

# Reticular Erythematous Mucinosi s on the Midline of the Back

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## Practice Points

- Reticular erythematous mucinosi s is a midline mucinosi s that presents with reticular macular lesions or erythematous indurations that typically are localized on the anterior and posterior aspects of the trunk.
- Reticular erythematous mucinosi s can be triggered by sun exposure and also can be associated with lupus erythematosus.

*We report the case of a 51-year-old man who presented with reticular macular erythema and erythematous plaques on the midline of the back of 10 years' duration that seemed to worsen on exposure to sunlight. Based on the clinical and histopathologic findings, a diagnosis of reticular erythematous mucinosi s (REM) was made. The patient was treated with hydroxychloroquine 400 mg daily and the lesions resolved by the end of the second week of therapy.*

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## Case Report

A 51-year-old man presented to our outpatient clinic with reticular macular erythema and erythematous plaques on the midline of the back of 10 years' duration (Figure 1). The patient noted that the lesions seemed to worsen on exposure to sunlight. He used high-potency topical corticosteroids for 1 month prior to presentation but only a minimal response was observed and he discontinued the treatment. Physical examination was otherwise unremarkable. No lymphadenopathy or organomegaly was noted. The results of laboratory examination including a complete blood cell count, liver and renal function tests, and erythrocyte

sedimentation rate were within reference range, and screening for hepatitis B and C as well as human immunodeficiency virus infection was negative. Antinuclear antibody titers were less than 1:100 (reference range, <1:160). There was no family history of similar cutaneous lesions. Hematoxylin and eosin staining of biopsy specimens from lesional skin revealed a periadnexal lymphocytic cell infiltrate (Figure 2A). Separation of dermal collagen bundles with homogenous basophilic material in the upper dermis was exhibited. Alcian blue pH 2.5 staining revealed positive mucin deposits in the upper dermis (Figure 2B). The epidermis appeared normal.

Based on the clinical and histopathologic findings, a diagnosis of reticular erythematous mucinosi s (REM) was made. Following a normal ophthalmologic examination the patient was treated with hydroxychloroquine 400 mg daily for 3 months. The skin lesions resolved by the end of the second week of therapy (Figure 3).

## Comment

Cutaneous mucinosi s is a heterogeneous disease in which mucin accumulates in the skin.<sup>1</sup> Reticular erythematous mucinosi s and the plaquelike form of cutaneous mucinosi s are classified as midline mucinoses.<sup>2</sup> Reticular erythematous mucinosi s has been documented more frequently in women than in men; it rarely arises in children younger than 10 years or in adults older than 60 years.<sup>3</sup> It typically presents as a midline mucinosi s with a predilection for the anterior and posterior aspect of the trunk.<sup>2,4</sup> The arms, face, and abdomen are less commonly affected.<sup>5</sup> Reticular erythematous mucinosi s presents with reticular macular lesions or erythematous indurations, papules, and plaques.<sup>3,5,6</sup> Affected patients frequently are asymptomatic; however, pruritus

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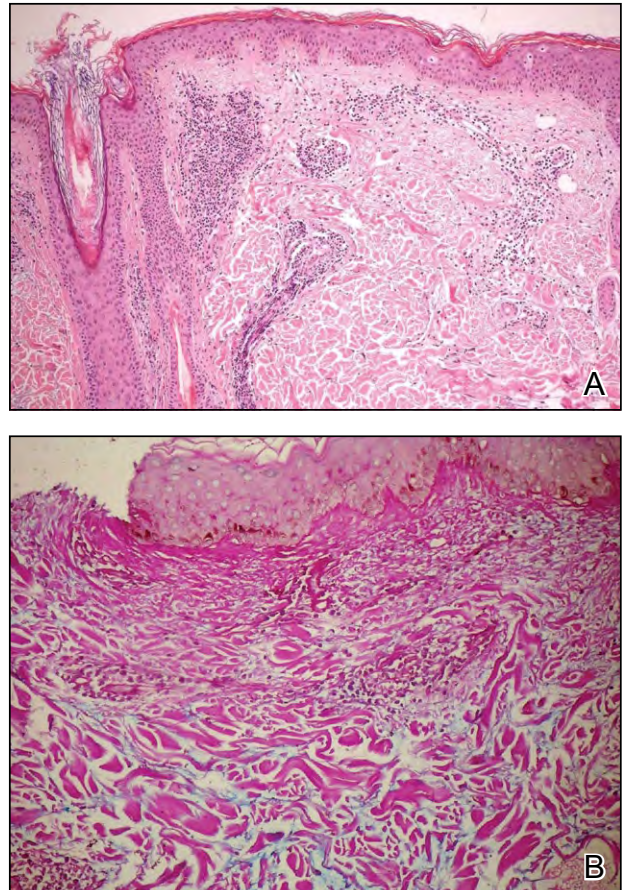
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**Figure 1.** Reticular macular erythema and erythematous plaques on the midline of the back (A and B).

is reported in 20% to 30% of cases.<sup>3</sup> Our patient presented with mild pruritus on the back.

Histopathologic findings predominantly include expanded vessels, perivascular and occasional perifollicular round-cell infiltration, and Alcian blue–positive mucin (glycosaminoglycans) depositions between the collagen fibers of the upper dermis. The epidermis generally appears normal.<sup>3-5,7</sup> Direct immunofluorescence usually is negative for immunoglobulins, fibrin, and



**Figure 2.** Histopathology revealed a periadnexal lymphocytic cell infiltrate (A)(H&E, original magnification  $\times 100$ ). Positive mucin deposits were noted in the upper dermis (B)(Alcian blue pH 2.5, original magnification  $\times 40$ ).

complement; however, a few cases of REM with positive direct immunofluorescence and granular deposits of IgM and C3 have been reported.<sup>3,8,9</sup> In REM skin lesions, mucin deposits mainly consist of hyaluronic acid, which is clear on analysis of glycosaminoglycan content.<sup>10</sup> Mucin synthesis is modulated by different cytokines, including transforming growth factor  $\beta$ , interleukins, tumor necrosis factor, and interferon.<sup>3</sup>

The etiology of REM remains unclear.<sup>3,5,6</sup> Some investigators consider REM to be a part of the lupus erythematosus–like disease spectrum due to its clinical findings, mucin deposition on histologic examination, progression after exposure to UV radiation, and good response to antimalarial drugs.<sup>3,6</sup> Del Pozo et al<sup>11</sup> reported a case of REM that developed into systemic lupus erythematosus after 6 years. In 20% of patients with REM, there is an association with autoimmune diseases such as discoid lupus erythematosus, idiopathic thrombocytopenic purpura, and diabetes mellitus.<sup>2</sup> Associations with malignancy, thyroid disease, monoclonal IgG  $\kappa$  paraproteinemia, tumors, and myopathy



**Figure 3.** Skin lesions regressed with hydroxychloroquine therapy.

also have been reported.<sup>3,5,6</sup> None of these associated factors existed in our patient.

The clinical course generally is cyclic, with alternating periods of remission and exacerbation.<sup>3</sup> Several factors that have been known to exacerbate REM include viral processes, immunologic disturbances, solar irradiation, menstruation, pregnancy, and radiography.<sup>5</sup> In our patient, exposure to sunlight seemed to aggravate his condition.

Treatment options for REM include antimalarials; topical and systemic corticosteroids, sometimes in combination with UVB irradiation; tacrolimus; pimecrolimus; and cyclosporine.<sup>3,5,6</sup> Greve and Raulin<sup>12</sup> reported successful treatment of REM in 2 patients with a pulsed dye laser as an alternative method due to its efficacy in treating vascular skin diseases. Antimalarial therapy has been reported to be the most effective treatment, resulting in prompt clinical improvement within a month or less in the majority of patients.<sup>5</sup> The mechanism of action for antimalarials is not well known. These drugs can inhibit the release of IL-2 from helper T cells (CD4<sup>+</sup>) and may inhibit macrophage expression of major histocompatibility complex antigens. It is known that hydroxychloroquine 200 to 400 mg daily is the treatment of choice. Relapses often occur.<sup>3</sup> Because of the dose-related toxicity, a referral

to an ophthalmologist is necessary for examination at baseline with follow-up every 6 to 12 months thereafter. Laboratory abnormalities do not commonly occur.<sup>13</sup>

## Conclusion

Our case is important due to the rare presentation of REM in men. Photosensitivity should be kept in mind as a feature of REM. Antimalarial agents are still considered the first-line therapy for REM and have been associated with substantial improvement of skin lesions in patients with REM.

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