

# Acute Hemorrhagic Edema of Infancy: Report of Two Cases Report

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## Abstract

**Introduction:** Acute hemorrhagic edema of infancy (AHEI) is a cutaneous leukocytoclastic vasculitis of infants, clinically characterized by acute development of peripheral edema and targetoid purpuric lesions on face and extremities. It is considered to be an uncommon form of cutaneous vasculitis occurring in children younger than 2 years old.

The clinical picture has a violent onset, a short benign course followed by spontaneous complete recovery.

**Case presentation:** We describe two males with acute hemorrhagic edema. In one case the disease appeared after upper respiratory tract infection.

**Conclusion:** AHEI is a benign disorder despite its dramatic appearance. The most striking classic features of the disease are the contrast between the acuteness of the cutaneous lesions and the good general condition of the patient. Considering its clinical features, AHEI can be justifiably characterized as a unique disorder, distinct from Henoch–Schönlein Purpura (HSP).

**Key words:** Acute hemorrhagic edema, Infancy, Vasculitis, Leukocytoclastic

## Introduction

Acute hemorrhagic edema of infancy (AHEI) is a form of benign cutaneous leukocytoclastic vasculitis.<sup>1</sup> It usually affects children younger than two years of age. AHEI is characterized by fever, peripheral edema, and rosette-shaped purpuric plaques and shows leukocytoclastic vasculitis of small dermal vessels on histological examination.<sup>2</sup> AHEI has some common clinical features with Henoch-schönlein purpura (HSP), and is considered by some authors to be a variant of this syndrome<sup>3</sup>, but in patients with AHEI most of the time there is only cutaneous involvement rather than visceral, thus it differs from HSP. The diagnosis of

the syndrome is based on clinical symptoms and definitive diagnosis of vasculitis when the clinical picture is typical. It is no need for skin biopsy to confirm the diagnosis of vasculitis. The course is generally favorable without requiring treatment and it has a good prognosis ending in complete cure.

## Report of cases:

### Case 1

A 9-month-old male presented with erythematous plaques on his cheeks, ears and ecchymotic lesions on the distal part of his legs

**Fig 1.** erythematous plaques on the leg



from 2-3 days before admission (Fig 1, 2). He had a history of swelling of legs and periorbital area from 1 day before admission. He also had a 5 day history of upper respiratory infection and fever approximately 2 weeks prior to admission, taking diphenhydramin and acetaminophen and cephalixin for 1 week.

Laboratory findings revealed a white blood cell count of 13000/mm<sup>3</sup>. C-reactive protein was 3+ and erythrocyte sedimentation rate was 85. Other blood chemistry profiles were within normal range including Prothrombin Time: 13, Partial Thromboplastin Time: 27.

The patient was treated with oral ketotifen, which resulted in rapid fading of the lesions and complete recovery within 10 days, physical examination was normal in follow up during next 20 days.

### Case 2

A 14-month-old male presented with violaceous edematous purpuric rash on his lower extremities and edematous purpuric plaques on his face (Fig 3).

Laboratory studies revealed the following: hemoglobin 9.2gr/dl, leukocytes 6300/mm<sup>3</sup> (20% neutrophils, 77% lymphocytes, 3% Eosinophils),

Erythrocyte Sedimentation Rate: 65.

Clinical features of our patients were consistent with acute infantile hemorrhagic edema.

The patient was treated with oral ketotifen, which resulted in rapid fading of the lesions and complete recovery within 10-14 days.

## Results

The ages of our patients ranged between 9-14 months and they were both male. AHEI episodes in our patients occurred during the winter and one patient had a history of recent upper respiratory infection and fever. Leukocytosis was noted in one patient and an increased erythrocyte sedimentation rate was also noted. Complete recovery time ranged between 10-14 days. We believe that AHEI is a benign disorder, despite its dramatic appearance and routine laboratory test are non-diagnostic. There is no specific treatment for AHEI.

When the clinical picture is typical there is no need for skin biopsy to confirm the diagnosis of vasculitis.<sup>4</sup>

## Discussion

Acute hemorrhagic edema of infancy is a variant

Fig 2. erythematous plaques on the cheek



Fig 3. Purpuric rash on the lower extremities



of leukocytoclastic vasculitis and presents clinically as petechiae and ecchymoses which become edematous and develop a target like appearance. This usually occurs in infants younger than 2 years old.<sup>5,6</sup> It was first described by Snow<sup>7</sup> in the United States in 1913. Since then, many cases have been reported in European literature under different clinical terms, including Finkelsten disease<sup>8</sup> and Seidlmayer's "Cockade" purpura or syndrome.<sup>9</sup> Until now, approximately 80 cases of AHE have been reported in the literature, although the disease may be underreported.<sup>3,6,10</sup>

Its etiology remains unknown, although a history of recent upper respiratory or urinary tract infection or immunization is found in 75% of patients.<sup>11-13</sup> Thus AHEI is believed to represent an immune complex-mediated disease. Of interest is that our 9-month-old patient had a history of upper respiratory infection. The most often reported infective agents include staphylococci, streptococci and adenovirus, although many other agents, such as *Escherichia coli* and mycobacteria, have been reported in association with AHEI.

A history of drug intake before the onset of the cutaneous eruption is present in many cases of AHEI. These drugs include various antibiotics and anti-inflammatory.<sup>6-13-14</sup> Conditions that should be considered in the differential diagnosis are Sweet's syndrome, meningococemia, erythema multiform, Kawasaki disease, purpura fulminans, and trauma induced purpura. These disorders can be differentiated from AHEI history, physical examination and appropriate laboratory studies, including examination of the skin biopsy specimen.

Acute hemorrhagic edema of infancy should especially be distinguished from HSP. Vasculitis is revealed in skin biopsy of those cases who have doubtful clinical features.<sup>15</sup> Since there was a typical clinical picture in our 2 cases, no skin biopsy was required.

Some authors have speculated that AHEI and HSP are separate entities, whereas others believe that they are different expressions of the same disorder and that the unifying factor is a hypersensitivity vasculitis.<sup>5</sup>

The prognosis of the disease is usually good and the evolution is self-limited, lasting from one to three weeks.<sup>16</sup>

The clinical features of our patients, characterized by a dramatic, acute onset of typical

large purpuric plaques on the face, ears and limbs that resolved spontaneously within 2 weeks, were consistent with the diagnosis of acute hemorrhagic edema. Routine laboratory tests of patients with acute hemorrhagic edema are not diagnostic, disclosing normal results, or as in our patients, an elevated ESR.

#### Conclusion:

AHEI is a benign disorder despite its dramatic appearance. The most striking classic features of the disease are the contrast between the acuteness of the cutaneous lesions and the good general condition of the patient. Considering its clinical features, AHEI can be justifiably characterized as a unique disorder, distinct from Henoch-Schönlein purpura (HSP).

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