Treatment of Reticular Erythematous Mucinosis With UV-A1 Radiation

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RETICULAR ERYTHEMATOUS MUCINOsis (REM) is a rare chronic dermatosis that is typically characterized by reticular macular erythema or erythematous papules and plaques on the central area of the chest and back of middle-aged women. The pathogenesis and etiology of REM remain undefined at present. Several factors have been associated with the induction of the syndrome, including viral processes, immunological disturbances, and solar irradiation.1-8

REPORT OF CASE

A 44-year-old woman presented to our outpatient clinic with a 4-year history of symmetrical netlike macular erythema and erythematous papules on the central area of her chest (Figure 1). She stated that exposure to light did not seem to aggravate her condition. Her medical history was unremarkable except for a goiter, and laboratory tests revealed normal plasma levels of thyroxine and thyrotropin. Her family history was not contributory. Thyroid hormone, antinuclear antibody, IgA, IgG, IgM, and serum complement levels were normal. Antibodies to native (single- and double-stranded) DNA were absent. Hematoxylin-eosin staining of biopsy specimens of lesional skin revealed a perivascular and perifollicular lymphomononuclear cell infiltrate. Alcian blue staining revealed mucin deposits in the upper dermis and a basement membrane of normal thickness. Direct immunofluorescence examination was negative for immunoglobulin depositions and slightly positive for complement (C3) depositions. On the basis of the clinical picture and the histologic findings, REM was diagnosed.

THERAPEUTIC CHALLENGE

Successful therapy of REM with chloroquine has been reported.1-12 Other treatment alternatives (UV-B, laser, glucocorticoids, cyclosporine) have had variable efficacy.1-12 Since treatment with chloroquine was refused by the patient, another therapeutic option was needed. Therefore, UV-A1 (340-400 nm) therapy was considered.

SOLUTION

To exclude photosensitivity, photoprovocation testing was performed with UV-A and UV-B on the patient’s normal back skin using a commercially available phototesting device (SBB LT 400 Multitester; Saalman GmbH, Herford, Germany). The minimal erythema dose was determined by applying graded doses of UV-A (1-8 J/cm²) and UV-B (10-80 mJ/cm²). It was defined as the lowest irradiation dose necessary to produce minimal perceptive erythema of the test site at 20 minutes (UV-A) and 24 hours (UV-B) after irradiation. Also, UV-A (60 J/cm²) and UV-B (90 mJ/cm²) irradiation was applied for a photoprovocation test on the next 3 days, and the irradiated
sites were checked for skin changes at days 4 and 5 and weeks 1, 3, and 6 after the first application. After no abnormal response was observed, UV-A1 irradiation of the lesional skin of the chest was started at a dose of 40 J/cm$^{-2}$. Irradiations were performed 5 days a week, and the doses were increased by 10 J/cm$^{-2}$ daily to a maximum dose of 90 J/cm$^{-2}$. After 18 UV-A1 irradiations and a cumulative dose of 1210 J/cm$^{-2}$, the patient's lesions completely cleared (Figure 2). Her skin remained clear for 10 months. A minor recurrence was treated with UV-A1 at doses of up to 90 J/cm$^{-2}$. After 16 additional irradiations (cumulative dose, 1420 J/cm$^{-2}$), the lesions again completely cleared. No further recurrence was observed until December 2002.

Initially reported as a “plaque-like [form of] cutaneous mucinosis (PCM)” and REM syndrome, these conditions were recognized as part of the spectrum of the same disorder. The cutaneous lesions range from erythematous, indurated papules and plaques to netlike macular erythema. The central chest and/or upper back area is a preferentially involved location. The arms, face, and abdomen are less frequently affected. Predominant histopathologic findings include expanded vessels, perivascular and perifollicular round cell infiltration, and (acian blue–positive) mucin depositions between the collagen fibers of the upper dermis. Anti-malarial therapy has reportedly been the most effective treatment, resulting in prompt clinical improvement within a month or less in the majority of patients. Other treatment regimens include topical and systemic corticosteroids, sometimes in combination with UV-B irradiation and cyclosporine. In approximately 20% of cases, REM may be associated with a variety of systemic disorders, especially autoimmune diseases such as discoid lupus erythematosus, idiopathic thrombocytopenic purpura, and diabetes mellitus, but also with malignancies and thyroid diseases. Other factors that have been observed to promote exacerbation of REM include menstruation, pregnancy, heat, x-ray therapy, and excitement. Sun exposure has also been reported to induce new skin lesions and to worsen the condition in several patients. Interestingly, an abnormal response was not observed in most patients when they were phototested with UV-A and UV-B irradiation.

In REM skin lesions, mucin deposits consist mainly of hyaluronic acid, as has clearly been shown by analysis of the glycosaminoglycan content. Ultraviolet A1, directly or indirectly, via induction of proinflammatory cytokines such as interleukin 1-β, can induce matrix-degrading enzymes such as proteoglycanase in dermal fibroblasts, which may lead to an increased degradation of hyaluronic acid depositions. In a similar way, these cytokines can result in a down-regulation of hyaluronic acid synthesis in REM fibroblasts. Moreover, reactive oxygen species such as singlet oxygen, superoxide anion, and the hydroxyl radical—as they all are produced on UV (especially UV-A) irradiation—have been shown to lead to fragmentation and damage of hyaluronic acid.

Because of their different wavelengths, UV-A, and particularly UV-A1, is able to permeate deeper into the dermis than UV-B. This physical quality might explain why UV-B irradiation, even in large doses, was an ineffective monotherapy in this case, while UVA-1 treatment led to a complete resolution of skin changes.

In summary, UV-A1 therapy represents an effective alternative in the treatment of REM.

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REFERENCES


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