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The Clinical Picture

An erythematous plaque on the nose

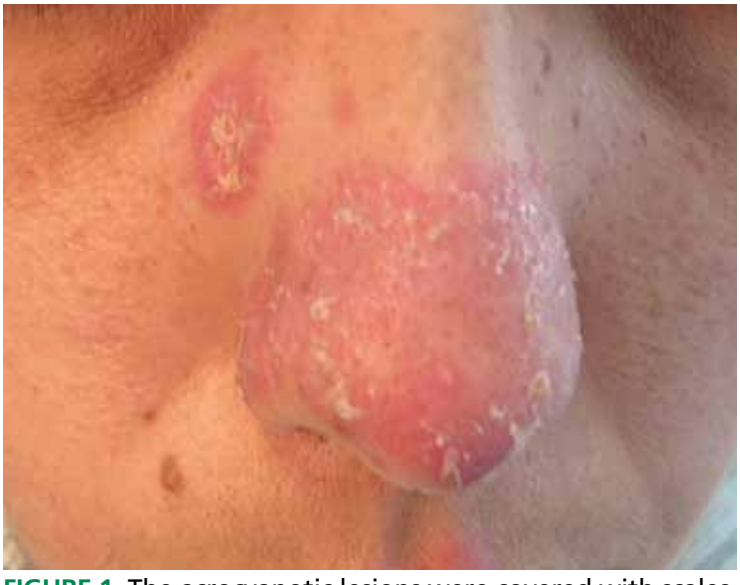

FIGURE 1. The acrocyanotic lesions were covered with scales.

A 38-year-old woman presented with a pruriginous and erythematous lesion on her nose that appeared during periods of cold weather. She said she is completely asymptomatic during the summer months.

A physical examination revealed acrocyanotic lesions on the nose that were covered with scales (FIGURE 1). Laboratory testing showed increased cholesterol levels, a positive antinuclear antibody titer (1:160 or higher is positive), and a positive anti-Ro/SS-A antibody titer (1:80 or higher is positive). Tests for cryoglobulin, cold agglutinins, anti-double-stranded DNA antibody, anti-extractable nuclear antigens, C3 and C4 complement proteins, and anticardiolipin antibody were normal or negative.

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Histologic examination revealed degeneration of the basal layer of the dermis, with periadnexal and perivascular inflammatory infiltrates (FIGURE 2). On immunofluorescence testing, linear deposits of immunoglobulin M were noted at the dermoepidermal junction.

Q: What is the most likely diagnosis?

- ☐ Lupus pernio
- ☐ Rosacea
- ☐ Seborrheic dermatitis
- ☐ Chilblain lupus erythematosus
- ☐ Lupus vulgaris

A: The diagnosis is chilblain lupus erythematosus.

The differential diagnosis of an erythematous lesion on the nose of a middle-aged woman also includes rosacea, lupus pernio, lupus vulgaris, and seborrheic dermatitis. Some of these lesions are exacerbated by cold. Usually, the diagnosis is based on clinical findings, but in some cases histologic features on biopsy study confirm the diagnosis.

Lesions of **lupus pernio (sarcoidosis)** remain unaltered with changes in temperature, and biopsy study usually shows granulomas without caseous necrosis with little inflammatory infiltrate at the periphery.

Rosacea usually gets worse with heat and with alcohol consumption, although it can be exacerbated by cold. Biopsy study shows a nonspecific perivascular and perifollicular lymphohistiocytic infiltrate accompanied occasionally by multinucleated cells.

Seborrheic dermatitis is a papulosquamous disorder characterized by greasy scaling over

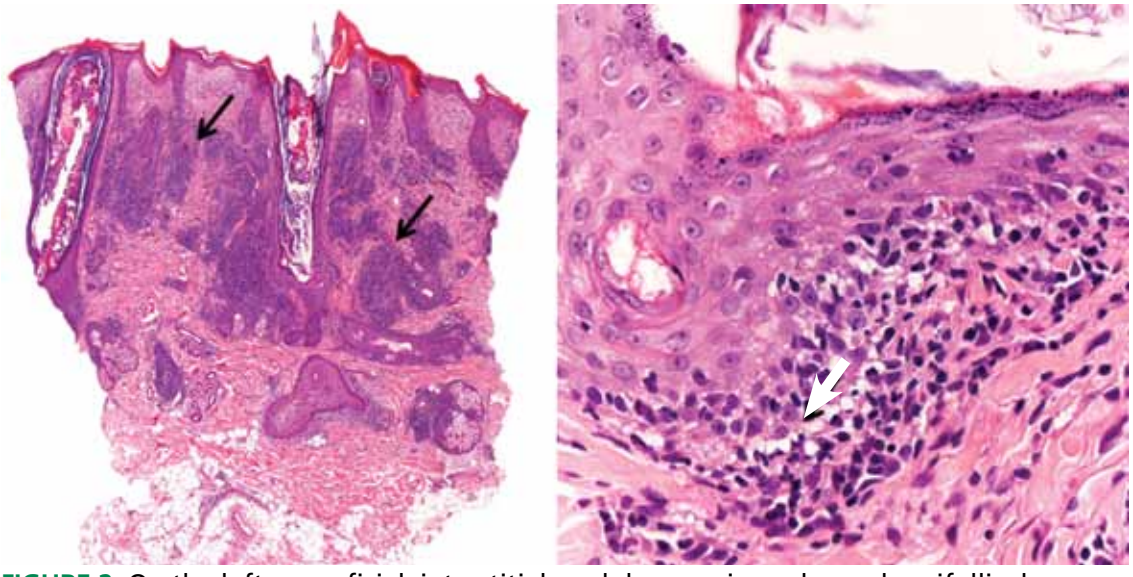

FIGURE 2. On the left, superficial, interstitial, and deep perivascular and perifollicular dense infiltrate of lymphocytes is seen (arrows) (hematoxylin-eosin, × 4). On the right, hydropic degeneration of the basal cell layer is seen (arrow) (hematoxylin-eosin, × 40).

inflamed skin on the scalp, face, and trunk. Disease activity is increased in winter and spring, with remissions commonly occurring in summer. The histologic features of seborrheic dermatitis are nonspecific; in this case, the histologic features were compatible with chilblain lupus without changes of seborrheic dermatitis.

Lupus vulgaris is a chronic form of cutaneous tuberculosis characterized by red-brown papules with central atrophy. The nose and ears are usually affected. Histologically, granulomatous tubercles with epithelioid cells and caseation necrosis are usually found.

CHILBLAIN LUPUS ERYTHEMATOSUS

Pernio, or chilblain, is a localized inflammatory lesion of the skin resulting from an abnormal response to cold.[1] The cutaneous lesions of chilblain may be classified as idiopathic, autoimmune-related (as in systemic lupus erythematosus, subacute cutaneous lupus), and induced by drugs such as terbinafine (Lamisil)[2] or infliximab (Remicade).[3]

Chilblain lupus is a rare form of cutaneous lupus erythematosus and should not be confused with lupus pernio, which is a misleading name used for a type of cutaneous sarcoidosis.[4]

Chilblain lupus is characterized by reddish-purple plaques in acral areas (more often the hands and feet, but also the nose and ears) that are induced by exposure to cold—unlike other lesions of lupus erythematosus, which worsen with exposure to sunlight. The main difference from the cutaneous variety of sarcoidosis (lupus pernio) is the histopathologic appearance. In patients with chilblain lupus, epidermal atrophy, perivascular and periadnexal inflammatory infiltrates, and degeneration of the basal layer are found, whereas in lupus pernio (sarcoidosis), we observe granulomas without caseous necrosis, but with few inflammatory infiltrates on the periphery.

Chilblain lupus should not be confused with lupus pernio, a misleading name for a type of cutaneous sarcoidosis

PROPOSED DIAGNOSTIC CRITERIA

Su et al[5] have proposed diagnostic criteria for chilblain lupus. Their two major criteria are 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 immunofluorescence study. Both of these criteria must be met, plus one of three minor criteria: the coexistence of systemic lupus erythematosus or of skin lesions of discoid lupus erythematosus; response to lupus therapy; and negative results of testing for cryoglobulin and cold agglutinins.

CHILBLAIN LUPUS VS SYSTEMIC LUPUS

Chilblain lupus is an uncommon manifestation of systemic lupus erythematosus, and it is reported to occur in about 20% of patients with that condition.[6] Often, the onset of chilblain lupus precedes the systemic disease. Patients with systemic lupus erythematosus and chilblain lupus do not usually present with renal disease, mucosal lesions, or central nervous system involvement. However, Raynaud phenomenon and photosensitivity have been reported to be more frequently associated with chilblain lupus.[7]

A disorder of peripheral circulation could be involved in the pathogenesis of chilblain lupus, and the association with Raynaud phenomenon, livedo reticularis, antiphospholipid syndrome, and changes in nailfold capillaries supports this hypothesis. Antinuclear antibody and anti-Ro/SS-A antibody are commonly detected in the serum of patients with chilblain lupus, and anti-Ro/SS-A antibody seems to be a major serologic marker of chilblain lupus in patients with systemic lupus erythematosus.[7]

TREATMENT

Protection from cold by physical measures is very important, as well as the use of topical or oral antibiotics if the lesions are infected. In severe cases unresponsive to topical corticosteroids, a calcium channel blocker is a good therapeutic option; antimalarials, commonly used in the treatment of lupus erythematosus, can also have a positive effect in patients with chilblain lupus.

CASE CONCLUDED

Our patient was advised to protect herself from the cold. Topical corticosteroids and oral hydroxychloroquine (200 mg/day) were prescribed, and they produced a good response. In severe cases, oral corticosteroids, etretinate (Tegison), mycophenolate (CellCept), or thalidomide (Thalomid) may be used.[8]

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