Acute hemorrhagic edema of infancy

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A 15-month-old male child presented with sudden onset of red-colored lesion over lower extremities and face along with mild fever and swelling of extremities. The patient's mother denied any history of drug intake or vaccination before the development of lesion. There was no history of any trauma.

On cutaneous examination, large, cockade, annular purpuric plaques over the lower extremity with involvement of face was present [Figures 1 and 2].

The child also had edema of hands and feet, which were not tender. Routine hematological investigation was normal. Coagulation profile, ultrasonography of abdomen, and routine urine analysis was also normal. Histopathology of erythematous plaque showed leukocytoclastic vasculitis with fibrinoid necrosis and perivascular neutrophilic infiltrate [Figure 3]. Therefore, on the basis of clinicohistopathological correlation, the diagnosis of acute hemorrhagic edema of infancy (AHEI) was confirmed. The patient was followed-up for 14 days and was started on symptomatic treatment. There was complete spontaneous subsidence of lesions within 2 weeks.

AHEI (Finkelstein disease, Medallion-like purpura, Seidemayer›s syndrome, infantile postinfectious iris like purpura and edema, and Purpura en cocarde avec oedeme) is a rare immune complex-mediated small vessel cutaneous leukocytoclastic vasculitis. It is characterised by triad of large purpuric lesions, fever, and peripheral acral edema. It occurs almost exclusively in children between 4 months to 2 years of age with recent history of upper respiratory tract infection (75%) or intake of antibiotics. Maximum cases present in winter season with a male predominance.[1]

AHEI was first described by Snow in 1913 in United States. It is characterised by sudden onset of large, annular, targetoid purpuric lesions with cockade pattern involving face and extremities with characteristic sparing of trunk. Along with

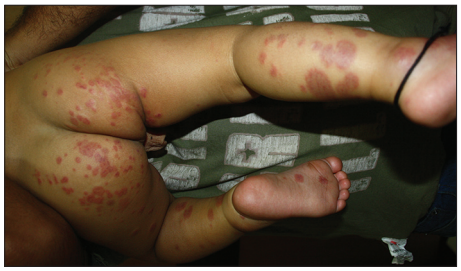

Figure 1: Annular, purpuric lesion over extremities

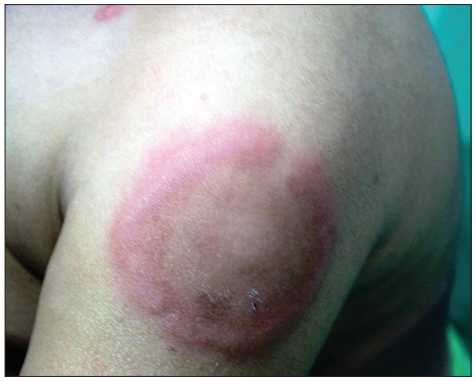

Figure 2: Cockade-like purpuric lesion over upper extremity

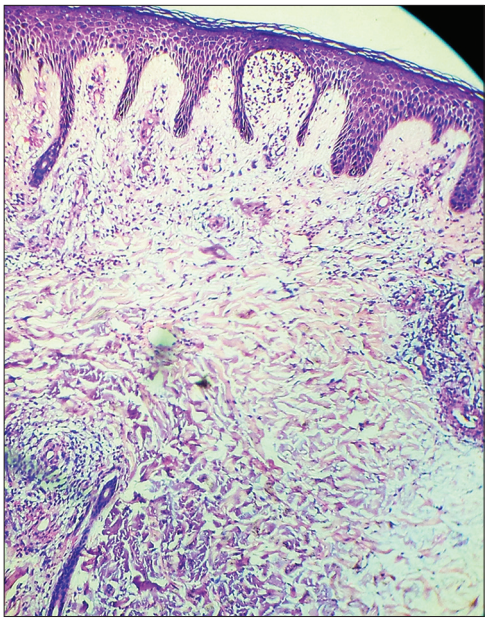

Figure 3: Leukocytoclastic vasculitis with fibrinoid necrosis and perivascular neutrophilic infiltrate (H and E, ×40)

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purpura, acral edema develops that is not tender and spreads proximally. Fever is usually associated with purpura and edema. Systemic involvement is rare with usually gastrointestinal tract (29%) and renal involvement (23%). It is necessary to distinguish this benign entity from henoch schonlein purpura, kawasaki disease, meningococcemia, urticaria, and erythema multiforme.[2,3] The age of onset in HSP is later age, i.e., after 5 years characterised by palpable purpura, abdominal pain, and arthritis. Erythema multiforme present with target lesion and does not show features of leukocytoclastic vasculitis. Urticarial vasculitis have chronic course with recurrences.

Treatment is usually supportive and proper counselling of parents about benign course and good prognosis of the disease is important. Spontaneous recovery usually occurs within 2-3 weeks.[1]

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