Reticular Erythematous Mucinosis: Partial Response to Treatment With Topical Tacrolimus

Mucinosis eritematosa reticular: respuesta parcial al tratamiento con tacrolimus tópico

To the Editor:

Reticular erythematous mucinosis (REM) is a rare form of cutaneous mucinosis that is occasionally associated with systemic diseases and tumors.[1] Hydroxychloroquine is frequently used as the first-line treatment for this condition.[2] We present a case of REM treated with topical tacrolimus.

The patient was a 48-year-old man with a history of pulmonary tuberculosis treated during childhood, who consulted for the presence of a skin lesion that had appeared 12 years earlier on the trunk, showing slow and progressive growth and causing occasional pruritus. Dermatological examination revealed a clearly delimited reticulated erythematous plaque of 12 × 6 cm in the sternal region, with slight infiltration of the borders (Figure 1). There was a similar lesion of a smaller size in the interscapular region. Incisional biopsy and pathology study showed discrete vacuolar degeneration in the basal layer and a moderate perivascular and perifollicular lymphocytic infiltrate in the papillary and reticular dermis. Alcian blue stain revealed abundant mucin deposits in the papillary dermis. Diagnosis of REM prompted an extensive series of additional tests including: complete blood count, serum biochemistry, coagulation study, erythrocyte sedimentation rate, rheumatoid factor, ferritin, autoantibodies (antithyroid, antinuclear, and extractable nuclear antigens), complement levels, protein electrophoresis, immunoglobulins, thyroid function tests, plain chest x-ray, abdominal ultrasound, and serology (hepatitis, human immunodeficiency virus and syphilis). All the results obtained were within normal limits, except that subclinical hypothyroidism was detected (thryotropin [TSH]: 11.32 μU/ml [normal range: 0.2–5.0 μU/ml]; free T4: 1.2 ng/dL [normal range: 0.8–1.9 ng/dL]; antithyroid antibodies: negative). The patient refused treatment with oral hydroxychloroquine but accepted treatment with tacrolimus ointment 0.1% twice daily. At 3 months the REM showed clear improvement and treatment was suspended. The lesions were stable when revised 2 months later (Figure 2).

REM is an uncommon, chronic form of cutaneous mucinosis. Steigleder et al[3] first described the condition in 1974 and some 70 cases have been published since. Hydroxychloroquine is the first line treatment.[2] The condition has a certain degree of overlap with lupus erythematosus as they share a series of common elements including: predominance in women, exacerbation after exposure to sunlight, clinical manifestations, mucin deposits, granular IgM deposits along the dermal-epidermal junction, and response to antimalarial drugs.[2-5] REM affects patients of both sexes, but most cases occur in middle-aged women.[3,5] The rash is usually asymptomatic, but pruritus occurs in approximately 30% of cases. Lesions are typically located on the central region of the thorax and the upper central region of the back; they occur less frequently on the face, arms, and abdomen. Two clinical forms have been described: infiltrated plaques and erythematous reticulated macules. Telangiectasia is observed on rare occasions.[1] Deterioration has been reported with exposure to sunlight, menstruation, pregnancy, and the use of oral contraceptives.[3,4] The cause is unknown, but sensitivity to light and immunological and viral mechanisms have been implicated.[1] Pathology findings are characterized by the presence of edema, a perivascular and periadnexal mononuclear infiltrate, and mucin deposits in the mid-dermis and papillary dermis.[2] In some cases there is slight degeneration of the basal layer.[4] REM has been associated with thyroid disorders, diabetes mellitus, lupus erythematosus, neoplasms, idiopathic thrombocytopenic purpura, myopathy, polyneuropathy, and human immunodeficiency virus infection.[1-5] In our patient we were able to detect a subclinical abnormality of thyroid function.

Antimalarial drugs are the treatment of choice for REM. They produce a rapid clinical improvement, often within a month of starting treatment; however, recurrence is common. Hydroxychloroquine at a dose of 200 to 400 mg/day has been effective in treating this condition.[1,2,4] In patients with no response or in whom this drug is contraindicated, other therapeutic options that have been effective include

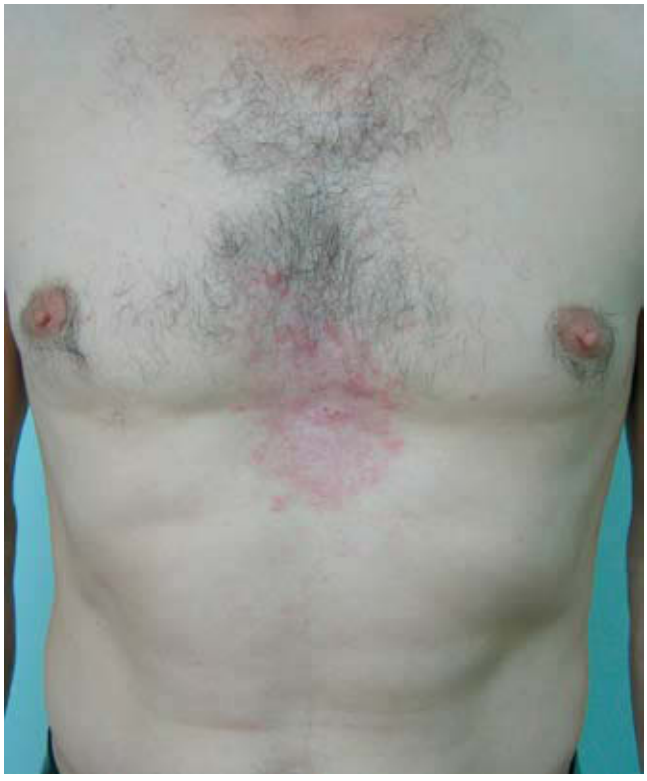

Figure 1 Reticulated erythematous plaque on the sternal region prior to treatment.

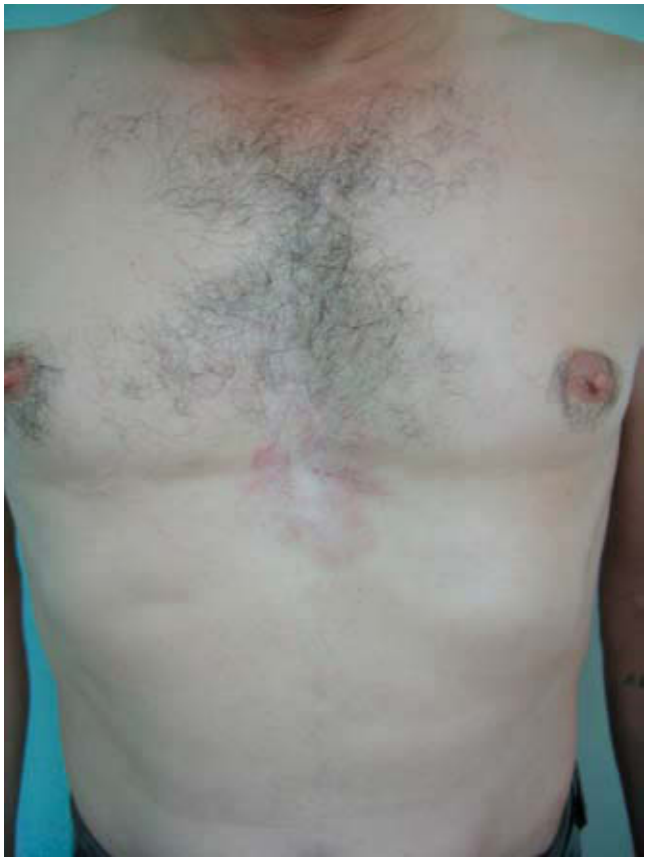

Figure 2 Appearance of the lesion 2 months after treatment with tacrolimus.

colchicine,[3] topical corticosteroids associated with UV-B,[6] phototherapy with UV-A1,[7] and pulsed dye laser.[8,9] In the English-language medical literature (bibliographical search on PubMed) there are 2 articles relating to the use of topical calcineurin inhibitors in REM.[9,10] In 1 case, pimecrolimus cream 1% was applied twice daily for 5 months with almost complete resolution of the lesions.[9] In another patient, a vast improvement was achieved through treatment with 0.1% tacrolimus ointment twice daily for 2 weeks and then 0.03% twice daily for 2 months.[10] The efficacy of tacrolimus in chronic inflammatory diseases like REM is believed to be due to its potent immunosuppressant effect on T-cells by blocking the action of calcineurin.[10] As our patient refused oral treatment with hydroxychloroquine, we decided to use topical 0.1% tacrolimus twice daily, which produced a substantial improvement in the lesions after 3 months of treatment.

References

1. Leon-Mateos A, Ginarte M, Leon L, Toribio J. Reticular erythematous mucinosis (REM) with telangiectasias associated with essential thrombocytosis and lung carcinoma. Eur J Dermatol. 2005; 15:179-81.
2. Woollons A, Darley CR. Erythematous rash on the chest. Arch Dermatol. 2002; 138:1245-50.
3. Serrano A, Rodríguez L, Conejo-Mir J. Mucinosis eritematosa reticular. Actas Dermosifiliogr. 2004; 95:317-9.
4. Sidwell RU, Francis N, Bunker CB. Hormonal influence on reticular erythematous mucinosis. Br J Dermatol. 2001; 144:633-4.
5. Gasior-Chrzan B, Husebekk A. Reticular erythematous mucinosis syndrome: Report of a case with positive immunofluorescence. J Eur Acad Dermatol Venereol. 2004; 18:375-8.
6. Yamazaki S, Katayama I, Kurumaji Y, Yodozeki H, Nishioka K. Treatment of reticular erythematous mucinosis with a large dose of ultraviolet B radiation and steroid impregnated tape. J Dermatol. 1999; 26:115-8.
7. Meewes C, Henrich A, Krieg T, Hunzelmann N. Treatment of reticular erythematous mucinosis with UV-A1 radiation. Arch Dermatol. 2004; 140:660-2.
8. Greve B, Raulin C. Treating REM syndrome with the pulsed dye laser. Lasers Surg Med. 2001; 29:248-51.
9. Mansouri P, Farshi S, Nahavandi A, Safaie-Naraghi Z. Pimecrolimus 1 percent cream and pulsed dye laser in treatment of a patient with reticular erythematous mucinosis syndrome. Dermatol Online J. 2007; 13:22.
10. Rubegni P, Sbano P, Risulo M, Poggiali S, Fimiani M. A case of reticular erythematous mucinosis treated with topical tacrolimus. Br J Dermatol. 2004; 150:173-4.

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