Discussion

Erythema multiforme-like lesions are a classically self-limited reaction to a drug or infective agent without any specific serological antibodies. It is characterized by acral and/or mucosal targetoid lesions with a central dusky necrotic zone, a middle-skin-colored zone, and a peripheral erythematous zone[2, 4]. While our patient had no trigger factors, she was positive for ANA, dsDNA, and RO, which differentiated RS from erythema multiforme [5].

The diagnostic criteria for RS were redefined in 2000, whence three major criteria and one minor criterion were to be satisfied for diagnosis. The major criteria were LE in any form, EM-like lesions (mucosa may or may not be involved), and ANA positivity (speckled pattern). The minor criteria consisted of the presence of chilblains, RF, and anti-Ro, or anti-La antibody positivity [1]. Later, it was suggested that RS may be considered a rare subtype in the spectrum of lupus-specific skin lesions, and the latest diagnostic standards for RS were proposed. This diagnosis of RS required all four major and at least one minor criterion [2]. The major criteria were chronic cutaneous LE, EM-like lesions (typical target or targetoid lesions), at least one ANA positivity speckled pattern, anti-Ro/SSA and anti-La/ SS-B antibodies positivity, and negative direct immunofluorescence (DIF) on EM-like lesions. The minor criteria were the absence of triggering factors (infections or medications) and typical location (acral and mucosal) of EM, the presence of at least one of the criteria for an SLE diagnosis provided by the American College of Rheumatology (ACR) except for discoid or malar rash, photosensitivity, chilblains, oral ulcers, and ANA positivity. Our patient met the former diagnostic criteria for RS, although she had no history of chronic cutaneous lesion, and DIF was not performed to fully match later criteria. Furthermore, based on her high SLEDAI-2K score, a flare-up of SLE was considered to have possibly been caused by adalimumab. A review of other case reports of RS for lupus disease activity in other organs revealed interesting results that confirmed our presumption that RS occurred in the context of SLE activity. This syndrome has been reported in a patient with SLE in the context of stopping immunosuppressive drugs; the patient, who had withdrawn his treatment one month earlier, referred with skin rash all over his body, laboratory evidence of hemolysis, low serum levels of C3 and C4, raised ESR and CRP, red cell casts in U/A, positive ANA with a speckled pattern, a positive anti-La, and anti-Ro. The patient did not experience a recurrence of skin lesions after restarting treatment with oral prednisolone, HCQ, and azathioprine [6]. RS has also been reported as the first manifestation of SLE in an 18-year-old girl who presented with skin rashes, fever, polyarthralgia, alopecia, lymphadenopathy, pancytopenia, raised ESR, low serum levels of complement C3, C4, microalbuminuria, as well as positive ANA and anti-RO antibodies. She was treated with prednisolone and HCQ and responded well, which led to the disappearance of her skin lesions and oral ulceration over the next days [7]. RS has even been reported in a patient with end stage renal disease caused by lupus nephritis who had skin rash, pancytopenia, and low C3 and C4 simultaneously; treatment with pulses of methylprednisolone and rituximab maintained the remission of skin disease and cytopenia [8]. Gallo et al. reported a unique case of RS. Their patient presented with lymphadenopathy and hepatomegaly, rash, fever, arthralgia, leukopenia, and elevated ESR levels; prednisone and HCQ administration led to gradual improvement of the patient's skin manifestations and elimination of related symptoms after two months of follow-up [9]. Another case of RS was reported in a patient with multiple reddish lesions, fever, joint pain, photosensitivity, anemia, raised ESR, the presence of cellular casts in U/A, a positive test for ANA speckled pattern, and anti-ds-DNA [10]. A review of the mentioned cases established that skin symptoms lead to earlier referral of patients, which may be considered a leading indicator for the diagnosis of SLE flare-up. According to the mentioned case reports, RS has similar prognoses and treatments to SLE or DLE which occur alone (7-10). Likewise, our patient responded well to corticosteroids, HCQ, and MMF, and 6 months of follow-up indicated no further recurrence of skin lesions.

Conclusion

Because RS may be a symptom of SLE activity

and organ involvement, it is concluded that this is more of a subtype of SLE than a separate clinical entity.

Acknowledgment

Not applicable.

Conflict of interest

None

Funding

No funds.

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