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CHILBLAIN LUPUS: A RARE FORM OF CUTANEOUS LUPUS ERYTHEMATOSUS - A CASE REPORT

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Chilblain lupus erythematosus is a rare, cold-induced form of chronic cutaneous lupus that can occur in both genetic and sporadic forms. It is characterized by acral skin lesions and is commonly associated with autoimmunity and type I interferon pathway activation. Diagnosis is based on clinical features and histological findings. While topical and systemic therapies can be effective, the disease is often chronic and relapsing. We describe a case report of a 48-year-old woman with a history of chronic spontaneous urticaria and Raynaud's phenomenon presented with a history of recurrent nodular lesions on the toes since 2015. The lesions, characterized by a burning sensation and worsening during winter, appeared as erythematous to violaceous plaques with light scaling. Laboratory investigations showed positive antinuclear antibodies (1:160) and anti-Smith antibodies, while cryoglobulins and cold agglutinins were absent. The diagnosis of CHLE was confirmed by skin biopsy, which showed perivascular lymphocytic infiltrate, interface dermatitis, and increased dermal mucin, and by nailfold capillaroscopy, which revealed severe microangiopathy and giant capillary loops. Initial management with hydroxychloroquine (200 mg twice daily), topical tacrolimus 0.1%, and warmth protection resulted in only partial improvement. However, after adding methotrexate (12.5 mg/week) to the regimen, the patient achieved almost complete clinical remission within six months. This stable remission has been maintained for six months to date, highlighting methotrexate as an effective add-on therapy for CHLE cases refractory to antimalarials.

References

1. Vinister G, Roongta R, Sinha D, et al. Chilblain lupus. *Mediterr J Rheumatol* 2023; 34(2): 269-270.

2. Doutre MS, Beylot C, Beylot J, et al. Chilblain lupus erythematosus: report of 15 cases. *Dermatology* 1992; 184(1): 26-28.
3. Millard LG, Rowell NR. Chilblain lupus erythematosus (Hutchinson). A clinical and laboratory study of 17 patients. *Br J Dermatol* 1978; 98(5): 497-506.
4. Lambertini M, Vincenzi C, Dika E, et al. Chilblain lupus with nail involvement: a case report and a brief overview. *Skin Appendage Disord* 2018; 5(1): 42-45.
5. Dubey S, Joshi N, Stevenson O, et al. Chilblains in immune-mediated inflammatory diseases: a review. *Rheumatology* 2022; 61(12): 4631-4642.
6. Cribier B, Djeridi N, Peltre B, et al. A histologic and immunohistochemical study of chilblains. *J Am Acad Dermatol* 2001; 45(6): 924-929.
7. König N, Fiehn C, Wolf C, et al. Familial chilblain lupus due to a gain-of-function mutation in STING. *Ann Rheum Dis* 2016; 76(2): 468-472.
8. Lee-Kirsch MA, Gong M, Schulz H, et al. Familial chilblain lupus, a monogenic form of cutaneous lupus erythematosus, maps to chromosome 3p. *Am J Hum Genet* 2006; 79(4): 731-737.
9. Fiehn C. Familial chilblain lupus—what can we learn from type I interferonopathies?. *Curr Rheumatol Rep* 2017; 19: 61.
10. Günther C, Meurer M, Stein A, et al. Familial chilblain lupus—a monogenic form of cutaneous lupus erythematosus due to a heterozygous mutation in TREX1. *Dermatology* 2009; 219(2): 162-166.
11. Günther C, Berndt N, Wolf C, et al. Familial chilblain lupus due to a novel mutation in the exonuclease III domain of 3' repair exonuclease 1 (TREX1). *JAMA Dermatol* 2015; 151(4): 426-431.
12. Ali MSM. LP-204 chilblain lupus erythematosus—a rare encounter. *Lupus Sci Med* 2023; 10(suppl 1): A1-A171.
13. Wenzel J, Brähler S, Bauer R, et al. Efficacy and safety of methotrexate in recalcitrant cutaneous lupus erythematosus: results of a retrospective study in 43 patients. *Br J Dermatol* 2005; 153(1): 157-162.



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