Subacute Cutaneous Lupus Erythematosus with Erythema Multiforme-like Lesions: Consideration for Rowell Syndrome

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Case Presentation:

Patient: 83 year-old Caucasian male.

History of Present Illness: The patient presented to the emergency department with a ten day history of a mildly pruritic generalized rash which developed after completing a course of high dose oral prednisone for previously diagnosed idiopathic thrombocytopenic purpura (ITP). He denied any other new medications. The patient had a 10 year history of discoid lupus erythematosus (DLE) which was under control with only sun protection. A review of systems was negative for joint pains, oral ulcers, headache, and other systemic findings except for fatigue, which began around the time of the patient’s skin findings. He denied any recent treatments for the rash and had no history of a similar problem in the past. There was also no reported history of herpetic lesions.

Medical History/Surgical History: Discoid lupus erythematosus, ITP, cutaneous disease, GERD, history of prostate cancer status post radiation therapy

Current Medications: Pantoprazole, tamsulosin, zinc oxide ointment

Physical Examination: The patient had multiple erythematous thin papules, some with dusky centers giving a targetoid appearance, coalescing into plaques on the scalp, neck, ears, trunk and extremities. Thin red annular plaques were seen on the palms. There were scattered vesicles on the trunk and extremities that desquamated with light pressure. Purpuric thin papules and plaques were present on the legs. The face was spared and there was no oral or conjunctival involvement. Lesions consistent with discoid lesions were not identified.

Laboratory Data: CBC WNL except platelets 81,000 (150,000-400,000/microliter); ANA 1:2560 (<40 titer) speckled pattern; SSA 155, SSB 167 (normal <20); Anti-dsDNA, RNP, Smith, Sc-70, anti-histone, p-ANCA, and c-ANCA WNL.

Studies: Chest x-ray: Negative

Biopsy: Health Network Labs (S13-25193, 7/30/2013) Left lateral arm: Interface and subepidermal vesicular dermatitis with scattered single dyskeratotic keratinocytes. Regions of confluent thickness keratinocytic necrosis, consistent with erythema multiforme. DIF from the left lateral arm with IgG deposition in a stippled, granular pattern over keratinocyte cytoplasm. This finding is reported in, though not exclusive to, EM-like lesions. ANA positive for speckled pattern ANA, SS-A and SS-B, findings reported in RS but also fairly specific for SCLE. Direct immunofluorescence (DIF) revealed granular IgG deposition over epidermal keratinocytes. This finding is reported in, though not exclusive to, EM and thought to be due to anti-Ro antibodies. In contrast, EM rarely has positive DIF, though when present is typically IgM or C involving the basement membrane zone and superficial blood vessels.

Application of the aforementioned diagnostic criteria for RS reveals this patient met the criteria presented by Zeitouni et al. However, the positive DIF and lack of discoid lupus lesions clinically would result in exclusion from the criteria described by Torchia et al. Furthermore, the patient responded well to Plaquenil and corticosteroids, treatment commonly used for SCLE, though corticosteroids may also be effective in EM. Considering the lack of agreement in current literature, failure to meet the most recent proposed criteria, findings and response to therapy more consistent with SCLE without precipitating factors of EM, this patient best fits the description SCLE without EM-like lesions, as opposed to RS.

The question remains as to whether or not RS exists. There may be a subset of patients with typical RS findings and response to therapy more consistent with SCLE without EM-like lesions. Whether or not RS exists, this patient best fits the description SCLE without EM-like lesions, much like this case.

References:

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