Dapsone hypersensitivity syndrome during Henoch–Schonlein purpura treatment

Sir,

An 11-year-old girl was admitted with high-grade fever for 2 days and pruritic rashes all over the body for 1 day after completing a course of oral dapsone (50 mg/day) for 2 weeks for biopsy-proven Henoch – Schonlein purpura. On examination, she was febrile and had maculopapular rash without pustules over her forehead, neck, trunk, back, and extremities [Figures 1 and 2]. She had tender enlarged liver 4 cm below the right costal margin. Multiple bilateral tender cervical and retroauricular lymph nodes measuring 1-2 cm were noted. There were no Koplik's spots. The peripheral smear showed mild neutrophilia and toxic granulations. Her liver enzymes, aspartate aminotransferase (178 U/L) and alanine aminotransferase (229 U/L) were elevated. Abdominal ultrasound showed uniform liver enlargement with a slight increase in echo texture. Urine microscopy revealed 25-30 pus cells per high power field.

Paul–Bunnel test was negative. Antinuclear antibody profile was negative. A diagnosis of dapsone hypersensitivity syndrome (DHS) was made and she was treated with oral prednisolone for 2 weeks. Her clinical condition improved over 10 days.

DHS is characterized by the sudden onset of papular or exfoliative rash, accompanied by fever, malaise, and weakness, followed by jaundice and lymphadenopathy, resembling infectious mononucleosis.[1-3] Anemia, oral erosions, conjunctivitis, splenomegaly, eosinophilia, atypical lymphocytosis, and rise of liver enzymes are other corroborative findings. It is important to emphasize that all those features need not necessarily be present.[2-4] It is believed that hydroxylated metabolites and genetic susceptibility are important in the pathogenesis of DHS.[4] Patients on dapsone should be monitored closely for new skin rashes and systemic organ involvement, for early recognition of this potentially fatal adverse drug reaction. A high index of suspicion with temporal correlation is required for early recognition and prompt discontinuation of dapsone.[4,5] Dapsone challenge test is not recommended in previously sensitized patients due to its life threatening reactions.[5] The

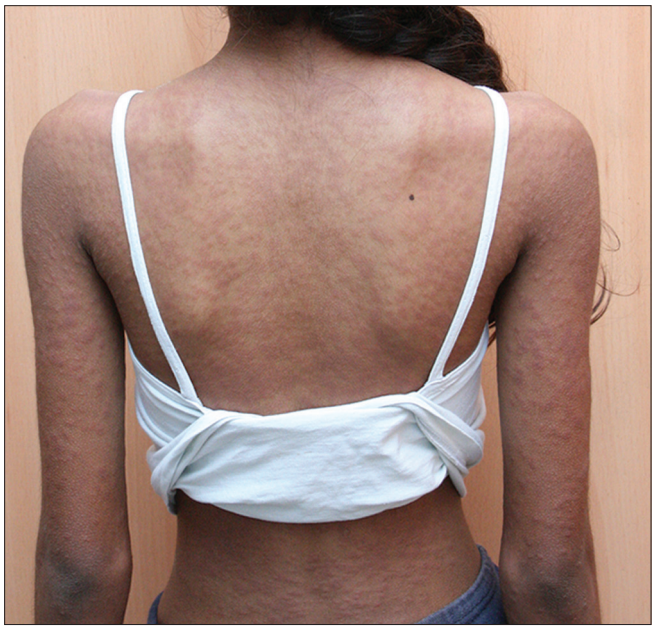

Figure 1: Maculopapular rash over back in a patient with dapsone hypersensitivity syndrome

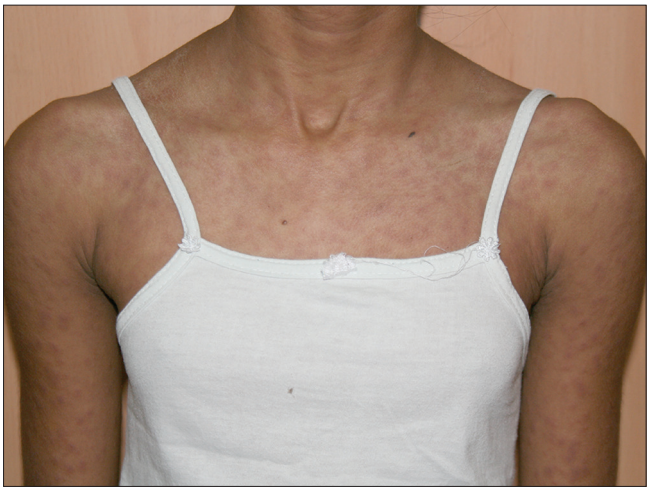

Figure 2: Maculopapular rash over upper trunk

management of DHS is supportive, involving management of shock, fluid, and electrolyte balance, pancytopenia, fastidious skin care, and prevention of secondary infection. Steroids may be used in severe cases not responding to supportive management.[1,2,5]

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