Bulgarian patient with Rowell syndrome!

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We present a 47 year-old woman with histologically and serologically confirmed subacute cutaneous lupus erythematosus since 2009. The initial clinical appearance of the lesions was that of polycyclic and monocyclic annular plaques, located on the face, trunk and extremities (Figs 1a, 1b). The patient was initially treated with oral chloroquine phosphate 250mg/daily in combination with 40 mg intravenous methylprednisolone, leading to control of the disease, and subsequently tapered until a maintenance oral dose of 4 mg over time.

With the 8 cycles of combined systemic therapy with chloroquine phosphate 250mg/daily and methylprednisolone under a reduction scheme, complete but short-lived remissions were observed over the last 6 years. In the recent recurrences of the disease, additional symptoms appeared as a possible sign of initial systemic involvement, in the form of subfebrile temperatures, joint pain, weakness and prostration, accompanied by the eruptive appearance of multiple new cutaneous lesions.

The patient underwent, for two times, a treatment scheme with azathioprine 50 mg/ twice daily in combination with a

systemic steroid, with a dose and tapering regimen similar to those described above for the chloroquine therapy.

At the time of the current hospitalisation the patient presented with pruritic, annular, erythematous, partly urticarial plaques, at some places with bizonal, erythema multiforme-like appearance (Figs. 1a, 1b) all over the body, that were present for a few months and whose worsening was related to sun exposure. Physical examination revealed two types of lesions: lesions on the extremities with a clearly bizonal configuration, displaying a well-defined peripheral border and blistering center, which clinically corresponded to a picture of erythema multiforme (EM) (Figs. 1a, 1b). On the trunk and extremities, a second type of lesion was seen, namely annular, partially urticarial plaques, sometimes with fine scaling in the inner portion of the border, and with a clear central area (figs. 1c,1d). Repeated histopathological examination of skin biopsies from lesional tissue showed hyperkeratosis, vacuolar degeneration of the basal cell layer, inflammatory infiltrate in upper dermis composed of lymphocytes and plasma cells in the upper dermis, particularly around the pilosebaceous units, findings consistent with subacute cutaneous lupus erythematosus.

Figure 1a: A oval erythematous lesions with erythema multiforme-like appearance on the right lower leg.

Figure 1b: erythematous plaques with erythema multiforme-like appearance in the area of the right upper limb.

Figure. 1c-d: annular, partially urticarial plaques with fine scaling in the inner portion of the border located along the lower limbs.

A direct immunofluorescence was performed in another department a few years ago, with the results identifying typical changes to subacute cutaneous lupus erythematosus, namely epidermal deposits of IgG.

Serology revealed positive ANA (1:1280), positive anti-La/SS-B (96 IU/ml; > 50), strongly positive anti-Ro/SS-A (95 IU/ml; >50) and anti-Ro-52 (112 IU/ml; >50); C3 of 1,32 g/L (normal: 0,90-1,80) and C4 of 0,15 g/L (normal 0,10-0,40) were within the reference range; highly positive RF (59,4 IU/ml; normal 0-14); and normal anti-phospholipid antibodies. Based on the clinical manifestations erythema multiforme-like and previously established LE, based on clinicopathological correlation a diagnosis of Rowell syndrome was made. Systemic therapy with chloroquine 250 mg/day was initiated in combination with 40 mg of intravenous prednisolone for 4 days under stable clinical status. Dose reduction was planned within the hospitalization. Additional therapeutic measures included esomeprazole 40 mg/day, topical mometasone furoate 0,1% and sun protection.

Rowell syndrome is mainly found in middle-aged women, and the major criteria include coexistence of lupus erythematosus and erythema multiforme-like lesions, accompanied by positive ANA (1). In turn, minor criteria include chilblains, positive anti-

La (SS-B) or anti-Ro (SS-A) antibodies, and reactive RF, which were also observed in our patient, except for the absence of chilblains (1). However, it should be mentioned that, according to certain authors it is still questionable whether RS is an overlap syndrome, a real association, or just a coincidence of LE and EM (2). Some of them even classify the syndrome as an autonomous type of cutaneous LE (3). The treatment of LE and RS is similar and good results are reported with therapeutic regimens that include azathioprine, antimalarials, prednisone, dapsone or cyclosporine (2). According to the literature, cases of RS have a good general prognosis with complete remission of skin lesions within 1 year. In order to avoid relapses, continuous treatment with hydroxychloroquine 200 mg/day has been described (1).

References

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