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Chilblain Lupus Erythematosus in an Adult Male

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INTRODUCTION

- Chilblain Lupus Erythematosus (CHLE) is a rare form of lupus seen in middle-aged females. Also known as “Hutchinson lupus” after Jonathan Hutchinson (1888).
- Pathogenesis not well understood-- vasoconstriction or microvascular injury secondary to exposure to cold? (Contrary to traditional systemic lupus erythematosus (SLE) in which skin lesions are exacerbated by sun exposure).
- CHLE has been found to progress to SLE in 20% of cases.
- The lesions in CHLE are painful and pruritic violaceous plaques on the palms, soles, and rarely on the ears, nose, and trunk.
- CHLE lesions are similar to those found in lupus pernio, erythema nodosum, cutaneous lesions after embolisation, and idiopathic chilblains. Consequently, Su et al. have suggested the “Mayo Clinic Diagnostic Criteria”:

Major Criteria:

- skin lesions in acral locations induced by exposure to cold
- evidence of SLE in skin lesions by histopathologic examination or indirect immunofluorescence

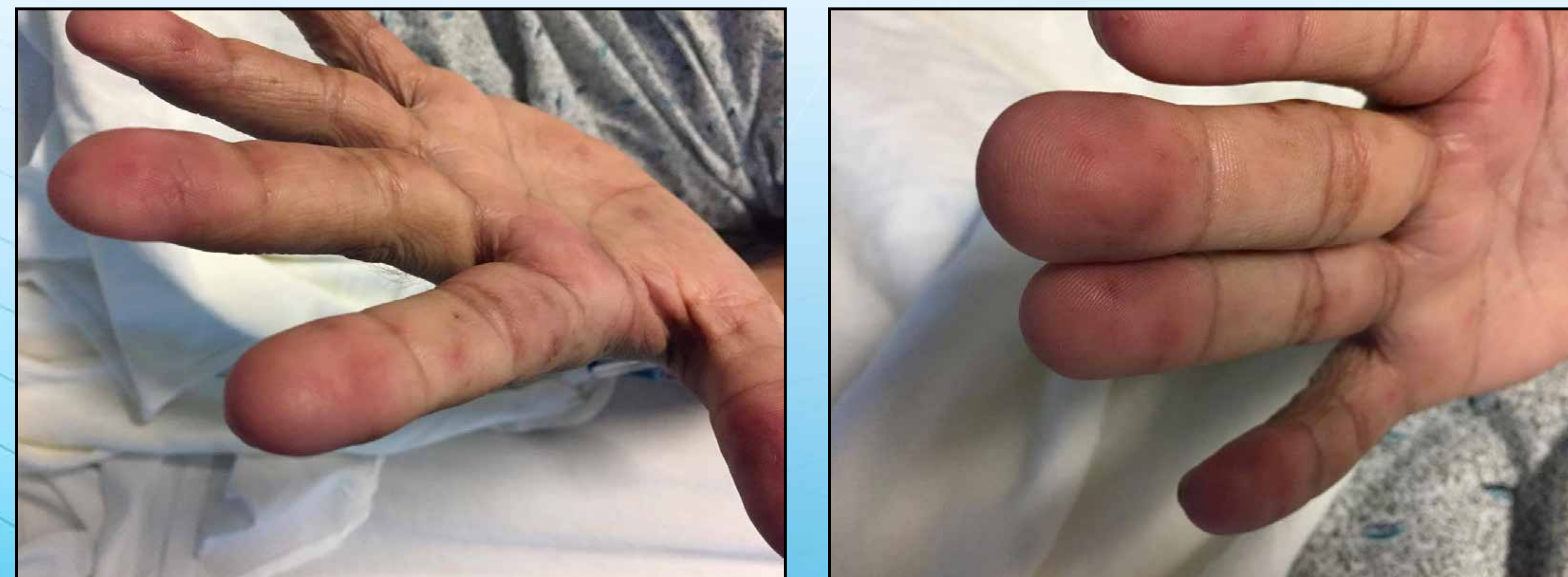
Minor Criteria:

- the coexistence of SLE or lesions consistent with Discoid LE
- response to anti-SLE therapy
- negative results of testing for cryoglobulin and cold agglutinins

Diagnosis of CHLE is confirmed if a patient fulfills both the major and any one of the minor criteria.

CASE PRESENTATION

- A 40 year-old Hispanic male presented in November with arthralgias, night sweats, fevers, weight loss, and violaceous skin lesions in an acral distribution. Medical history included arthritis and a penile ulcer that developed two months ago.
- He was urged to go to the ED after an outpatient CBC revealed pancytopenia.
- A CT scan in the ED showed widespread lymphadenopathy. The patient was admitted and a lymph node biopsy was performed; however, it showed no signs of lymphoma.
- Extensive infectious workup was unrevealing, and penile ulcer biopsy was negative for organisms.
- Biopsy of a hand lesion showed interface change with a lympho-plasmacytic infiltrate.
- Anti-dsDNA was negative, but ANA was highly elevated at 1:2,560 with a speckled pattern. Anti-Smith was also markedly elevated, and complement levels were low.
- The patient was diagnosed with SLE in combination with CHLE.
- Prednisone and hydroxychloroquine were initiated after confirming that G6PD was normal. The patient’s symptoms and skin lesions improved on this regimen.



DISCUSSION

- Our patient was not the typical age or gender for CHLE; however, he met both of the major and all of the minor criteria.
- Treatment of CHLE consists of avoidance of cold weather and treatment with antibiotics if lesions become secondarily infected. In addition, topical steroids and calcium channel blockers have been shown to be effective in treating CHLE in 50% of cases.
- Antimalarial agents (chloroquine or hydroxychloroquine) are effective in treating SLE symptoms; however, their effect on CHLE has been debated.
- In refractory cases, some reports have shown success with mycophenolate mofetil and full-thickness skin grafts (from an unaffected region).

LEARNING POINTS

- Chilblain Lupus Erythematosus should be suspected when any patient presents with violaceous, acral skin lesions that are exacerbated by cold weather and are associated with other nonspecific systemic symptoms.
- If left untreated, the disease can progress to SLE and can lead to complications of kidney failure, pericarditis, vasculitis, increased risk of infections, and worse overall prognosis.
- Making the diagnosis of CHLE can be aided by reference to the Mayo Clinic Diagnostic Criteria.

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