

Short Report

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Chilblain lupus erythematosus- A rare encounter

*Corresponding Author: **Mohamed Shafi Bin**

Mahboob Ali

Advanced Medical and Dental Institute (AMDI),
Universiti Sains Malaysia.

Email: mshafix_7@yahoo.co.uk

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Abstract

Chilblain lupus erythematosus is a rare, chronic variant of cutaneous lupus erythematosus that occurs during cold or damp periods on the hands, fingers, or feet. It is often associated with other forms of cutaneous lupus. Like idiopathic chilblains such as perniosis, CHLE presents with tender, reddish-blue papules, nodules, or plaques on the toes, fingers, nose, or ears precipitated by cold exposure. The prevalence of the disease is about 3-20%; moreover, it occurs more frequently in women. Purpuric erythematous-violet plaques characterize it in the distal regions, such as the ears, nose, fingers and toes. It has a symmetrical distribution, usually edematous, itchy and sometimes painful lesions. They generally occur as a result of exposure to cold. Chilblain lupus is defined by the Mayo Clinic criteria, including two major and four minor. Histologic features are identical to those of discoid lupus erythematosus. The damaged skin gives a positive fluorescent band test. Various medications have been tried, including steroids, mycophenolate mofetil, calcium channel blockers, and hydroxychloroquine, but the symptoms do not remit completely. Managing chilblain lupus erythematosus is still unclear, and more research needs to be conducted. Treatment aims are to reduce disease activity and damage control. The first-line treatment for mild and localised cutaneous lupus erythematosus is topical corticosteroids, while the second-line systemic treatments consist mainly of immunomodulators and immunosuppressants. Clinicians should recognise the importance of early diagnosis and prompt treatment initiation to reduce associated morbidity and possible disfigurement of the patients.

Introduction

Chilblain lupus erythematosus (CHLE) or Perniosis is a rare and chronic form of lupus involving the toes, fingers, nose, and ears precipitated by cold exposure. The prevalence is 3-20%, affecting mostly women and can be divided into primary and secondary. The primary or idiopathic form is not associated with an underlying disease, while the secondary form is associated with an underlying condition such as connective tissue disease, monoclonal gammopathy, cryoglobulinemia, or chronic myelomonocytic leukaemia. It is often associated with other forms of cutaneous lupus, and about 20% of patients develop systemic lupus erythematosus (SLE). The patient usually comes with symptoms of purple plaques or nodules and oedematous skin, mainly around the acral regions of the body. Histologic features

are identical to those of discoid lupus erythematosus. The damaged skin gives a Positive fluorescent band test picture. CHLE is defined by the Mayo Clinic criteria, which include two major and four minor. Two major and at least one minor criterion are required to diagnose a patient. Patients with chilblain lupus erythematosus may also display hypergammaglobulinaemia, positive rheumatoid factor, antinuclear antibody, antiphospholipid or anti-Ro antibodies. They are usually negative for anti-double-stranded DNA antibodies. The first-line treatment for mild and localised symptoms is topical corticosteroids. Second-line systemic treatments consist mainly of immunomodulators and immunosuppressants. Studies have shown benefits from the use of topical tacrolimus and pimecrolimus. We want to report a case of a young lady that presented to our centre with CHLE.



Figure 1: Patient's hand showing red and dusky purple plaques after cold exposure.

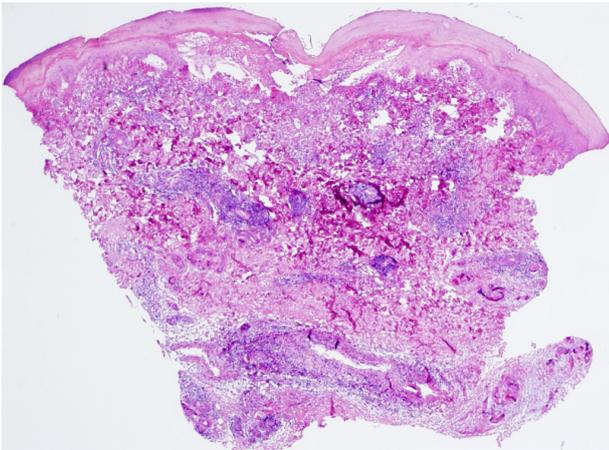


Figure 2: Histology slide depicting vacuolar interface dermatitis, perivascular lymphocytic infiltration and papillary oedema.

Conclusion

Chilblain lupus erythematosus is a rare and chronic disease mainly affecting women. Although it is not as severe as Systemic Lupus Erythematosus (SLE), it may be the sentinel sign of a range of underlying auto-immune diseases. Physicians should be vigilant in dealing with CHLE as their symptoms may be subtle and mimic other similar pathologies.

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