Acute Hemorrhagic Edema of Infancy: A Case Report

Ali Tariq Abd Al Hussan, Fatima Abd Allah Al Hassany

Dermatology Department/Al-Husseiny Teaching Hospital/ Krbala, Iraq

Abstract

Acute hemorrhagic edema of infancy (AHEI) is an uncommon form of eukocyteclastic vasculitis that affects children younger than 3 years and frequently is preceded by drug intake, vaccination, or a variety of infections. It is characterized by an abrupt onset of fever, purpuric lesions, and peripheral edema on the face and extremities & ears. The course is benign with spontaneous resolution.

Key words: Acute hemorrhagic edema, infancy.

Introduction

Snow first described acute hemorrhagic edema of infancy (AHEI) as a purely cutaneous variant of Henoch-Schönlein purpura (HSP) in the United States in 1913 (1). Europeans have recognized Finkelstein’s description of AHEI since his publication in 1938 and until recently, most reports of AHEI occurred in the European literature under the terms Finkelstein’s disease (2).

The etiology of AHEI is not very clear; 6 of 8 patients had history of recent infection, drug administration or immunization (3). AHEI is an uncommon disease. This may reflect either a low incidence or underdiagnosis. Another reason for its rarity could be that it is considered to be a variant of Henoch Schönlein purpura with only skin involvement (4).

Although some have suggested that AHEI is a purely cutaneous variant of Henoch-Schönlein purpura (HSP), most authors prefer to regard it as a separate clinical entity among the cutaneous small vessel vasculitic diseases of childhood.

The Case Report

A 13 months old baby presented at the dermatology outpatient at Al Hussein Teaching Hospital at Krbala city. His mother said that he was feverish three days before but now he is doing well. He had received drugs for his fever, no history of antibiotic or vaccination was revealed.

Physical examination revealed that multiple erythematous papules with central vesicles were present all over the body with purpuric target-like skin lesion involving the face and back of wrist joint, the knee joints and the ankle joints in symmetrical distributions. Both ears were involved, this purpuric lesions associated with edema especially in the hands and foot (Figure 1, 2 & 3).

The baby was admitted to the pediatric hospital, where complete investigations were done for him including complete blood count, ESR, coagulation tests, chest x-ray, abdominal ultrasound, general urine examination. All results of investigations were normal. Our case was a typical case of AHEI without any complications, so a skin biopsy was not necessary.

Discussion

The clinical features of our patient, characterized by a dramatic, acute onset of typical large, symmetrical, annular purp puric plaques on the face, ears, and limbs
that resolved spontaneously within 2 weeks, were consistent with the diagnosis of AHEI. Routine laboratory tests of patients with AHEI are not diagnostic, disclosing normal results.

Since it is a leukocytoclastic vasculitis, there are authors who consider AHEI to be a variant of HSP (5).

Figure-1 Target like purpuric lesion involving the cheeks and ears.

Figure-2 Purpuric lesion involving the knee and ankle joints with edema of foots

Figure-3 Purpuric target lesions involving the wrist joints with edema of the hands
Acute hemorrhagic edema of infancy: a case report

On the other hand, given the early age of onset, benign clinical course that generally does not require treatment other authors consider AHEI to be a different entity from the classic HSP (6).

Approximately 100 cases have been reported in the English and European literature since the disease was first described. The low number of reported cases may represent low incidence, disease underdiagnosis, or confusion with other similar entities (7).

There is a slight male predominance and the majority of the cases have been seen to occur in winter (8, 9). This coincided with our case. The head and distal portion of the extremities are preferred sites in AHEI, and the disease is limited to the skin (10). Visceral involvement is rare but it has been reported involving the kidneys and intestines, causing symptoms such as hematuria, mild proteinuria, and bloody diarrhea (7).

Other systemic symptoms such as abdominal pain, gastrointestinal bleeding, arthritis, and nephritis, have been rarely reported (11). Severe articular involvement has been described in one case (12). The disease had been reported as not involving the mucosa, but rare cases have demonstrated mucosal involvement (8, 13).

Also, one case of severe genital and trunk involvement has been described. 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 AHE.

Diarrhea has been observed during episodes of AHE and related to Coxsackie virus (14, 15).

Our 13-month-old patient had no gastrointestinal symptoms during the skin eruption just fever without evidence of tonsillitis or any sign of upper respiratory tract infections.

We must remember this entity wherever we faced with purpuric lesion in infant within a context of male patient with fever without evidence of coagulopathy or nephrological problems.

Acute hemorrhagic edema of infancy should especially be distinguished from HSP, acute febrile neutrophilic dermatosis, meningococcemia, septicemia, purpura fulminans, child abuse, Kawasaki disease, and other diseases leading to cockade eruptions, such as erythema multiforme and urticaria (16, 17, and 18).

References

Acute hemorrhagic edema of infancy: a case report

Ali Tariq Abd Al Hassan