Acute hemorrhagic edema of infancy-a rare entity

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ABSTRACT

A patient presenting with fever and purpura often presents a diagnostic challenge for the dermatologist. Acute hemorrhagic edema of infancy (AHEI) is a rare acute cutaneous leukocytoclastic vasculitic syndrome of infants leading to this presentation. We present an 18-month-old girl with AHEI who presented with fever, edema, and purpuric lesions involving the face, ears and extremities with uneventful complete recovery.

Key words: Acute hemorrhagic edema, cockade purpura, henoch-schönlein purpura, leukocytoclastic vasculitis

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INTRODUCTION

AHEI, also called postinfectious cockade purpura, Finkelstein's disease and Seidlmayer's disease is a leukocytoclastic vasculitis characterized by inflammatory acral edema and ecchymotic purpura in a target or cockade pattern usually without visceral involvement. The cutaneous findings of AHEI resemble those of Henoch-Schönlein purpura (HSP). Infection, drugs and immunization have been considered as precipitating factors. Although some have suggested considering AHEI a purely cutaneous variant of HSP, most authors regard it as a separate entity amongnst cutaneous small vessel vasculitis of childhood. The recognition of AHEI allows making the diagnosis of a benign entity rather than a more serious disease.

CASE REPORT

An 18 month-old girl was referred from the paediatric department for purpuric skin lesions since 3 days. She had an upper respiratory infection 5 days prior to the development of the skin lesions. Her mother reported seeing reddish colored spots initially on the legs which gradually increased to involve the face, legs and arms. There was no recent history of vaccination or any drug intake. On general examination the child was irritable and mildly febrile. The cutaneous examination showed many symmetrically distributed, round and oval, ecchymotic, purpuric, targetoid plaques localized on the cheeks and extremities, in particular lower limbs. The ears appeared oedematous [Figure 1]. Soles showed violaceous targetoid macules [Figure 2]. Some plaques showed a cockade pattern [Figure 3], while others coalesced to form large purpuric lesions with polycyclic borders. Moderate non-pitting edema was noted over her hands and feet [Figure 4]. There was no mucosal involvement. The remainder of the physical examination was unremarkable and there were no signs of systemic involvement. Urinalysis, liver and renal function test, and complete blood count were within normal limits. Stool was negative for occult blood. Erythrocyte sedimentation rate was elevated. Histopathological examination of cutaneous lesion showed features of leucocytoclastic vasculitis [Figure 5]. The skin lesions, as well as fever resolved without treatment within two weeks.

DISCUSSION

The clinical features of our case, characterized by a dramatic onset of typical symmetrical, annular purpuric plaques on the face, ears and limbs, histology showing leukocytoclastic vasculitis and a spontaneous recovery within two weeks, were consistent with the diagnosis of AHEI. A clinical disorder corresponding to AHEI was first described in 1913 by Snow, *et al*. who reported a patient as "Purpura, urticaria and angioneurotic oedema of the hands and feet in a nursing baby".[1] AHEI is a disease that affects children between 4 and 24 months of age and is characterized by a triad

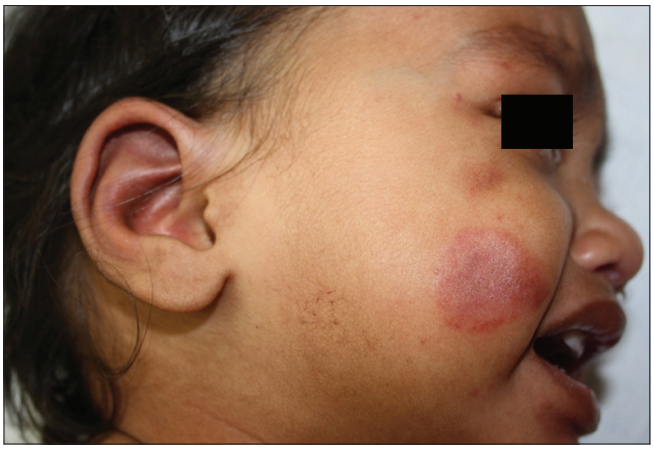

Figure 1: Round to oval purpuric targetoid plaque on the face with redness and edema of right pinna

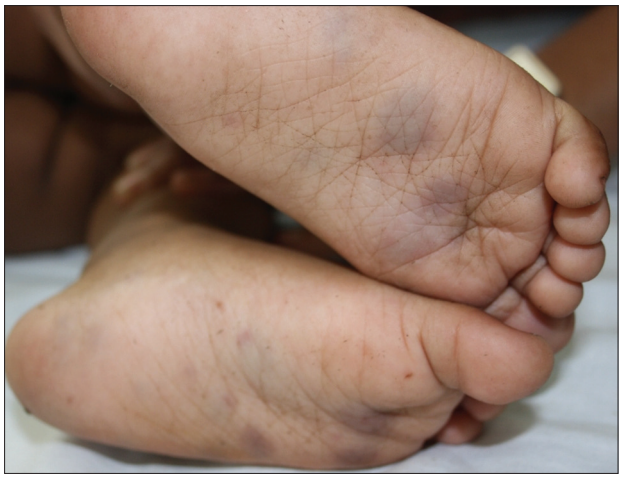

Figure 2: Violaceous plaques on both soles

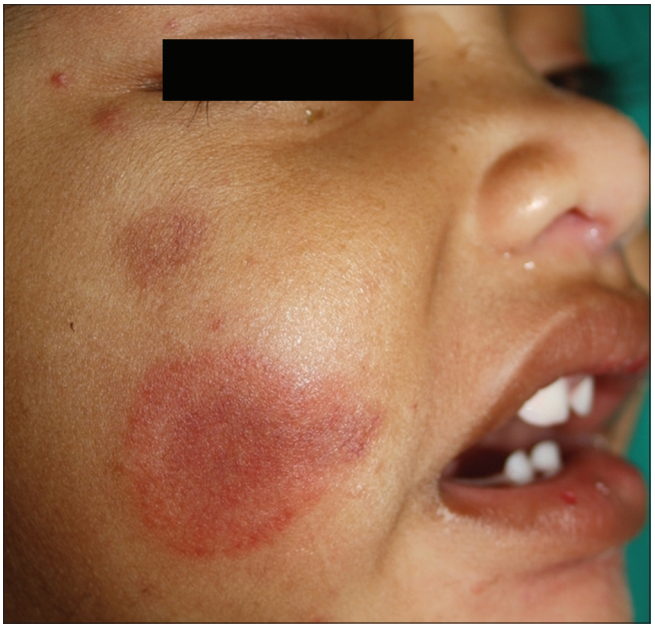

Figure 3: Close up of targetoid plaque on cheek

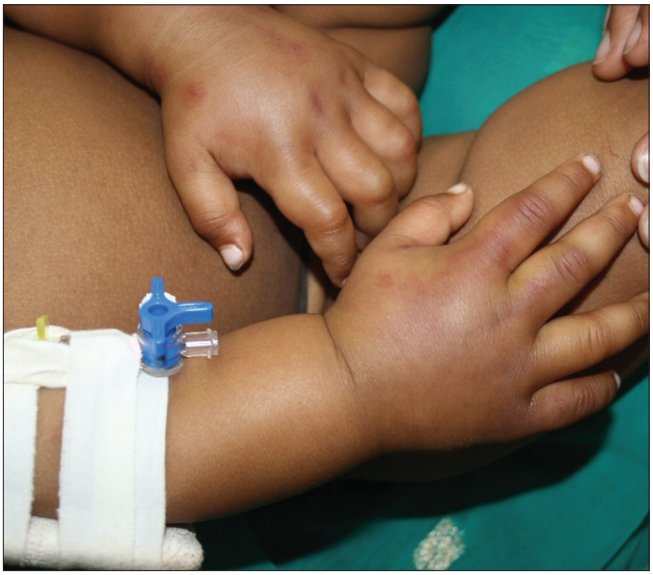

Figure 4: Inflammatory acral oedema

of fever, large palpable purpuric skin lesions, and edema.[2] Classically, there is a contrast between the dramatic skin findings and relative absence of ill health. It starts with a sudden appearance of well demarcated, annular, medallion-like, rosette shaped purpuric plaques, almost entirely limited to extremities and face, with relative sparing of trunk. Fever and painful edema of the distal extremities, ears, and eyelids are associated features of AHEI.[3] Since there are no diagnostic laboratory tests, the history and physical examination provide clues to the successful recognition of the disease. Its aetiology remains unknown, although a history of recent upper respiratory or urinary tract infection treated with antibiotics or immunizations is found in 75% of patients.[4] Thus, AHEI is considered to represent an immune-complex mediated disease. The most reported infective agents include Staphylococci, Streptococci and among viruses, *Adenovirus*, although many other agents, such as *Escherichia coli*, or *Mycobacteria*, have been reported. A history of drug intake before the onset of the cutaneous eruption is present in many cases of AHEI. The drugs include various antibiotics and anti-inflammatory drugs.[5] Visceral involvement is rare but it has been reported involving the kidneys and intestines, causing symptoms such as hematuria, mild proteinuria, and bloody diarrhea.[6] Laboratory tests typically show normal results. Elevated erythrocyte sedimentation rate, leucocytosis, thrombocytosis, and eosinophilia have been seen in AHEI.[7] Histologic features of AHEI are consistent with small-vessel vasculitis of both capillaries and post capillary venules of the upper and the middle dermis, showing typical leukocytoclastic vasculitis with or without fibrinoid necrosis and a deep perivascular and interstitial infiltrate composed mostly of neutrophils with abundant nuclear dust.[5] The disease runs a benign course with complete spontaneous recovery occurring in 1-3 weeks,

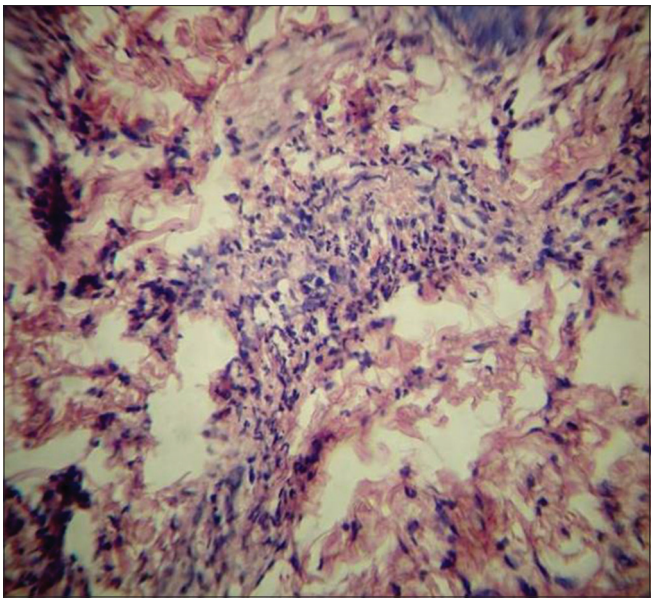

Figure 5: Histopathology showing fibrinoid vessel wall necrosis with perivascular neutrophilic infiltrate and leucocytoclasis

Table 1: Differences between AHEI and HSP

	AHEI	HSP
Age	4-24 months	Peak between 3 and 6 years
Purpura	Ecchymotic cockade pattern: Limbs and face	Papular, Petechial, urticarial: Predominantly lower legs
Edema	Constant, often extensive	Inconsistent
Visceral Involvement	Uncommon	Frequent
Leucocytoclastic vasculitis	+	+
Fibrinoid necrosis	Frequent	Uncommon
Perivascular IgA deposits	Often negative (30% +)	+
Perivascular C1q deposits	+	-
Mean duration	12 days	30 days
Relapses	No	Frequent

AHEI: Acute hemorrhagic edema of infancy, HSP: Henoch-schönlein purpura

although relapses have been reported rarely.[2,5] Treatment with oral corticosteroids has been reported, but this is unnecessary due to its spontaneous recovery.[8] It is important to remember that purpura in infants needs complete work up to exclude more serious conditions like HSP, meningococcemia, septicemia, Kawasaki disease, erythema multiforme and purpura fulminans.

Amitai *et al*. suggested that AHEI cannot be considered a distinctive syndrome, but a benign variant of HSP pointing out a similar pathogenesis of both diseases, but a different distribution of purpuric lesions and with predilection of the face in infants (AHEI) and buttocks and lower extremities in older children (HSP). In addition, they postulated that the proportionally larger head and face in infant with a corresponding increase in blood supply would render them more susceptible to facial purpura.[9] Cases of AHEI and HSP overlap have also been reported. However, most authors consider these to be distinct clinical and pathological entities.[5] The differences between AHEI and HSP have been summarised in the Table 1.[10]

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