Hemorrhagic Lacrimation and Epistaxis: Rare Findings in Acute Hemorrhagic Edema of Infancy

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PRACTICE **POINTS**

- Acute hemorrhagic edema of infancy (AHEI) is clinically characterized by a triad of large purpuric lesions, low-grade fever, and peripheral acral edema. Although joint pain, bloody diarrhea, hematuria, and proteinuria can accompany AHEI, most cases are devoid of systemic symptoms.
- It is a self-limited condition typically lasting 1 to 3 weeks and requires only supportive care.
- On rare occasions, AHEI may be accompanied by hemorrhagic lacrimation and epistaxis. Patients should be assured that the condition is self-limited and resolves without permanent sequalae.

To the Editor:

Hemorrhagic lacrimation and epistaxis are dramatic presentations with a narrow differential diagnosis. It rarely has been reported to present alongside the more typical features of acute hemorrhagic edema of infancy (AHEI), which is a benign self-limited leukocytoclastic vasculitis most often seen in children aged 4 months to 2 years. Extracutaneous involvement rarely is seen in AHEI, though joint, gastrointestinal tract, and renal involvement have been reported.[1] Most patients present with edematous, annular, or cockade purpuric vasculitic lesions classically involving the face and distal extremities with relative sparing of the trunk. We present a case of a well-appearing, 10-month-old infant boy with hemorrhagic vasculitic lesions, acral edema, and an associated episode of hemorrhagic lacrimation and epistaxis.

A 10-month-old infant boy who was otherwise healthy presented to the emergency department (ED) with an acute-onset, progressively worsening cutaneous eruption of 2 days' duration. A thorough history revealed that the eruption initially had presented as several small, bright-red papules on the thighs. The eruption subsequently spread to involve the buttocks, legs, and arms (Figures 1 and 2). The parents also noted that the patient had experienced an episode of bloody tears and epistaxis that lasted a few minutes at the pediatrician's office earlier that morning, a finding that prompted the urgent referral to the ED.

Dermatology was then consulted. A review of systems was notable for rhinorrhea and diarrhea during the week leading to the eruption. The patient's parents denied fevers, decreased oral intake, or a recent course of antibiotics. The patient's medical history was notable only for atopic dermatitis treated with emollients and occasional topical steroids. The parents denied recent travel or vaccinations. Physical examination showed an afebrile, well-appearing infant with multiple nontender, slightly edematous, circular, purpuric papules and plaques scattered on the buttocks and extremities with edema on the

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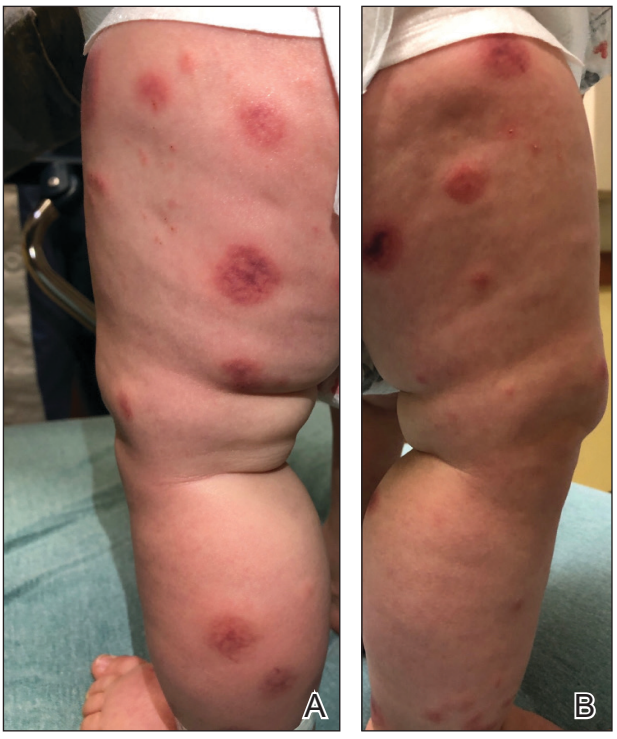

FIGURE 1. A, Targetoid hemorrhagic and purpuric lesions with scalloped margins of varying sizes involving the distal and proximal left leg. B, A targetoid hemorrhagic and purpuric lesion with scalloped margins and a necrotic center surrounded by additional coin-shaped lesions of varying sizes involving the distal and proximal right leg.

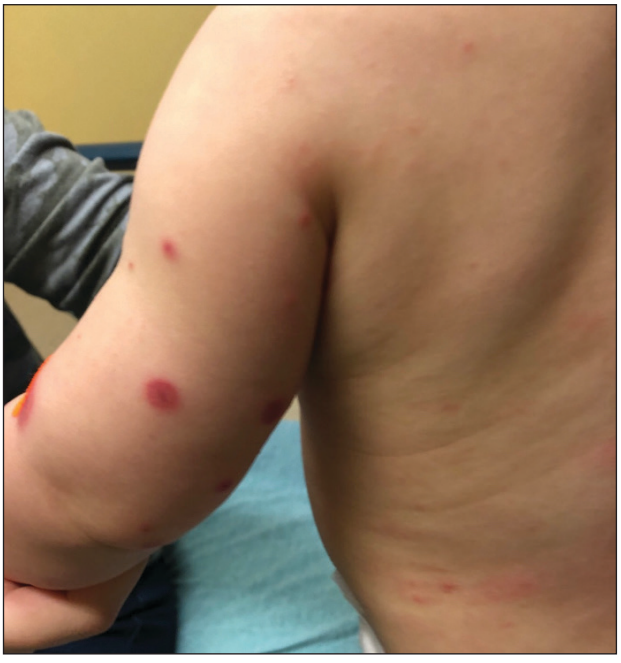

FIGURE 2. Several coin-shaped hemorrhagic lesions of varying sizes on the left arm.

dorsal feet. The remainder of the patient's workup in the ED was notable for mild elevations in C-reactive protein levels (1.4 mg/dL [reference range, 0–1.2 mg/dL]) and an elevated erythrocyte sedimentation rate (22 mm/h [reference range, 2–12 mm/h]). A complete blood cell count; liver function tests; urinalysis; and coagulation studies, including prothrombin, partial thromboplastin time, and international normalized ratio, were unremarkable. Acute hemorrhagic edema of infancy was diagnosed based on the clinical manifestations.

Acute hemorrhagic edema of infancy (also known as Finkelstein disease, medallionlike purpura, Seidemayer syndrome, infantile postinfectious irislike purpura and edema, and purpura en cocarde avec oedeme) is believed to result from an immune complex–related reaction, often in the setting of an upper respiratory tract infection; medications, especially antibiotics; or vaccinations. The condition previously was considered a benign form of Henoch-Schönlein purpura; however, it is now recognized as its own clinical entity. Acute hemorrhagic edema of infancy commonly affects children between the ages of 4 months and 2 years. The incidence peaks in the winter months, and males tend to be more affected than females.[1]

Acute hemorrhagic edema of infancy is clinically characterized by a triad of large purpuric lesions, low-grade fever, and peripheral acral edema. Edema can develop on the hands, feet, and genitalia. Importantly, facial edema has been noted to precede skin lesions.[2] Coin-shaped or targetoid hemorrhagic and purpuric lesions in a cockade or rosette pattern with scalloped margins typically begin on the distal extremities and tend to spread proximally. The lesions are variable in size but have been reported to be as large as 5 cm in diameter. Although joint pain, bloody diarrhea, hematuria, and proteinuria can accompany AHEI, most cases are devoid of systemic symptoms.[3] Hemorrhagic lacrimation and epistaxis—both present in our patient—are rare findings with AHEI. It is likely that most providers, including dermatologists, may be unfamiliar with these striking clinical findings. Although the pathophysiology of hemorrhagic lacrimation and epistaxis has not been formally investigated, we postulate that it likely is related to the formation of immune complexes that lead to small vessel vasculitis, underpinning the characteristic findings in AHEI.[4,5] This reasoning is supported by the complete resolution of symptoms corresponding with clinical clearance of the cutaneous vasculitis in 2 prior cases[4,5] as well as in our patient who did not have a relapse of symptoms following cessation of the cutaneous eruption at a pediatric follow-up appointment 2 weeks later.

Acute hemorrhagic edema of infancy is a clinical diagnosis; however, a skin biopsy can be performed to confirm the clinical suspicion and rule out more serious conditions. Histopathologic examination reveals a leukocytoclastic vasculitis involving the capillaries and postcapillary venules of the upper and mid dermis. Laboratory

test results usually are nonspecific but can help distinguish AHEI from more serious diseases. The erythrocyte sedimentation rate and C-reactive protein level may be slightly elevated in infants with AHEI. Urinalysis and stool guaiac tests also can be performed to evaluate for any renal or gastrointestinal involvement.[6]

The differential diagnosis includes IgA vasculitis, erythema multiforme, acute meningococcemia, urticarial vasculitis, Kawasaki disease, and child abuse. IgA vasculitis often presents with more systemic involvement, with abdominal pain, vomiting, hematemesis, diarrhea, and hematochezia occurring in up to 50% of patients. The cutaneous findings of erythema multiforme classically are confined to the limbs and face, and edema of the extremities typically is not seen. Patients with acute meningococcemia appear toxic with high fevers, malaise, and possible septic shock.[5]

Acute hemorrhagic edema of infancy is a self-limited condition typically lasting 1 to 3 weeks and requires only supportive care.[7] Antibiotics should be given to treat concurrent bacterial infections, and antihistamines and steroids may be useful for symptomatic relief. Importantly, however, systemic corticosteroids do not appear to conclusively alter the disease course.[8]

Acute hemorrhagic edema of infancy is a rare benign leukocytoclastic vasculitis with a striking presentation often seen following an upper respiratory tract infection or course of antibiotics. Our case demonstrates that on rare occasions, AHEI may be accompanied by hemorrhagic lacrimation and epistaxis—findings that can be quite alarming to both parents and medical providers. Nonetheless, patients and their caretakers should be assured that the condition is self-limited and resolves without permanent sequalae.

REFERENCES

1. Emerich PS, Prebianchi PA, Motta LL, et al. Acute hemorrhagic edema of infancy: report of three cases. *An Bras Dermatol*. 2011;86:1181-1184.
2. Avhad G, Ghuge P, Jerajani H. Acute hemorrhagic edema of infancy. *Indian Dermatol Online J*. 2014;5:356-357.
3. Krause I, Lazarov A, Rachmel A, et al. Acute haemorrhagic oedema of infancy, a benign variant of leucocytoclastic vasculitis. *Acta Paediatr*. 1996;85:114-117.
4. Sneller H, Vega C, Zemel L, et al. Acute hemorrhagic edema of infancy with associated hemorrhagic lacrimation. *Pediatr Emerg Care*. 2021;37:E70-E72. doi:10.1097/PEC.0000000000001542
5. Mreish S, Al-Tatari H. Hemorrhagic lacrimation and epistaxis in acute hemorrhagic edema of infancy. *Case Rep Pediatr*. 2016;2016:9762185. doi:10.1155/2016/9762185
6. Savino F, Lupica MM, Tarasco V, et al. Acute hemorrhagic edema of infancy: a troubling cutaneous presentation with a self-limiting course. *Pediatr Dermatol*. 2013;30:E149-E152.
7. Fiore E, Rizzi M, Ragazzi M, et al. Acute hemorrhagic edema of young children (cockade purpura and edema): a case series and systematic review. *J Am Acad Dermatol*. 2008;59:684-695.
8. Fiore E, Rizzi M, Simonetti GD, et al. Acute hemorrhagic edema of young children: a concise narrative review. *Eur J Pediatr*. 2011;170:1507-1511.