Case Report: Chilblain Lupus Masquerading as Tinea Pedis

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Abstract

Chilblain lupus erythematosus is a condition that is frequently misdiagnosed. Our case report describes a young male from the Middle East who underwent a skin biopsy that revealed the diagnosis of chilblain lupus erythematosus after multiple incorrect diagnoses, emphasizing the importance of skin biopsy in establishing the correct diagnosis and avoiding unnecessary treatment.

Keywords: SLE; Chilblain Lupus Erythematosus; Middle East; dermatology; Rheumatology.

Introduction

Chilblain lupus erythematosus, also known as Hutchison lupus, is a rare and chronic form of cutaneous lupus erythematosus that manifests as lesions occurring during cold periods and tend to be pruritic erythematous to purplish papular, nodular and/or plaque tender lesions on the ears, nose, fingers and toes.[1] According to the Mayo Clinic Diagnostic Criteria, the diagnosis is made based on the presence of two major criteria (skin lesions in acral locations induced by exposure to cold or a drop in temperature and evidence of lupus erythematosus in the skin lesions by histopathologic examination or indirect immunofluorescence study) and four minor criteria (coexistence of systemic lupus erythematosus (SLE) or other skin lesion of discoid lupus erythematosus, response to anti-lupus erythematosus treatment, negative results of cryoglobulin and cold agglutinin studies).[2] To diagnose chilblain lupus erythematosus, both major criteria and one minor criterion must be met, which our patient fulfilled. Our case describes the experience of a young man who received multiple initial diagnoses and treatments before being diagnosed with chilblain lupus erythematosus.

Case Report

A previously healthy 22-year-old Jordanian male began experiencing a scaly rash with peeled off skin patches involving the fingers, and feet, which was associated with nail changes and bluish to purplish discoloration of the toes since December 2019. Symptoms were initially mild involving the tips of the digits and gradually progressed to involve the fingers and feet associated with ulcer formation at the tips of the toes and heels (Figure 1). Symptoms are triggered by cold exposure. Family history significant for psoriasis in his sister who was commenced on a biologic therapy. He denied recurrent fever, photosensitivity, malar rash, unintentional weight loss, night sweats, history of acute uveitis, scarring alopecia, recurrent oral or genital ulcers, dysphagia, cough, wheezing, hemoptysis, recurrent abdominal pain, heartburn, change in bowel habits, urinary symptoms, numbness or tingling extremities, weakness in extremities, pitting distal ulcers, peripheral joint pain, swelling or stiffness. On exam, he had warm extremities with slightly dusky to erythematous purple patches on the fingers of both hands and feet. They were more prominent on the toes bilaterally and associated with subungual hyperkeratosis and dystrophic nail changes (Figure 2). He was seen by multiple dermatologists throughout 2020 and was initially diagnosed with eczema and was commenced on topical

glucocorticoids with no notable improvement of his symptoms. Later he was diagnosed with fungal infection, despite negative nail cultures, and was prescribed antifungal therapy with partial improvement of his symptoms. Subsequently, he was diagnosed with possible psoriasis and was prescribed topical glucocorticoids without improvement of his symptoms. He was reevaluated in December 2020 by another dermatologist, who referred him to a rheumatologist to rule out an underlying connective tissue disease in view of the discoloration of the affected areas.

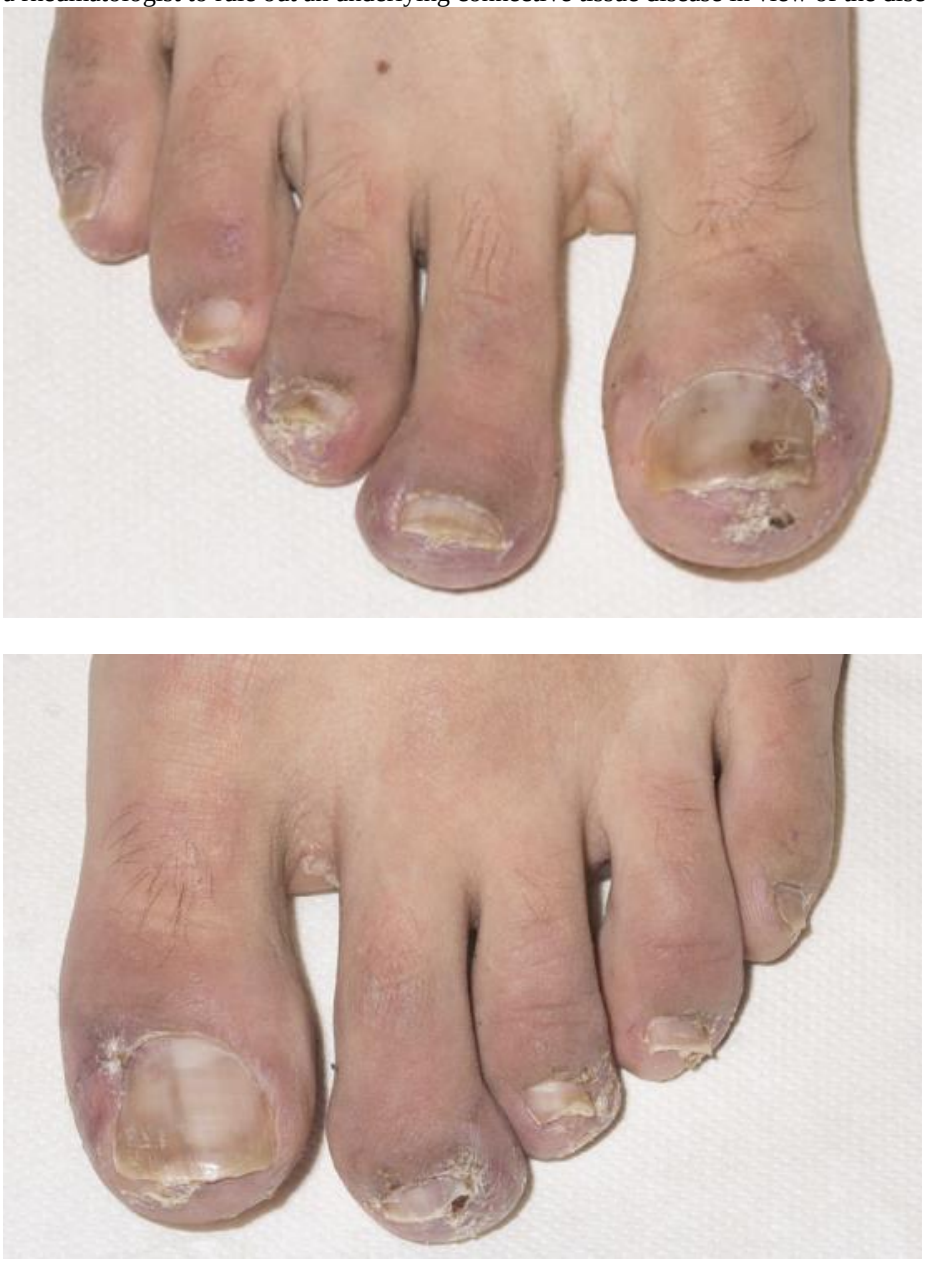

Figure 1. Feet prior to treatment. Red to violaceous pigmented patches around the toes and the nails of both feet, associated with superficial ulceration and scarring. The nail plate is partially destroyed in few toenails with subungual hyperkeratosis and petechiae. Nail fold capillaroscopy of the toes showed tortuous capillaries.

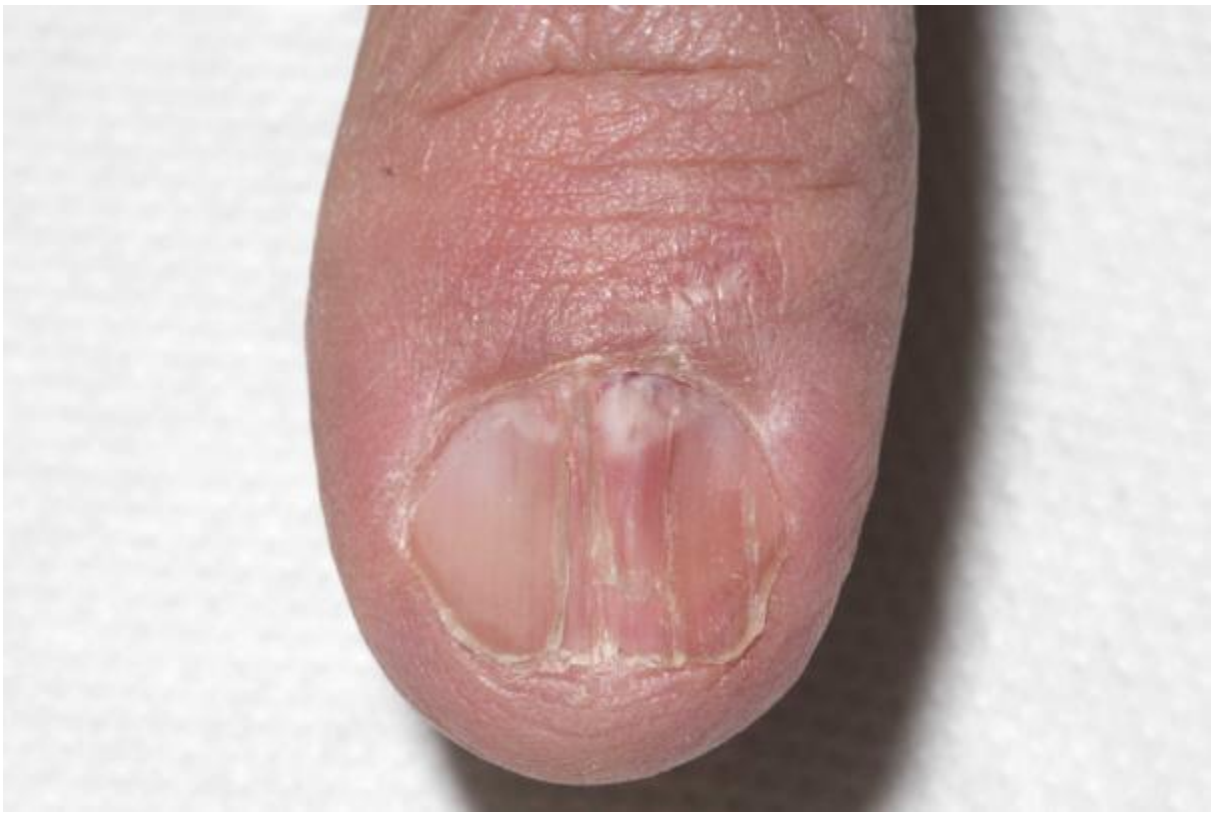

Figure 2. Right thumb prior to treatment. Longitudinal wedge-shaped nail overgrowth.

He underwent a skin biopsy of the right toe in December 2020 which showed interface dermatitis with vasculitis-like changes consistent with chilblain lupus erythematosus (Figure 3). Laboratory investigations at that time revealed a borderline elevation of the ESR [29 mm/hr] and a low titer positive immunofluorescence antinuclear antibody, [1:80 with nucleolar pattern], otherwise the rest of autoimmune work-up was unremarkable including SSA, SSB, Smith antibody, Scl-70, Smith antibody, RNP antibody, anticentromere antibody, p and c-antineutrophil cytoplasmic antibodies, C3, C4, cryoglobulins, antiphospholipid antibody profile including negative anticardiolipin IgG and IgM antibodies, beta-2 glycoprotein's IgM and IgG antibodies and lupus anticoagulant. In January 2021, he was commenced on a tapering dose of glucocorticoids starting at 20 mg with taper of 5 mg per week, in addition to hydroxychloroquine 200 mg twice daily, nifedipine 20 mg daily, aspirin 75 mg daily, topical clobetasol cream, tacrolimus ointment and topical nitroglycerin with significant improvement of his skin lesions by (Figure 4,5). He continues to follow up regularly with rheumatology as an outpatient on topical clobetasol cream without need for oral glucocorticoids.

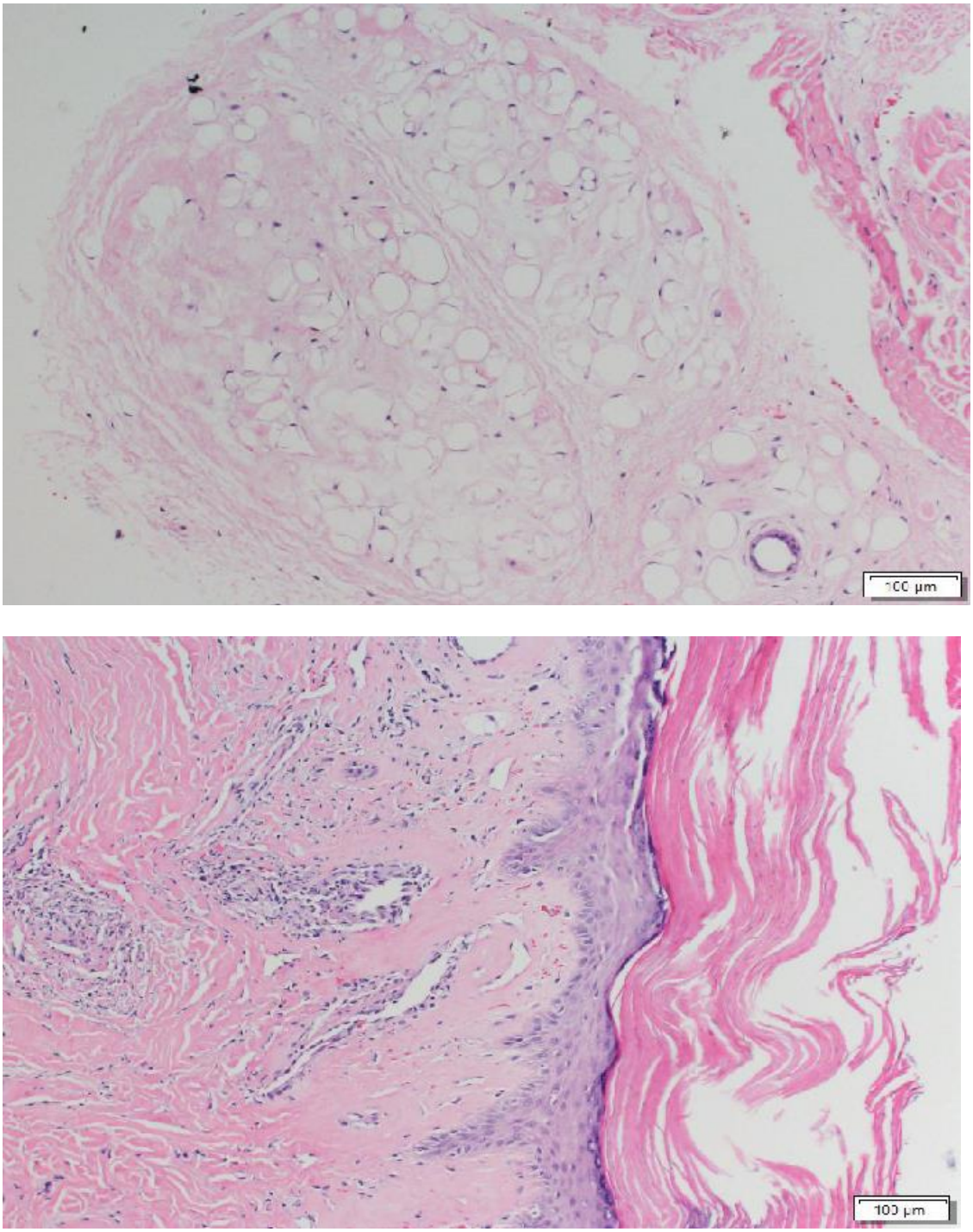

Figure 3. Right toe skin biopsy confirming chilblain lupus. The epidermis shows a thick layer of surface hyperkeratosis with associated keratin plugging of hair follicles. The underlying epidermis is thinned with focal lymphocytic infiltration in the basal layer. There is limited basal hydropic change. The basement membrane zone appears to be mildly thickened. In the underlying dermis, the stroma appears hyalinized in association with a sparse perivascular lymphocytic infiltrate present around ectatic dermal vessels and also in the intervening stroma. The infiltrate extends to a lesser extent around deep dermal vessels. The subcutis shows adipose tissue with some changes of myxoid degeneration.

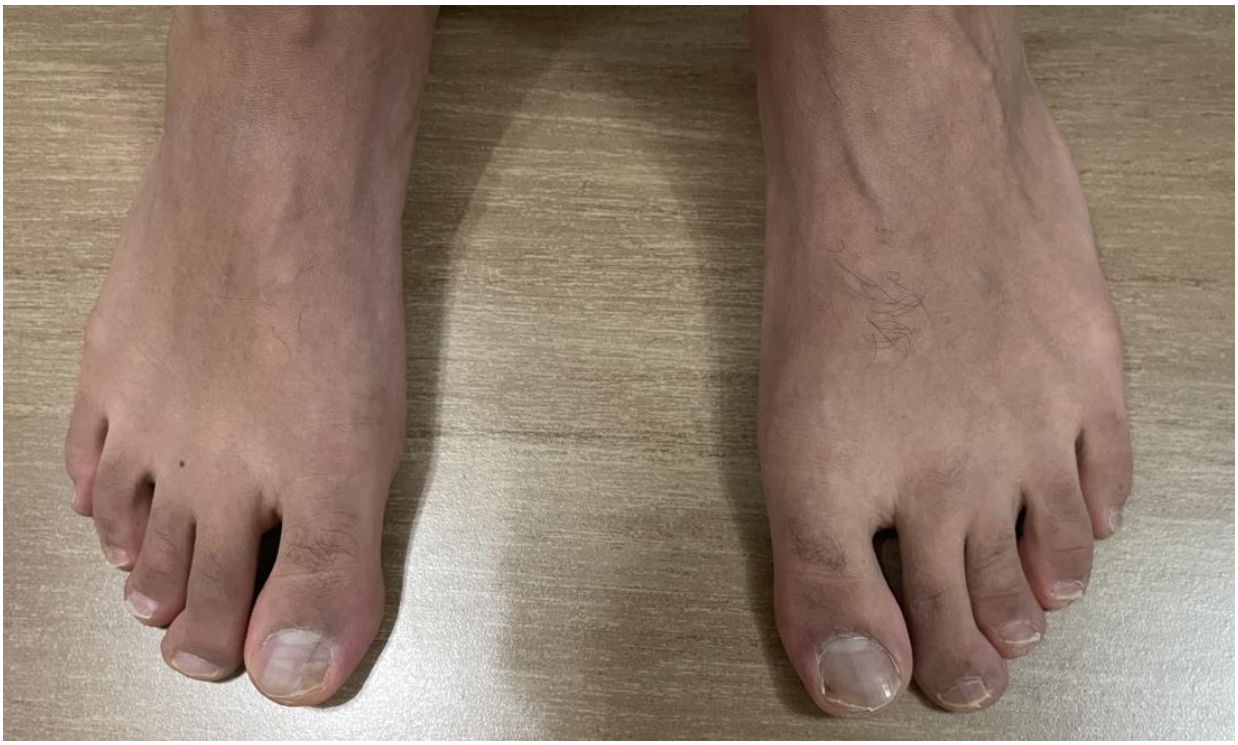

Figure 4. Bilateral feet after treatment. Significant improvement of the lesions after one year of treatment.

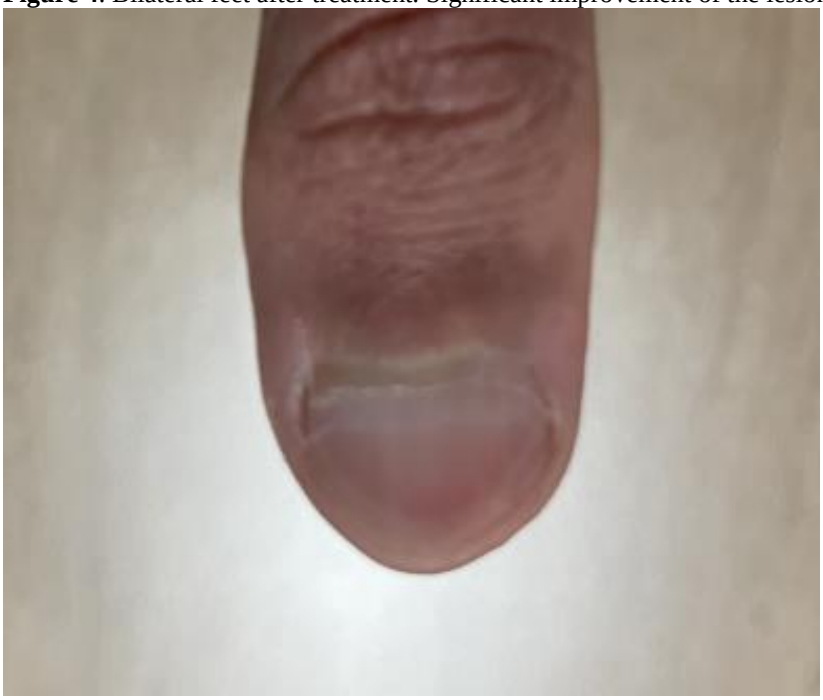

Figure 5. Right thumb after treatment. Significant improvement in the nail changes noted previously.

Discussion

In a review article published in 2008, approximately 70 cases have been reported in the literature, with only few in the adolescent age group (Hedrich et al., 2008). Additionally, no reports were found in the literature surrounding chilblain lupus erythematosus in the Middle East specifically, likely making our patient the first case report from the region. Our case report also highlights the importance of skin biopsy in establishing the diagnosis of chilblain lupus erythematosus to avoid incorrect diagnoses and further unnecessary, and potentially harmful, treatment. The first line measures consist of protection of the acral sites from low temperatures and cold weather to prevent recurrences.[1] Other treatment options include topical steroids, which have been showed to be beneficial in up to half of the patients, topical calcineurin inhibitors and calcium channel blockers.[1]

References

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