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Retiform Purpura with Overlying Bullae: Unusual Presentation of Systemic Lupus Erythematous in Children

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Bullous lesions in the form of sub-epidermal blisters or due to severe edema and hydropic degeneration of the basal layer rarely occur in children with systemic lupus erythematous (SLE).[1] Here, we report a 13-year-old girl presenting with anemia and concurrent retiform purpura with overlying bullae on the dorsum of feet; she was diagnosed with SLE.

A 13-year-old girl was referred to our hematology-oncology clinic due to severe anemia, fever, and musculoskeletal pain. The fever was the intermittent and low grade for the last 2 months. Physical examination revealed pallor, arthritis, and arthralgia of both knee, wrist, and ankle joints, violaceous purpuric lesions with overlying bullae on the dorsum of feet and the lower legs (Figure 1A), and palpable purpura in a livedoid, reticulate pattern on the dorsum of hands (Figure 1B). Abdominal examination revealed hepatosplenomegaly. Malar rash, oral ulcers, and cutaneous lesions on the face, neck, or trunk were not identified, but the patient was complaining of photosensitivity.

The complete blood count was as follows: white blood cell: 9.7 × 10^3/mm^3, neutrophil: 83%, hemoglobin: 6.5 g/dL, MCV: 94 fL, platelet: 82 × 10^3/mm^3. The direct Coombs test was positive. Erythrocyte sedimentation rate was 117 mm/h, and lactate dehydrogenase (LDH) was 1808 IU/L. The levels of blood urea nitrogen, creatinine, liver function tests, and alkaline phosphatases were within normal limits. Urinalysis was negative for proteinuria or cell casts. Anti-double stranded DNA (anti-dsDNA) was 256 IU/mL (>24 IU/mL positive), and antinuclear antibody (ANA) was 2.36 (>1.2 positive). Complement levels were normal (C3: 94 mg/dL [normal: 90-180], C4: 14 mg/dL [normal: 10-40]). Anti-tissue transglutaminase IgA (tTG-IgA) was 38 U/mL (>18 positive). Blasts were not seen in peripheral blood smear. No evidence of lupus nephritis was found in this patient. The SLE was diagnosed with positive ANA, positive anti-dsDNA, autoimmune hemolytic anemia, thrombocytopenia, photosensitivity, and arthritis.

Treatment was started with intravenous methylprednisolone pulse (30 mg/kg/day for 3 days) therapy and was continued with oral prednisone (1 mg/kg/day in 3 divided doses) and hydroxychloroquine (200 mg/day); systemic manifestations and skin lesions gradually improved after a week.

Skin is the second most commonly affected organ by SLE. Cutaneous eruptions are lupus-specific (e.g., malar rash) or non-specific (e.g., alopecia). Bullous lesions are non-specific for SLE.[2,3] Shin et al.,[4] in a cohort study showed that mucocutaneous manifestation is the most common clinical feature of juvenile SLE.

Vesiculobullous lesions occur in less than 5% of children with SLE; they might be isolated or accompanied by other cutaneous lesions.[5] These patients are usually young women in their second decade of life.[6] These lesions should be considered in the differential diagnosis of other cutaneous vesiculobullous lesions of infectious and non-infectious origin and can cause diagnostic and therapeutic challenges.[7] Although, in some research, there was a correlation between these lesions and lupus nephritis,[1,8] there was no evidence of lupus nephritis in this patient. In conclusion, SLE should always be considered in children presenting with purpuric eruptions with overlying bullae.

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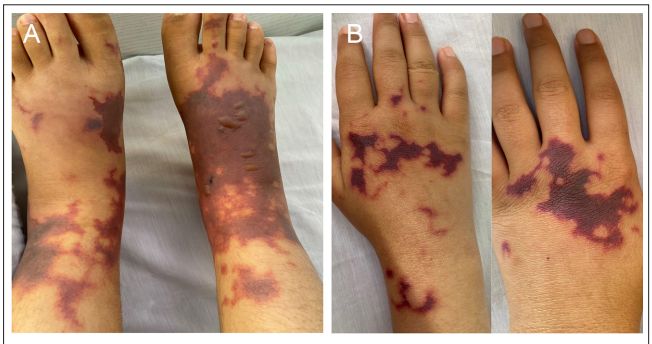

Figure 1. Retiform, violaceous purpuric lesions with overlying bullae on dorsum of feet and on the lower legs (a) and palpable purpura in a livedoid, reticulate pattern on the dorsum of hands (b).

Informed Consent: Written informed consent was obtained from the patient's parents.

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