

Brownish discoloration with some erythematous plaques on the chest in a young woman: a quiz

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A 25 year-old woman with a history of having erythematous macules and papules with a reticular pattern on her chest, spreading to the upper abdomen and neck for about three months has visited outpatient clinic for consultation. She was in a good general condition and only complained of a mild pruritus within skin lesions. She tried topical steroids of moderate potency in the past to treat skin lesions; however, without a big success. She was otherwise healthy and took no other medications.

Which diagnosis is most likely in our patient?

- A. Polymorphic light eruption
- B. Seborrheic dermatitis
- C. Prurigo pigmentosa
- D. Reticular erythematous mucinosis
- E. Livedo reticularis

See next page for answer.



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ANSWER: RETICULAR ERYTHEMATOUS MUCINOSIS

Reticular erythematous mucinosis (REM) syndrome is a condition first described by Steigleder et al. [1] in 1974. However, the first description of the entity currently known as REM has been reported already by Perry et al. [2] in 1960 as a plaque-like form of cutaneous mucinosis. REM is a rare, chronic and persistent disorder that affects patients of all ages and both sexes, but most cases are middle-aged women. The aetiology of REM remains unknown. REM has been considered by some authors to be an idiopathic, primary form of cutaneous mucinosis, while by others as a disorder closely related to or associated with cutaneous lupus erythematosus, in particular with lupus erythematosus tumidus. Both diseases share common features such as the plaque-like clinical aspect of lesions, the lack of significant immune serological abnormalities, histology, and response to antimalarial drugs [3, 4]. Other disorders reported in association with occasional cases of REM syndrome include malignancies (breast and rectal carcinoma, Hodgkin disease) and autoimmune disorders, including thyroid disease, diabetes mellitus and ulcerative colitis. There are some reports linking REM to hormonal factors such as contraceptive pills, pregnancy and menses [3]. Viruses may also play a causal role, but this is still controversial and not well documented in the literature [5]. The photosensitivity and photodistribution of skin lesions observed in many patients suggest that sun exposure may play a role in the pathogenesis of REM. However, the recent review of 25 cases has reported that sun exposure worsened or triggered REM in five patients, but also improved the disease in another four [3]. Thus, the role of sunlight remains controversial. Regarding our case, the patient admitted, that erythematous lesions improve in summer, despite a clearly photodistributed location of skin abnormalities.

Clinically, REM syndrome manifests as the persistent erythema in a reticular pattern and/or confluent erythematous papules and plaques that lack scales or other surface changes. Skin lesions are typically located on the central area of the chest or on the upper back but may also spread to the upper abdomen. The arms, face, and abdomen are less frequently affected [6]. Mucous membranes, genitalia, and internal organs are not involved, but in 1977 Keczkes and Jadhav reported a case with gum involvement and cervical lymphadenopathy [7]. Most of the affected patients are asymptomatic. However, in 20–30 percent of cases pruritus is reported [8]. Slight burning sensation may also occur in some cases [3]. The clinical course is cyclic with remission and exacerbation [9].

A perivascular, and occasionally perifollicular, mononuclear cell infiltrate with increased dermal mucin deposition is a characteristic histologic feature. The epidermis appears normal. Direct immunofluorescence is usually negative for immunoglobulins, fibrin, and complement, although a few cases of REM with positive direct immunofluorescence and granular deposits of immunoglobulin IgM and C3 have been reported [4].

There is no single evidence-based treatment for REM, but antimalarials should be considered as a first-line therapy. Hydroxychloroquine 200–400 mg daily is considered the treatment of choice [3, 4, 6]. Numerous treatment options for REM syndrome have been examined. Successful treatment of REM syndrome with topical tacrolimus or pimecrolimus has been reported [4, 10]. Another effective treatment option is pulsed-dye laser (PDL) [4, 11]. UVA-1 (340–400 nm) therapy [12] and UVB excimer lamp (308 nm) [13] also represent an effective alternative in the treatment of REM. To exclude photosensitivity, photoprovocation testing should be performed on the patient's healthy back skin before proceeding with phototherapy. However, further studies on large case series are needed to evaluate the efficacy of available treatment options.

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