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# An Infant's Sudden-Onset Rash and Swelling: Can You Identify the Cause?

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A previously healthy 5-month-old boy presented to the emergency department (ED) within a few hours of the sudden onset of a rash on his hands, feet, and face along with painful bilateral swelling of the hands and feet. He also had a 4-day history of fever, congestion, and rhinorrhea. The child had no family history of bleeding disorders, and he lived with his parents, his grandmother, and 2 older brothers.

Gross inspection revealed ecchymosis on the infant's face (**Figure 1**) and his bilateral upper and lower extremities (**Figure 2**). Also present was swelling of the bilateral hands and feet, with tenderness to palpation. He was otherwise well-appearing.



Results of a complete blood cell count (CBC), a basic metabolic panel (BMP), urinalysis, and urine and blood cultures were unremarkable except for an elevated white blood cell (WBC) count of  $19,000/\mu\text{L}$ . The erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) level were normal. Radiographs of the pelvis and the bilateral lower extremities showed mild subcutaneous tissue swelling on the bilateral legs and feet without fractures. The patient was discharged from the ED with a scheduled next-day orthopedics outpatient follow-up.

The patient presented to the orthopedics clinic the next day with worsening discoloration of the legs and to buttocks and was directly admitted to the hospital. In the hospital, the results of additional workup, including coagulation tests, a skeletal survey, and repeated CBC, CRP, ESR, and BMP, were unremarkable except for a WBC count of  $20,600/\mu\text{L}$ .

Which one of the following is causing the infant's symptoms?

- A. Erythema multiforme
- B. Acute hemorrhagic edema of infancy
- C. Cutaneous drug reaction
- D. Kawasaki disease
- E. Nonaccidental trauma

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## Answer: Acute Hemorrhagic Edema of Infancy

A hematologist was consulted, who made the diagnosis of acute hemorrhagic edema of infancy (AHEI).

AHEI is an immune complex–mediated small-vessel leukocytoclastic vasculitis. It initially had been considered a variant of Henoch-Schönlein purpura (HSP) but is now classified as a separate entity.<sup>1-3</sup> Only approximately 300 to 500 cases of AHEI have been reported worldwide.<sup>3,4</sup>

AHEI presents most often in patients between 4 months and 2 years of age, with most cases preceded by a viral infection, an immunization, or antibiotic use.<sup>2-4</sup> Classic symptoms include a rapid onset of a rash (petechiae, ecchymoses, and annular, nummular or targetoid purpuric lesions) that involves the face, the ears, and/or the extremities with associated edema.<sup>2-5</sup> Patients with AHEI are nontoxic, with or without fever, and, unlike with HSP, rarely have extracutaneous involvement.<sup>3,4</sup>

Laboratory test results typically are normal but can include mild leukocytosis, thrombocytosis, elevated CRP and ESR levels, and slightly elevated liver enzyme levels.<sup>3,5</sup> Symptoms self-resolve within 1 to 3 weeks.<sup>4</sup> Histologic findings of a skin biopsy, if performed, will show small-vessel leukocytoclastic vasculitis.<sup>3-5</sup>

AHEI is usually a clinical diagnosis. Among the differential diagnoses are erythema multiforme, drug reaction, Kawasaki disease, meningococemia, and nonaccidental trauma.<sup>2</sup> Treatment is supportive with ibuprofen and acetaminophen.

### OUTCOME OF THE CASE

At a follow-up visit to the hematology clinic 1 month after his hospital discharge, the infant's symptoms had resolved.

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