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Persistant Reticular Erythema After Severe Sunburn: Reticular Erythematous Mucinosis: Case Report

Şiddetli Güneş Yanığı Sonrası Ortaya Çıkan Persistan Retiküler Eritem: Retiküler Eritematozus Musinozis

ABSTRACT Reticular erythematous mucinosis (REM) is a rare, chronic and persistent disease characterized by reticular macular erythema or erythematous papules and plaques mostly on the central area of the chest and back of young to middle aged woman. Its etiopathogenesis is not known clearly. Sun light, pregnancy, menstruation, emotional changes, heat, oral contraceptive drugs may aggravate the lesions. Because of flare of lesions after exposure to ultraviolet, common clinical and histopathological features and good response to systemic antimalarials, it's considered in the spectrum of lupus erythematosus-like diseases by some authors. We report a case of REM with persistent reticular macular erythema started after sunburn with positive antinuclear antibody titers.

Key Words: Erythema; mucinoses; antibodies, antinuclear

ÖZET Retiküler eritematöz musinozis (REM) sıklıkla genç ve orta yaş erişkin kadınlarda göğüs ile sırt orta hatta ortaya çıkan retiküler maküler eritem veya eritemli papül ve plaklarla karakterize nadir ve kronik bir hastalıktır. Histopatolojik olarak perivasküler ve perifoliküler mononükleer hücre infiltrasyonu ve dermal musin birikimi ile karakterizedir. Etiyopatogenezi kesin olarak bilinmemektedir. Güneş yanığı, gebelik, menstruasyon, emosyonel değişiklikler, sıcaklık ve oral kontraseptif ilaçlar lezyonları alevlendirebilmektedir. Ultraviyole maruziyeti sonrası lezyonların alevlenmesi, ortak klinik ve histopatolojik bulgular ve sistemik antimalaryal ilaçlara iyi yanıt vermesi nedeniyle bazı otörlerce lupus-eritematozus benzeri hastalık spektrumunda olduğu düşünülmektedir. Burada güneş yanığı sonrası ortaya çıkan retiküler maküler eritemi ve pozitif antinükleer antikor değerleri olan bir REM olgusu sunulmaktadır.

Anahtar Kelimeler: Eritem; musinozis; antikorlar, antinükleer

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eticular erythematous mucinosis (REM) is a rare disease characterized by reticular macular erythema or erythematous papules and plaques on the central area of the chest and back. Herein a case of REM with persistent reticular macular erythema on the breasts started after sunburn was reported.

CASE REPORT

A 16-year old girl presented with a 1-year history of symmetrical reticular erythema and erythematous plaques on the breasts which started after a severe sunburn last summer (Figure 1). She stated that the rash was asymp-

tomatic however, exposure to heat aggravated the rash; in contrast exposure to cold decreased it. Her medical and family history was unremarkable and review of the systems were normal.

Routine complete blood count, blood chemistry, erythrocyte sedimentation rate, thyroid function tests, antithyroglobulin antibody, C3, C4 were within normal limits. Extractable nuclear antigens and anti-HIV antibody were negative. Antinuclear antibody (ANA) was present at a titer of 1/80. All other criteria necessary for a diagnosis of systemic lupus erythematosus (SLE) were lacking.

Hematoxylin–eosin staining of a punch biopsy specimen from the lesional skin revealed normal epidermis and basement membrane with a perivascular and perifollicular lymphomononuclear cell infiltrate in the dermis (Figure 2). Alcian blue staining revealed mucin deposits in the upper and mid-dermis (Figure 3). With the clinical and histopathological findings diagnosis of REM was made. An informed consent was obtained from the patient for the publication.

DISCUSSION

REM is a rare, chronic and persistent disease characterized by reticular macular erythema or erythematous papules and plaques on the central area of the chest and back of young to middle aged woman. It's usually asymptomatic. In rare instances male and children may be affected and lesions can be present on the arms, face, legs and abdomen.^{1.2}



FIGURE 1: Symmetrical reticular erythema and erythematous plaques on breasts.

(See color figure at http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/)

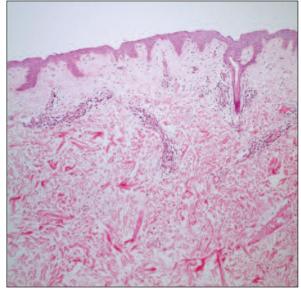


FIGURE 2: Normal epidermis and basement membrane with a perivascular and perifollicular lymphomononuclear cell infiltrate in the dermis (haematoxylin and eosin, original magnification x 10).

(See color figure at http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/)

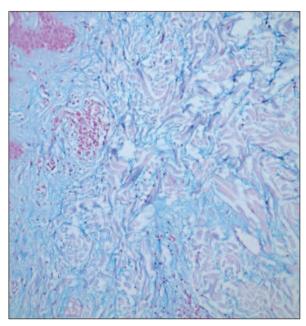


FIGURE 3: Mucin deposits in the upper and mid-dermis (Alcian blue, original magnification x40).

(See color figure at http://www.turkiyeklinikleri.com/journal/dermatoloji-dergisi/1300-0330/)

The etiopathogenesis of REM remain undefined at present. Several factors have been associated with the induction of syndrome, including immunological disturbances, viral processes and solar irradiation.^{1,3} Sun light, pregnancy, menstruation, emotional changes, heat, oral contraceptive drugs are thought to aggravate the lesions.¹ Some authors consider REM in the spectrum of lupus erythematosus-like diseases due to common features such as flare after exposure to ultraviolet radiation, clinical manifestations, histopathology and good response to systemic antimalarials.^{1,4,5}

Histopathological examination of REM is characterized by normal epidermis, mild or moderate mononuclear infiltrate located predominanly around blood vessels and hair follicles and variable amounts of mucin mostly in the upper and mid dermis.^{1,2} Although mucin can be recognized even in routinely stained sections in papular lesions, in macular lesions, the mucin may become apparent only on staining with Alcian blue.² In several cases, the deposition of immunoglobulins, particularly IgM, along the basal layer was shown in direct immunofluorescence examination.^{1,2,4}

In differential diagnosis of REM especially Jessner's lymphocytic infiltration of skin and tumid lupus erythematosus must be considered. In histopathological examination of these conditions perivascular and perifollicular lymphocytic infiltration and increased mucin are common. The lymphocytic infiltrate is usually much denser in tumid lupus and Jessner's lymphocytic infiltration than that in REM but often clinicopathologic correlation is essential in distinguishing these conditions.²

Systemic antimalarials are the main treatment of choice for REM. Topical and systemic corticosteroids, topical calcineurin inhibitors, UVB, UVA1, pulse dye laser, dapsone, oral antihistamines, tetracycline and cyclosporine have all been used with variable results.¹

In conclusion, in the light of this case we would like to underline that persistent reticular macular erythema on the central area of the chest after sunburn together with dermal mucin deposition on the biopsy specimen should alert the physician for a diagnosis of REM which is in fact a rare disease. It is wise to consider REM as part of the spectrum of lupus erythematosus-like diseases and that a long term follow-up for SLE criteria might be mandatory at least for patients with positive ANA.

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