Chronic Erythematous Plaques Around the Ears

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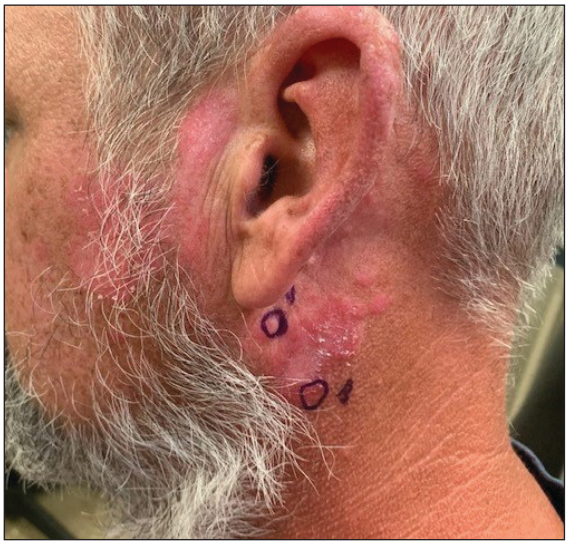

A 41-year-old man presented to the dermatology clinic with erythematous, pruritic, and painful plaques around the ears of 6 years' duration. He reported that application of topical steroids, antifungals, and most recently a topical calcineurin inhibitor did not change the appearance or symptoms. His medical history was notable for tobacco smoking and gastroesophageal reflux disease, for which he was taking omeprazole for the last 3 years. He was unsure if the lesions changed with UV exposure. He was an active-duty US military service member, and his job required frequently working outdoors. A review of systems was otherwise unremarkable. Physical examination revealed annular, erythematous, indurated plaques on both the preauricular and postauricular skin on the left ear with associated central atrophy and hypopigmentation. No alopecia was appreciated. The remainder of the skin examination was unremarkable. Ancillary laboratory test results were notable for a negative antinuclear antibody screen but positive (low titer) for Sjögren syndrome A and B antibodies. Two punch biopsies were performed.

WHAT'S YOUR **DIAGNOSIS?**

a. annular psoriasis
b. cutaneous sarcoidosis
c. discoid lupus erythematosus
d. granuloma annulare
e. Majocchi granuloma

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THE **DIAGNOSIS:** Discoid Lupus Erythematosus

The biopsies demonstrated vacuolar interface changes with superficial and deep perivascular and periadnexal inflammation as well as increased background mucin deposition. The clinical morphology and distributions of the plaques limited to the photoexposed areas of the head suggested a diagnosis of discoid lupus erythematosus (DLE). The interface changes on histopathology supported this clinical impression. Our patient was treated with limited application of triamcinolone ointment 0.1% twice daily around the ears and neck, tacrolimus ointment 0.1% twice daily on the face, and hydroxychloroquine, as well as sun protection instructions. Smoking cessation was strongly advised.

Discoid lupus erythematosus is a disorder with chronic, erythematous, scaly, coin-shaped (discoid) plaques and is the most common form of chronic cutaneous lupus erythematosus.[1] Lesions usually present on sun-exposed areas of the face, scalp, neck, ears, lips, or upper torso. They expand slowly with an active peripheral margin and a central scar that can result in induration, pigmentation changes, telangiectases, pruritus, or tenderness. Hair-bearing areas may be involved, causing hair loss due to follicular plugging; irreversible scarring alopecia can result. Facial DLE often spares the nasolabial folds. Ear involvement characteristically includes the conchal bowl and the outer external auditory canal. Discoid lupus erythematosus is considered localized if most of the head and neck region is involved or generalized if lesions also are present below the neck. Risk factors for DLE include genetic and environmental factors such as UV exposure, hormones, or exposure to toxins such as cigarette smoke.[1] The disorder most commonly affects females and has a higher prevalence in patients of African descent than in Asian and White patients. Disease can occur at any age but usually occurs between 20 and 40 years of age.[2] Discoid lupus erythematosus and other forms of chronic cutaneous lupus can occur independently or in conjunction with systemic lupus erythematosus (SLE), and approximately 15% to 30% of SLE patients develop DLE.[1]

Discoid lupus erythematosus is clinically diagnosed by the presentation of plaques in the characteristic distribution with confirmation via skin biopsy.[1] Elman et al[3] created a system for DLE classification that was only clinical and did not involve histopathology. Histologically, DLE often includes basement membrane thickening, follicular keratin plugs, mucin deposition, and vacuolar change with an interface, and a perivascular and periadnexal lymphocytic infiltrate.[3,4] Antibodies such as antinuclear antibodies, rheumatoid factor, anti–double-stranded DNA, anti-Smith, and Sjögren syndrome A and B antibodies may be present (albeit with low positive frequency) in cutaneous lupus erythematosus.[4] Characteristics of SLE also may be present, helping to confirm the diagnosis. Because there is an association of DLE with SLE, various laboratory tests should be ordered, including complete blood cell count, renal function panel, inflammatory markers, antibodies, and urinalysis for proteinuria.[2,4]

Treatment of DLE consists of preventative measures, such as sun protection with vitamin D supplementation, avoidance of drug triggers, and smoking cessation, as well as pharmacotherapy. The importance of wearing sun protective hats and garments with sunscreen use cannot be understated.[1] Smoking cessation should be advised because smoking reduces the efficacy of antimalarial treatment and potentially increases the likelihood of patients requiring a second antimalarial drug. Quinacrine often is noted in both the dermatology and rheumatology literature to be used for escalating cutaneous lupus erythematosus care when hydroxychloroquine is ineffective or not tolerated, but no US manufacturer produces this medication; thus, compounding is required, which may be financially prohibitive, making this recommendation difficult to translate into clinical practice.[5] First-line therapy for acute flares is high-potency topical corticosteroids. If lesions are primarily on areas other than the face, a medium-potency topical steroid may be used. Topical calcineurin inhibitors or intralesional corticosteroids may be used if minimal improvement is seen after initial topical corticosteroid therapy. Treatment for widespread disease or disease that is resistant to local treatment is systemic therapy with antimalarial agents, followed by antimetabolites, systemic retinoids, thalidomide, or dapsone.[1,2] The Cutaneous Lupus Erythematosus Disease Area and Severity Index is a valid tool to gauge the degree of disease and to help with disease progression and treatment response by noting the features of the plaques.[1]

Patients also should be educated that this disease can last for years, and long-standing DLE plaques infrequently can give rise to squamous cell carcinoma. In addition, isolated DLE can progress to SLE in 5% to 28% of patients.[2]

The differential diagnosis in our patient included other diseases with violaceous annular lesions and central clearing. Majocchi granuloma usually presents in areas of prior trauma, possibly due to shaving the face in our patient, or in the setting of topical corticosteroid use or immunosuppression. Scaling often is present within lesions, and histology shows fungal elements.[6] Cutaneous sarcoidosis usually presents on the face, with scarring alopecia when appearing on the scalp; histology shows noncaseating granulomas, and 70% of patients with cutaneous symptoms will have systemic sarcoidosis.[7] Granuloma annulare

most commonly presents on the extremities, and histology shows lymphohistiocytic granulomas in a palisaded or interstitial pattern with connective-tissue degeneration and mucinous deposits.[8] Annular psoriasis often is scaly and symmetric with parakeratosis, epidermal hyperplasia, dilated dermal capillaries, loss of granular layer, perivascular mononuclear cell infiltrate, and elongation of rete ridges on histology.[9] Drug-induced lupus erythematosus always should be considered in patients taking triggering drugs such as antihypertensives, lipid-lowering drugs, antifungals, anti–tumor necrosis factor drugs, and proton pump inhibitors—the latter being a drug our patient was taking.[10]

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