Cutaneous vasculitis in lupus treated with IV immunoglobulin

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Presentation

A 44-year-old man presented to the emergency department with a 3-week history of painful ulcers in the sacral region with a new skin rash. He had received an early diagnosis of systemic lupus erythematosus (SLE) at the age of 7 years and was using only prednisone 15 mg daily (he had stopped using hydroxychloroquine by himself 3 months earlier). On physical examination, multiple necrotic ulcers were noted with major livedo racemosa intermingled (Fig. 1a). The systemic examination was unremarkable. A differential diagnosis workup between vasculitis and antiphospholipid syndrome was performed, and a skin biopsy was promptly collected.

Histopathologic examination revealed diffuse neutrophilic infiltrate around small vessels (Fig. 1c). Direct immunofluorescence showed moderate homogeneous deposits of both immunoglobulin M (Fig. 1d) and C3 (Fig. 1e) along the basement membrane zone.

Laboratory tests revealed hemoglobin 11.8 g/dL; an elevated erythrocyte sedimentation rate of 47 mm, C-reactive protein 10 mg/dL, creatinine 1 mg/dL, urea 30 mg/dL, cryoglobulin negative, low C3 of 39 mg/dL and C4 of 8 mg/dL, ANCA and serology tests for HIV, hepatitis B and C, and VDRL were negative, ANA 1/640 nuclear homogeneous, anti-Smith > 180 IU/mL, anti-SSA > 240 IU/

mL, anti-SSB > 320 IU/mL, anti-dsDNA > 379 IU/mL, negative anti-phospholipid antibodies (anti-cardiolipin, anti-beta 2 glycoprotein I, and lupus anticoagulant), urinalysis without significant proteinuria; and no dysmorphic erythrocytes.

We opted to start prednisone 1 mg/kg daily with hydroxychloroquine 400 mg/day, and an improvement in the livedo was observed. However, the patient's ulcers became infected during the hospital stay (Fig. 1b), demonstrated by a positive culture for *Pseudomonas aeruginosa*, and they were treated with culture-guided ciprofloxacin and surgical debridement. Due to clinical instability from sepsis, a second immunosuppressive agent was reconsidered, and we chose to start immunoglobulin 1 g/kg for two consecutive days. Unfortunately, soon after, the patient died from septic shock.

Discussion

Cutaneous vasculitis may be seen in 19–28% of patients with SLE and accounts for 89% of vasculitis cases in this disease [1]. Male sex, young-onset SLE, disease duration, presence of livedo reticularis, hematological parameters (anemia and high erythrocyte sedimentation rate), anti-dsDNA, anti-SSA [2], anti-SSB, and anti-Smith are the most common parameters associated with cutaneous vasculitis in SLE patients [3].

Histopathology is characterized by small-vessel neutrophilic vasculitis, and under immunofluorescence, deposits of IgG, IgM, and C3 at the dermo-epidermal junction are common (lupus band test) [4]. However, it is important to evaluate both laboratory and clinical profile of the patient since lupus band test could be found in bullous pemphigoid, porphyrias, rosacea, skin infections and even in healthy sun-exposed skin [5].

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Fig. 1 **a** Livedo racemosa and necrotic ulcers at admission. **b** Lesion aspect after infection due to *Pseudomonas aeruginosa*. **c** Perivascular inflammatory infiltrate composed of neutrophils (hematoxylin and eosin $\times 400$). **d** Direct immunofluorescence showing moderate homogeneous deposits of IgM along the dermo-epidermal junction (lupus band test) (fluorescein isothiocyanate, $\times 100$). **e** Direct immunofluorescence showing moderate homogeneous deposits of C3 along the dermo-epidermal junction (lupus band test) (fluorescein isothiocyanate, $\times 200$)

Antimalarials, thalidomide and dapsone are treatment options for cutaneous vasculitis in lupus. For resistant cases immunoglobulin could be an option. The usual dose is 1 g/kg for 2 consecutive days followed by 400 mg/kg monthly until disease resolution or for 6 months [6]. The use of immunoglobulin is considered off-label since there is a lack of randomized controlled trials, but it is an excellent option, particularly in unstable patients who cannot tolerate another immunosuppressant drug.

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Declarations

Conflict of interest The authors have nothing to disclose. The authors declare that they have no conflict of interest.

Ethics approval and consent to participate Written informed consent was obtained from the patient for publishing this material.

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