VISUAL DIAGNOSIS

Ecchymotic Skin Lesions and Hand Edema in a 10-month-old Boy

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PRESENTATION

A 10-month-old boy presents to the emergency department with a 24-hour history of worsening targetoid papules and plaques over the face, trunk, and limbs. Some of the older lesions have become ecchymotic in appearance, and his hands are swollen. The eruption is nonpruritic and nonpainful. The patient is otherwise clinically well. There is no history of nausea, vomiting, or diarrhea. He completed a 7-day course of amoxicillin for community-acquired pneumonia, confirmed by chest radiography, the day before onset of the rash. His mother reports that she initially noticed a few pink papules on his face the day before presentation. When the lesions subsequently began to rapidly enlarge and spread to other parts of his body, she became alarmed and brought him in for assessment. His medical history is otherwise normal, and he is not taking any medications regularly. His immunizations are up to date.

On examination there are multiple erythematous to violaceous targetoid plaques with purpuric changes over the face, neck, trunk, and extremities (Figs 1 and 2). The child has edema of the dorsum of the left hand (Fig 3), extending from the wrist to the metacarpophalangeal joints, as well as edema of the ear pinnae bilaterally. He does not seem to be bothered by the presence of the skin lesions. He is moving all limbs appropriately and does not have any apparent joint effusions or joint tenderness. There is no periorbital edema, conjunctivitis, or mucous membrane involvement. Coryza is noted. The child is afebrile, and there is no lymphadenopathy. Abdominal examination findings are normal. Clinical examination confirms the diagnosis.

DIAGNOSIS

The patient has acute hemorrhagic edema of infancy (AHEI). The diagnosis can be made clinically based on the characteristic history and skin findings in an otherwise well child.

DISCUSSION

AHEI is classified as a cutaneous leukocytoclastic small-vessel vasculitis. It is a rare condition, and the exact etiology is unknown. Associated triggers include upper respiratory tract infections, antibiotic use, and vaccinations. (1)(2)(3) The pathogenesis may be related to immune complex deposition in the skin. (3) AHEI generally occurs in those younger than 2 years. Despite the dramatic skin appearance, most

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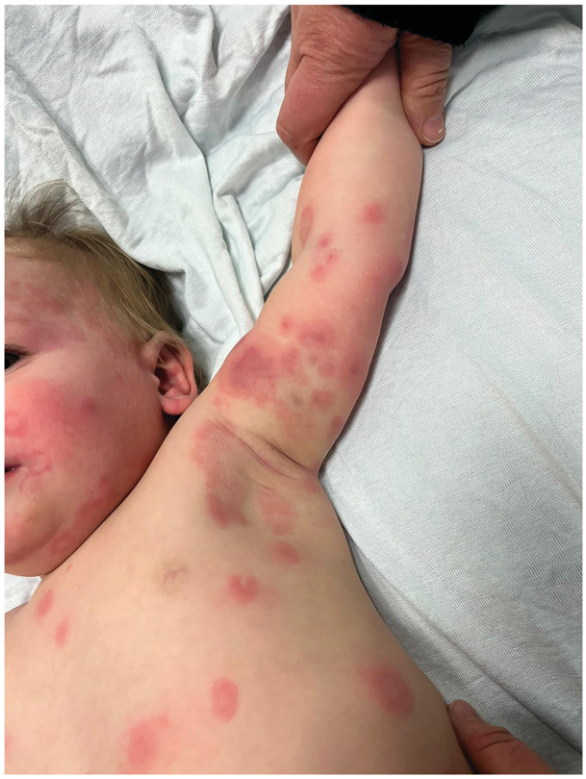


Figures 1 Classic presentation of acute-onset red, annular, purpuric plaques in a 10-month-old boy.

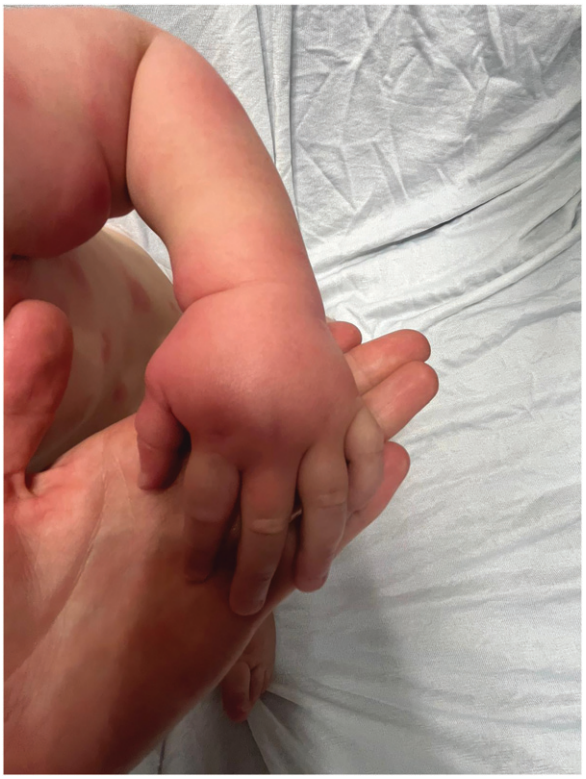


Figure 3. Image of the dorsal hand edema that was present in addition to the skin lesions.

affected children are otherwise systemically well. A low-grade fever may accompany the annular purpuric skin lesions and acral edema that are characteristic of this condition. (4)

A thorough history and physical examination should allow for distinction between this benign condition and other entities, such as Henoch-Schönlein purpura (HSP), urticaria multiforme, erythema multiforme (EM), serum sickness–like reaction, and child abuse. HSP is an IgA-mediated small-vessel vasculitis that tends to occur in children aged 4 to 8 years, with a predilection for males. The clinical manifestations of HSP include arthralgias, abdominal pain, and renal involvement, which are not observed in AHEI. (5) Urticaria multiforme lesions are transient, often pruritic, erythematous, edematous plaques that last less than 24 hours and resolve without ecchymotic and/or pigmentary alteration. Dermatographism may also be apparent. (6) The administration of antihistamines results in symptomatic relief and faster resolution of urticaria multiforme lesions but has no effect on the lesions of AHEI. (6) The target lesions seen in EM are also abrupt in onset, typically triggered by herpes simplex virus infection. EM tends to occur in older children and adults, but cases of infantile EM triggered by vaccinations have been reported. (7) The acral edema characteristic of AHEI is absent in patients with EM. (8) In addition to fever and malaise, serum sickness–like reaction is commonly associated with lymphadenopathy and arthralgias, which are typically absent in AHEI. (9) The resolving lesions of AHEI are ecchymotic in nature, as shown in Fig 3. As such, if a child presents for care later in the course of the cutaneous eruption, the bruiselike lesions may be mistaken for signs of nonaccidental injury. Finally, although rare, autoinflammatory conditions such as tumor necrosis factor receptor–associated periodic syndrome

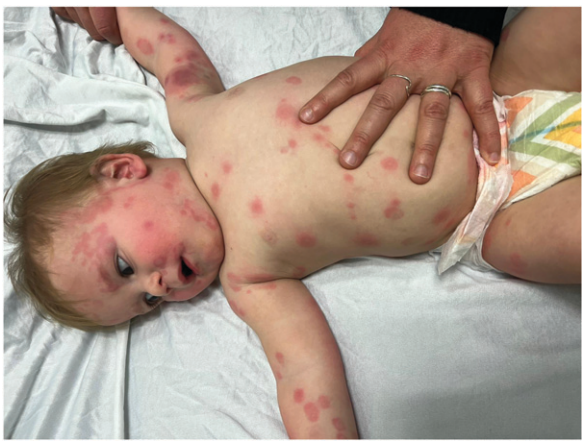


Figure 2. Classic presentation of acute-onset red, annular, purpuric plaques in a 10-month-old boy.

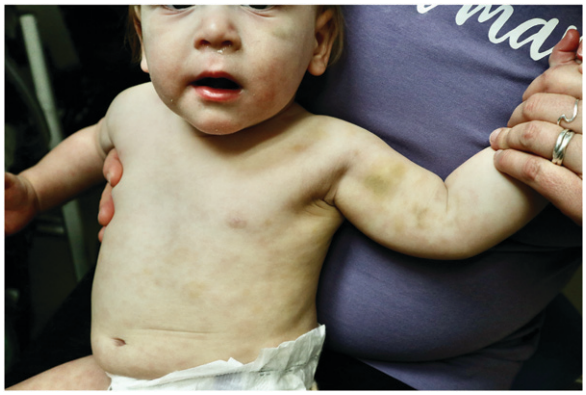
should also be kept in mind as potential mimickers of AHEI. (10) Specifically, recurrent episodes of AHEI-like lesions combined with extracutaneous symptoms should prompt a diagnosis other than AHEI.

An extensive evaluation is not necessary in classic cases of AHEI, as described herein. If performed, blood work results are generally nonspecific and may show leukocytosis and/or thrombocytosis with elevated inflammatory markers (C-reactive protein or erythrocyte sedimentation rate). (3) Urinalysis is typically normal. If a skin lesion is biopsied, routine histologic analysis would reveal typical findings of leukocytoclastic vasculitis (neutrophilic infiltrate/debris in the small vessels), and direct immunofluorescence is generally negative and demonstrates an absence of the IgA deposition seen in HSP. (3)

The eruption typically resolves spontaneously within 3 weeks without long-term sequelae. (1) Parents should be counseled about the self-limited, benign nature of this condition that requires only supportive care. However, appropriate follow-up should be arranged to ensure that the cutaneous eruption is resolving as expected.

PATIENT COURSE

Due to a high level of confidence in the clinical diagnosis of AHEI in this patient, no blood work or other investigations were ordered. The family was reassured about the benign, self-limited nature of the condition and the need for only supportive care. The patient was discharged with plans to follow up in the next few days. As a precaution, the family was instructed to return to the emergency department should the patient develop any additional worrisome features. At an outpatient dermatology follow-up appointment 2 days later, the child was still doing well clinically. His skin lesions were far less striking in appearance, with only some faint ecchymotic discoloration remaining (Fig 4). No recommendation to avoid amoxicillin in the future was made.



Figures 4 Follow-up image taken 2 days later showing evolution of the red edematous plaques to resolving yellow-brown ecchymotic patches.

Summary

- Acute hemorrhagic edema of infancy (AHEI) is an uncommon but very distinct entity that generally has a benign course.
- Its acute presentation and dramatic skin findings can alarm many health-care practitioners and lead to unnecessary testing and treatment.
- The distinct clinical features of this condition help differentiate AHEI from other worrisome entities.
- Laboratory findings (including biopsy) are nonspecific but may help rule out other diseases.
- Most patients have spontaneous resolution within 1 to 3 weeks and require only supportive care and reassurance for caregivers.

References for this article can be found at https://doi.org/10.1542/pir.2023-006114.