Acute hemorrhagic edema in a nursing infant - An unusual diagnosis

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ABSTRACT

Acute hemorrhagic edema of infancy is a rare, small vessel vasculitis of young children with characteristic skin findings. We have described an 8-month-old male infant who presented with upper respiratory infection followed by acute onset of generalized swelling of both lower limbs and appearance of erythematous, purpuric, and ecchymotic lesions on the lower limbs, face, and ear lobules. There were no systemic complications and the lesions resolved completely.

Key words: *Acute hemorrhagic edema of infancy, Purpura, Vasculitis*

Acute hemorrhagic edema of infancy (AHEI), also called infantile post-infectious cockade purpura, Finkelstein's disease, or Seidlmayer syndrome [1], is a rare form of leukocytoclastic vasculitis affecting children younger than 2 years of age [2]. It is clinically characterized by the triad: Fever, purpuric/ecchymotic skin lesions, and edema on the extremities and/or face [2]. The cutaneous findings are dramatic, both in appearance and rapidity of onset, and may cause significant anxiety for parents and clinicians [2]. Since 1913, when AHEI was first reported in the United States by Snow, only about 100 cases have been described in the literature [3]. The recognition of AHEI allows making the diagnosis of a benign entity rather than a more serious disease [1]. We report this case to increase awareness regarding this uncommon but benign form of vasculitis.

CASE REPORT

This 8-month-old male infant presented with acute onset of edema starting from the feet, progressing proximally, and involving both lower limbs within one day. The child's mother also noticed erythematous palpable spots over both lower limbs after the appearance of edema. There was a history of mild fever, cough, and cold for 5 days preceding the onset of edema and skin lesions. There was no history of septic focus, drug intake, hematuria, decreased urine output, diarrhea, refusal to feed, or recent vaccination. No history of similar illness was found in the family. On examination, the child was irritable but alert with mild fever and stable vitals. There was diffuse non-pitting, non-tender, tense edema of both the lower limbs; along with multiple erythematous, palpable, purpuric, and ecchymotic lesions over both the legs, feet and gluteal region varying in size from 1 to 5 cm in diameter (Fig. 1). There was no mucosal involvement. Systemic examination was normal except for features of viral upper respiratory infection without any tachypnea, and the infant was feeding well from the breast. During the hospital course, edema spread proximally to involve most of lower limbs over the next several hours.

New purpuric lesions continued to appear subsequently over the next 3–5 days over thighs, buttocks, face, and ear lobules sparing the trunk without any deterioration in the infant's health. Complete blood count and clotting screen were normal. Blood urea, creatinine, electrolytes, and urinalysis were normal. Blood culture was sterile, and chest X-ray was normal. Skin biopsy showed the features of leukocytoclastic vasculitis. Immunofluorescence examination could not be done. The patient was given symptomatic treatment. The skin lesions and edema resolved completely within 10 days.

DISCUSSION

AHEI is an uncommon form of cutaneous vasculitis that typically occurs in children between 4 months and 2 years of age. Most of the cases occur in the winter, probably precipitated by upper respiratory tract infections, immunizations, and hypersensitivity to drugs [4]. AHEI was once considered a purely cutaneous variant of Henoch–Schonlein purpura (HSP) but today is recognized as a distinct entity [1].

Krause *et al*. [5] proposed clinical criteria for the diagnosis of AHEI: Age <2 years, sudden appearance of well-demarcated annular, rosette-like, or targetoid purpuric or ecchymotic lesions, located primarily on the face, ears, and extremities with sparing of the trunk (Fig. 2). The lesions are accompanied by edema of the extremities. The most remarkable feature of AHEI is the contrast between the cutaneous signs, which are typical, and the child's overall good health [1]. Systemic affection and recurrence of lesions are rare; there is a spontaneous resolution within 1–3 weeks. Our case fulfills all above the above criteria.

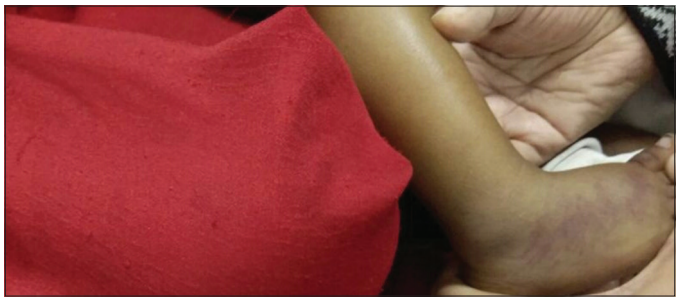

Figure 1: Diffuse inflammatory acral edema with violaceous ecchymotic lesion on the foot

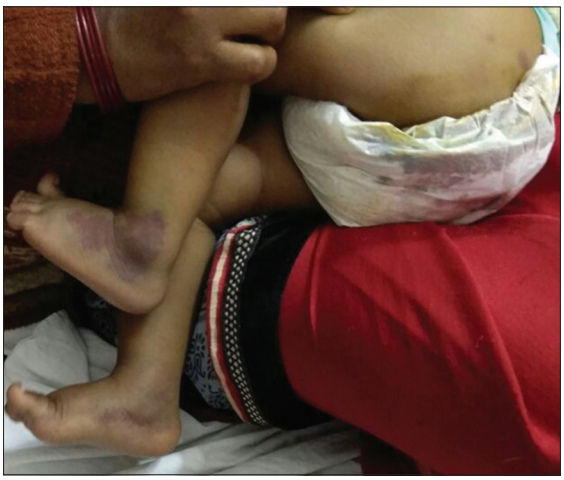

Figure 2: Large cockade (rosette-like) ecchymotic lesion characteristic of acute hemorrhagic edema of infancy

Eosinophilia, leukocytosis, and thrombocytosis may be found. Coagulation tests, kidney and liver function, and urine analysis are normal. On histopathological examination of the skin, the findings are leukocytoclastic vasculitis affecting small dermal vessels and fibrinoid necrosis with perivascular neutrophilic infiltrate with abundant nuclear dust [1]. Although the histopathological findings in HSP are similar to AHEI, the lack of vessel wall staining for IgA on direct immunofluorescence is diagnostic for AHEI [2].

The clinical features may be confused with HSP, erythema multiforme, meningococcemia, and septicemia [2]. All these entities are not difficult to differentiate from AHEI clinically. HSP affects mostly children between 4 and 7 years of age. The lesions are mainly palpable purpura on extensor aspect of lower limbs and rarely present subjacent edema. Systemic complications are often seen in HSP and are extremely rare in AHEI [4].

No specific treatment is required for AHEI [2]. Antibiotics may be given if there is a secondary infection; otherwise, the treatment is supportive. The role of corticosteroids is controversial [2]. Although the cutaneous findings are dramatic and of rapid onset, the disease runs a benign course. Recurrences are rare [1].

CONCLUSION

AHEI is a rare, small vessel vasculitis of young children with characteristic skin findings. A high index of suspicion is required to diagnose this condition.

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