopathology findings were not consistent and detailed history and temporal correlation indicated that these nail changes were related to severe chilblains. Both the patients were advised cold protection and oral nifedipine 10 mg twice daily which helped in improving the chilblains, however the nail condition persisted and appeared to be permanent.

Conclusions: The pathophysiology of chilblains is said to be an abnormal vascular response to cold temperatures. Chronic vasoconstriction leading to nail matrix ischemia can be postulated as a cause of the nail changes in our patients. Our patients developed severe cosmetically bothering nail changes mimicking lichen planus and even onychia which have not been described previously.

Keywords: Nail; Lichen planus; Chilblains; Perniosis; Inflammatory disorders

Introduction

Chilblains or perniosis is an inflammatory disorder characterized by development of pruritic or painful erythematous and violaceous papules, plaques or vesicles over acral areas on exposure to cold and humid conditions [1]. Most of the cases resolve on rewarming and cold protection without any adverse events. We describe two patients with chilblains who developed severe nail changes mimicking lichen planus due to their paronychia skin condition.

Case 1

A 29-year-old woman from a Himalayan state of India presented with a 15-year history of bluish discoloration of toes and fingers followed by development of flaccid blisters over proximal nail folds during winters, which ruptured painlessly with trivial trauma. This progressed to painless avulsion of the hand nails associated with itching followed by growth of flaky nails which would fall off spontaneously leaving behind pin-point bleeding spots from the nail bed. On examination, she had complete onychia involving almost all of the digits of both hands and feet. The leftover nails showed longitudinal fibrotic bands and striations. Few nails also showed pterygium formation. Loss of pulp of distal phalanges and depigmentation were also seen (Figure 1).



Figure 1: Picture of Case 1 showing (a) onychia and (b) severely dystrophic nails with depigmentation and loss of digital pulp space.

Nail biopsy showed hyperkeratosis with unremarkable epidermis with mild peri-vascular inflammatory cell infiltrate in the dermis. X-rays of bilateral feet and hands showed acro-osteolysis of the distal phalangeal bones.

Case 2

A 22-year-old Indian man presented with gradual thinning of toe-nails and finger-nails for 13 years. This was followed by shedding off of the nail plate during winters with trivial trauma to the digits and regrowth of thin, brittle nails during summers. He also had history of developing erythema and vesicles on the lateral borders of both hands and feet, associated with pruritis and pain, on cold exposure since the past 8 years. At the time of presentation, he also had longitudinal striations and pterygium. Nail matrix biopsy from the great toe showed unremarkable epidermis along with perivascular and interstitial chronic inflammation in the deeper dermis. Vascular thrombi and evidence of vasculitis were not seen. The skin biopsy showed hyperkeratosis, irregular acanthosis and mild upper dermal perivascular chronic inflammatory infiltrate. Rest of the dermis and adnexa were unremarkable (Figure 2).



Figure 2: Picture of Case 2 showing onychodystrophy, pterygium and anonychia.

Both the patients had low body mass indices which is common in patients with chilblains. There was no history of smoking or drug exposure. Both the patients had no history of Raynaud’s phenomenon or any other features suggestive of connective tissue disease or systemic illness. There was no co-existing cutaneous disease. Anti-nuclear antibodies, cryoglobulins and thyroid functions were within normal limits. Initial clinical diagnosis of nail lichen planus was considered in both cases, however, the histopathology findings were not consistent and detailed history and temporal correlation indicated that these nail changes were related to severe chilblains. Both the patients were advised cold protection and oral nifedipine 10 mg twice daily with

symptomatic improvement in chilblains, however the nail condition persisted and was apparently permanent after 2 years of follow-up (Table 1).

Comparative features of both cases
• Young adults with similar duration of disease and onset in childhood. • Lean built. • Winter aggravation. • Similar clinical and pathological features. • No triggering factors (smoking/drugs/comorbidities). • No associated cutaneous/systemic illness.
Contrasting features
• Depigmentation over areas of previous blistering in Case 1. • Loss of pulp of distal phalanges in Case 1 with penciling of fingers.

Table 1: Table depicting outline of the 2 cases.

Discussion

The pathophysiology of chilblains is said to be an abnormal vascular response to cold temperatures. It is aggravated with humid and damp conditions and commonly affects young women [2]. Genetic predisposition, inappropriate nutritional habits, anorexia, hormonal changes, systemic diseases, focal sepsis, dysproteinaemia and myelodysplastic diseases may also play a role in its etiopathogenesis [3].

Chronic vasoconstriction leading to nail matrix ischemia can be postulated as a cause of the nail changes in our patients. A single case report describes post-inflammatory melanonychia due to chilblains and another labels nail dystrophy in the form of longitudinal ridging and discoloration of nail plate in a patient with perniosis [4,5]. Most of the cases resolve with rewarming, cold protection and use of calcium channel blockers without any adverse or disfiguring events but our patients developed severe cosmetically bothering permanent nail changes mimicking lichen planus and even anonychia which have not been described previously.

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