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Refractory Subacute Cutaneous Lupus Erythematosus With a Response to Efalizumab

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To the Editor:

Subacute cutaneous lupus erythematosus is a specific form of lupus that occurs with annular or polycyclic erythematous plaques in areas exposed to sunlight.[1] Conventional treatments include the use of sunscreens, topical and oral corticosteroids, sulfones, antimalarial drugs, and immunosuppressive agents including cyclosporine, azathioprine, or methotrexate.[1]

We present the case of a 54-year-old woman—allergic to heparin, with a history of arterial hypertension, deep vein thrombosis in the lower right leg, and bilateral pulmonary thromboembolism resulting from an ankle fracture—who had been monitored for subacute cutaneous lupus erythematosus for 25 years. Since the initial diagnosis, the patient has presented continual outbreaks of cutaneous lesions consisting of annular erythematous plaques with a shiny surface and clear margin around the eyebrows, and on the cheeks and upper back (Figure 1). There were no associated systemic symptoms and tests for antinuclear antibodies (ANA) and anti-Ro antibodies were positive. The condition progressed with successive poorly controlled cutaneous outbreaks that were treated with medium to high potency topical corticosteroids, topical calcineurin inhibitors, oral prednisone (minimum of 15 mg on alternate days up to 60 mg daily), and systemic agents (hydroxychloroquine, azathioprine, methotrexate, and cyclosporine). All forms of treatment had been suspended due to ineffectiveness, poor tolerance, or adverse effects. As the daily dosage of 30 mg of prednisone could not be reduced, subcutaneous efalizumab 1 mg/kg/week was administered and the skin lesions resolved after 2 months (Figure 2). The patient was symptom free at the end of 4

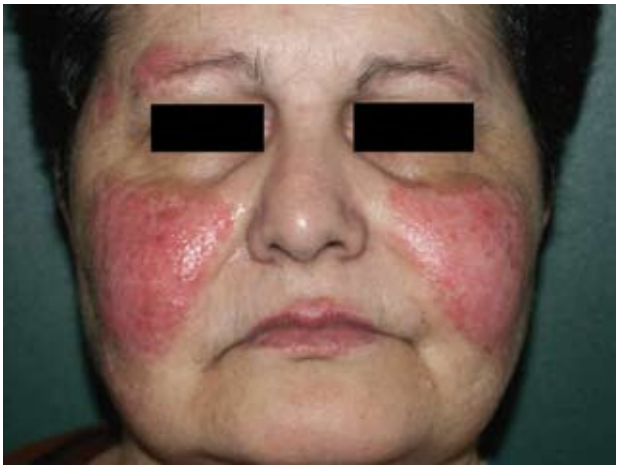

Figure 1. Clearly defined infiltrated erythematous plaques on the eyebrows, cheeks, and upper lip.

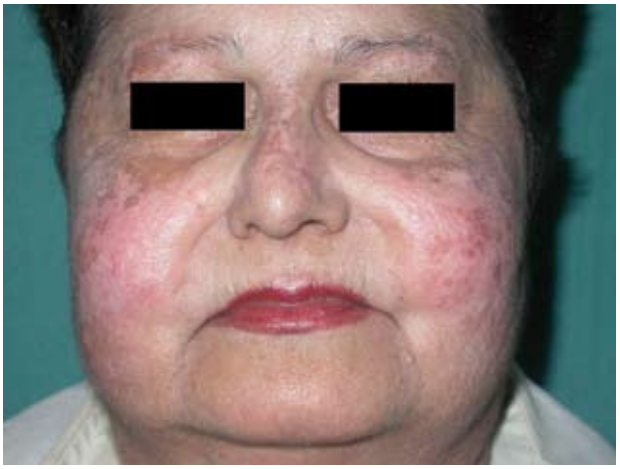

Figure 2. Resolution of the lesions after 2 months treatment with efalizumab.

months of follow-up during which the efalizumab dose was maintained while tapering the prednisone dose. She showed good tolerance to the treatment with no adverse effects, no changes in test outcomes, and a negative ANA titer. Efalizumab is a humanized monoclonal antibody that has proved effective in the treatment of inflammatory illnesses such as psoriasis[2] and lichen planus.[3] Its use has recently been proposed in the treatment of cutaneous lupus, on the assumption that the pathogenesis of this illness is mediated by T cells.[4,5] Clayton et al[4] described the case of a patient with refractory subacute cutaneous lupus who was treated with systemic efalizumab and who presented improvement of the lesions in 6 weeks with no adverse effects. Some 3 months later the patient presented an outbreak of lesions that was controlled by increasing the dose of efalizumab. Usmai and Goodfield[5] presented a series of 13 patients of discoid lupus refractory to multiple treatments in which 11 showed an excellent response to efalizumab. The mean response time was 5 weeks and the mean treatment duration was 14 months. However, concerns have been voiced about the safety of this treatment, and cases of efalizumab-induced lupus have even been reported.[6-8] Hamprecht et al[6] reported a woman with tumid lupus erythematosus and malar rash whose lesions failed to respond to systemic agents but resolved in 4 weeks with efalizumab combined with prednisolone. However, 6 weeks into treatment the patient developed lupus nephritis with increased antibodies that led to suspension of the efalizumab and an increased dose of corticosteroids. Bentley et al[7] described the case of a woman with erosive oral lichen planus who was being treated with efalizumab. She developed a lupus-like syndrome with discoid lesions in areas exposed to sunlight, positive antibodies, and histology compatible with lupus. The lesions healed in 8 weeks when treated with hydroxychloroquine. Durox et al[8] presented the case of a woman with psoriasis being treated with efalizumab who developed drug-induced lupus erythematosus. The condition resolved after the drug was suspended and treatment with corticosteroids and cyclosporine was initiated. We believe that efalizumab could be a useful drug in treating patients with cutaneous lupus erythematosus refractory to conventional therapies or when there is some contraindication for these. However, the fact that there are reports of efalizumab-induced elevated ANA and lupus-type syndrome means that, for the time being, this treatment should only be used in carefully selected cases with exhaustive clinical and laboratory follow-up. Randomized studies are needed with larger cohorts and long-term follow-up to evaluate safety and efficacy.

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Conflicts of Interest
The authors declare no conflicts of interest.

Addendum

On February 19, 2009, the Spanish Drug Agency (Agencia Española de Medicamentos y Productos Sanitarios [AEMPS]) informed health professionals that Raptiva (efalizumab) was withdrawn from the market following reports of 3 confirmed cases (2 fatal) and one probable case of progressive multifocal leukoencephalopathy in patients using efalizumab. In view of the risk-benefit relationship, some patients may benefit from future drugs with a similar action mechanism and reduced side effects.

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