

Positive photobiological investigation in reticular erythematous mucinosis syndrome

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Background: Reticular erythematous mucinosis (REM) syndrome is a rare disorder. Its clinical course is cyclic with remissions and exacerbations. In this disease, photosensitivity has previously been noticed but rarely demonstrated. We report three new cases with positive photobiological investigation.

Case reports: Three patients (two males, one female) with a mean age 47 years were seen with reticular erythematous papules on the upper chest and or back. After sun exposure, the lesions were exacerbated. Skin biopsies showed dermal lymphocytic perivascular infiltration with mucin deposition between collagen bundles. Direct immunofluorescence was negative. Antinuclear antibodies were absent. In cabin, ultra-

violet (UV)A exposure reproduced clinically and histologically REM lesions in our cases. UVA and UVB provoking phototests were negative. In all patients treatment with oral antimalarials and external photoprotection was effective.

Conclusions: In our patients, we confirm the photosensitive feature of REM syndrome by provocative irradiation in UVA cabin. The mechanism of triggering is actually unclear. It is supposed that UV radiation, heat, and perspiration are necessary to reveal this affection.

Key words: photosensitivity; reticular erythematous mucinosis syndrome.

Reticular erythematous mucinosis (REM) syndrome is classified in primary and idiopathic forms of cutaneous mucinosis. The term has been introduced by Steigleder et al. (1). This rare disorder affects patients of all age and both sexes but shows a predilection for young adult females. REM syndrome is characterized by erythematous papules coalescing in plaque with reticular pattern on the central chest and upper back. A mild-to-pronounced lymphocytic infiltrate and mucin deposition in dermis are characteristic histologic features. A similar disorder has been described in 1960 and called plaque-like cutaneous mucinosis (PCM) (2), but although REM and PCM were initially considered to be different, most current authors accept that two conditions are single process (3). In REM syndrome, photosensitivity is often noticed by authors (1, 4, 5) but rarely demonstrated (6). We report three new cases with positive photobiological investigation.

Patients and methods

Patients

Two men aged 25 and 51 years, respectively, and a 67-year-old woman were referred to our department

from February 1995 to November 2003. All presented reticulate erythematous eruption of the trunk (Fig. 1). Duration of the skin disease was 8 years (Table 1). After sun exposure, the patients noted that the lesions were more numerous and pruritic. Biopsy specimens from involved skin showed a normal epidermis and a moderate perivascular lymphocytic infiltrate with oedema in the papillar dermis. Alcian blue stain revealed deposits of mucin between the collagen bundles. Direct immunofluorescence was negative. Routine laboratory tests (complete blood cells count, ESR, serum protein electrophoresis) were normal. Antinuclear antibodies were absent. Diagnosis of REM syndrome was done for all patients. The case of a 51-year-old man has previously been reported (7).

Photobiological investigations

Phototesting was performed, using a solar simulator (Dermolum UM-W, Muller Elektronik, Moosinning, Germany) on the back. Minimal erythema dose (MED) of ultraviolet (UV)B was tested and determined 24 h after exposure. Provocative phototest with UVA ($3 \times 10 \text{ J/cm}^2$) and UVB ($3 \times 3 \text{ MED}$) were done in

two areas (10 × 10 cm) of uninvolved skin on 3 consecutive days. In an UVA cabin (Waldman 7001 K, Reichstett, France), body irradiation was done at 10 J/cm² per 24 h during 3 days. Positive reactions were defined as papular eruption appearing on irradiation areas. Skin biopsies and immunofluorescence studies were performed on triggered lesions after UV exposure.

Results

In all observations, photobiological investigations revealed normal MED and negative UVA and UVB phototests. Yet, these patients exhibited abnormal cutaneous reactions (Fig. 2) after UVA irradiation in cabin (Table 1). Skin biopsies from photoinduced lesions revealed dermal perivascular lymphocytic infiltrate. Alcian blue was positive between the dermal collagen fibres (Figs 3 and 4). Direct immunofluorescence was negative.

All patients were treated with oral hydroxychloroquine 200 mg/day and external photoprotection each summer which turned to be effective.

Discussion

In REM syndrome, some ambiguity exists about its relation with photodermatosis group such as lupus



Fig. 1. Case no. 3. Reticulate maculopapular erythema on the left shoulder.



Fig. 2. Case no. 2. Reticulate erythematous eruption on the back 168 h after irradiation by ultravioletA cabin.

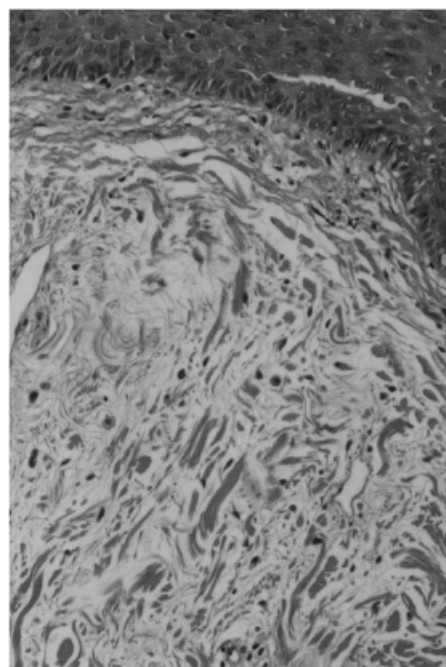


Fig. 3. Case no. 2. Lymphocytic infiltrate and dissociated collagen fibres in the upper dermis.

Table 1. Patients characteristics and data

Sex/age (years)	Duration of eruption (years)	Distribution	UVB and UVA phototest reaction*	Latency of positive reaction after UVA radiation in cabin (dose)	Aspect of photoinduced lesions by UVA cabin
M/51	20	Upper chest and back	Negative	24 h (10 J/cm ²)	Erythematous papules on the trunk
F/67	2	Central chest	Negative	168 h (3 × 10 J/cm ²)	Reticulate erythematous eruption on the upper back
M/25	1	Upper chest and left shoulder	Negative	48 h (2 × 10 J/cm ²)	Erythematous papules on the upper limbs

F, female; M, male; UV, ultraviolet.

*Provocative phototest performed with solar simulator (Müller Elektronik).

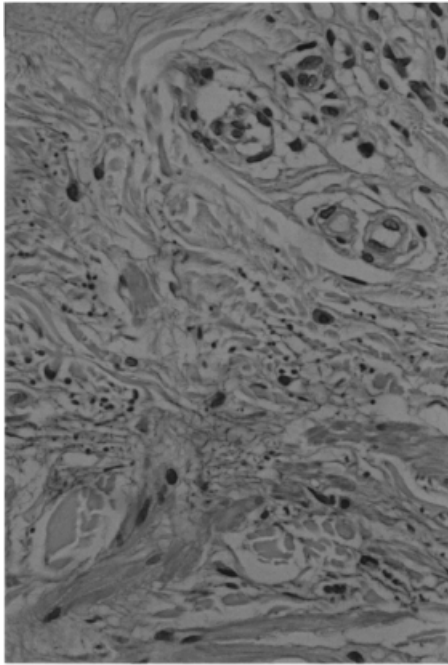


Fig. 4. Case no. 2. In the dermis, fibrillary deposits of mucin revealed by Alcian blue stain and perivascular lymphocytic infiltrate.

erythematosus (LE) polymorphic light eruption (PLE) and Jessner's lymphocytic infiltration (JLI) of the skin. Sun exposure exacerbated REM syndrome in nearly half of patients (8). Photoprovocation testing has been performed with disappointing result (1, 4–6, 9–11). Only one case had the rash reproduced 4 weeks after the UVB and UVC exposure (6). Furthermore, Mc Fadden and Larsen (9) induced histologically lymphocytic dermal infiltrate with traces of positive Alcian blue staining but without evidence of clinical lesion on test sites. In our study the patients were tested in an UVA cabin. This provocative method has not previously been used (1, 4–6, 9–11). In our cases, UVA irradiation of total body reproduced REM lesions. This finding indicates, that many conditions may operate to trigger REM syndrome including UV radiation, heat, and perspiration (4). Moreover, the test reactions appeared after a relative long interval of 1–7 days. This delay between first UV exposure and skin-induced lesions may explain the absence of photosensitivity history in most patients with REM.

The pathogenic mechanism of REM remains unknown. Thus the accumulation and role of mucin in upper dermis is unclear. Some authors believe REM to be a subset of LE (4, 12). Dermal mucin deposits is occasionally found in LE (13). In REM syndrome, direct immunofluorescence staining may be positive showing deposition of immunoglobulins and

complement along dermo-epidermal junction in lesional skin (4, 8, 9, 12). The improvement of this affection syndrome under antimalarial treatment is suggestive of similarities with LE. Furthermore, in LE photosensitivity often occurs and test reactions generally appeared in 1 week (14). Yet our patients with REM have the negative autoantibody profile.

PLE and JLI may be difficult to differentiate from the REM syndrome. Thus PLE contrasts with REM in which exacerbations occur out of the sunny period such as perspiration, menstruation, pregnancy, and stress (4). JLI is characterized by papular lesions located predominantly on the face with the onset in summer and persistence (15, 16). All three disorders show histologically perivascular mononuclear infiltrate but in PLE and JLI deposition of mucin is usually absent.

In conclusion, we confirm the photosensitive feature of REM syndrome by provocative irradiation in UVA cabin. The mechanism of triggering is actually unclear. Therefore we consider that provocative phototesting with standardized protocols will help identify the pathogenesis of the disease in the future.

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