

Case Report

Acute hemorrhagic edema of infancy: A case report

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Abstract Acute hemorrhagic edema of infancy (AHEI) is a rare skin disorder introduced for the first time by Snow (1913). The disorder which usually presents exclusively with skin manifestations occurs mostly in less than 2-year-old-age children. Most of the patients have suffered from a background of upper respiratory tract infection and have experienced vaccination or antibiotic treatment before beginning of the symptoms. Clinical sign of the disease is including petechiae, ecchymosis, and edema in head, face, and extremities. We present a typical case of AHEI in a 6-month-old infant.

Key words

Acute hemorrhagic edema, infancy.

Case report

The patient, the second child of family, was a 6-month-old boy who had fever two days prior to admission. After appearance of skin lesions in the form of ecchymotic and edematous plaques with high progressing trend, he was visited by a pediatrician and later referred for a dermatological consultation. The child, developed edema in face, ears, hands, feet, and scrotum, with ecchymotic plaques, in various sizes (**Figures 1 and 2**). In the centre of a facial lesion, a small necrotic area was visible (target sign) [**Figure 3**].

In spite of a low grade fever, the patient's general condition was well and lesions were painless throughout. Scrotum showed some edema with ecchymosis. Mouth and nasal mucosae were normal, and there was no sign of internal organ involvement. In past history, the

only important point was a common cold in the previous week, for which an antihistamine and acetaminophen syrup was prescribed.

Complete blood count, CRP, anti-streptolysin (ASO) titer, urinalysis, coagulation tests, liver and kidney function tests were normal. Punch biopsy from one of lesions showed a leukocytoclastic vasculitis of small vessels.

Keeping self-limited nature of this disorder, simple care of the lesions was undertaken. One week after the first visit, most of the lesions were remarkably cured and the patient's general condition was well (**Figure 4**).

Discussion

Acute hemorrhagic edema of infancy (AHEI) is a rare skin disorder.^{1,2} It was introduced for the first time in the United States in 1913 by Snow under the title of "Purpura, urticaria and angioneurotic edema of the hands and feet in a nursing baby". However, there is no report indicating the prevalence of the disease in the United States.^{1,3} Acute hemorrhagic edema of infancy is a leukocytoclastic vasculitis of

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Figure 1 Ecchymotic plaques on the face.



Figure 2 Ecchymotic plaques on the thighs.



Figure 3 Necrotic area in the center of purpuric plaque on right cheek – target sign.



Figure 4 Complete healing of lesions after one week.

cutaneous small vessels with characteristic clinical features and outstanding dermal findings in the very young children. The two clinical signs of the disease are purpuric cockade and circular (target) lesions, which mostly appear on face, ears, and extremities, in addition to the edema, which affects the face and extremities, as well.

The typical patient is an infant 2-24 months old who experienced an upper respiratory tract infection or received antibiotic.¹ However, one case has been reported in an infant from the birth.⁴ AHEI is an immune complex-mediated vasculitis which initiates from bacterial or viral infections (mostly related to the upper respiratory tract and urinary system) in addition to some medications specially antibiotics and, with less probability, vaccination.⁵

The peak prevalence of this disorder is in the winter, which may be related to the upper respiratory tract infection in the most cases of the disease.¹ Males are more susceptible than females.⁶

The clinical signs of the disease are very diagnostic, since in spite of the fact that the progression of disease is acute, the patient's general condition is well.⁷ Painless edema of

face, mostly asymmetric, may be the first sign of AHEI. Then painful ecchymoses and petechiae appear on the face and extremities or large target lesions may erupt suddenly. Skin lesions begin from the extremities and spread proximally. Sometimes involvement of scrotum is also observed in males. There is no specific trunk involvement. Although, fever might be present, the patient's general condition is well. Involvement of joints, gastrointestinal tract like bloody diarrhea and kidneys are rarely seen due to vasculitis.⁸

Histologic changes of the disease are based on a leukocytoclastic vasculitis, with neutrophilic infiltration of the walls of small vessels. Inflammation of the endothelial cells and fibrin deposition is observed around the vessels. RBCs extravasation is also a typical feature.

The main differential diagnosis of AHEI is Henoch-Schoenlein purpura (HSP), and the differentiation of these two diseases from each other is not difficult. The age of onset of AHEI (2-24 months) is lower as compared to the age of HSP (4-7 years). Systematic complications like arthralgia, GI bleeding and nephritis are common in HSP; whereas these complications are rare in AHEI.⁹ HSP is distinguished clinically by palpable purpura upon extensor surface of legs and buttock, whereas in AHEI face and the extremities are affected by ecchymoses and is accompanied by severe widespread edema.¹⁰ Both of these lesions are leukocytoclastic vasculitis, however, immunohistologic pattern in AHEI is different from the pattern in HSP. In AHEI, severe vasculitis with fibrin deposition is observed, and in some cases IgA deposits are found, as well. AHEI, like HSP, may show IgM, fibrinogen, and C3 deposition. AHEI, also shows perivascular deposition observed in HSP.¹¹ A

case of AHEI and HSP overlapping was also reported in a child.¹²

In addition to HSP, differential diagnosis of AHEI includes meningococemia, erythema multiforme, urticarial vasculitis, and child abuse.¹

There is no effective treatment for this disorder, and simple care of dermal lesions is sufficient. However, a report indicated healing within 24 hours from the beginning of disease with prednisone therapy.⁷

Spontaneous recovery is usually occurs within 1 to 3 weeks without any scars. Relapse of the disease is also reported.¹³

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