CASE REPORT

Acute Hemorrhagic Edema of Infancy: A Case Report

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Abstract

Acute hemorrhagic edema of infancy (AHEI) is an uncommon form of eukocytoclastic 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 Schonlein 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 Karbala 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 purpric 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 purpric 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 purpuric 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].

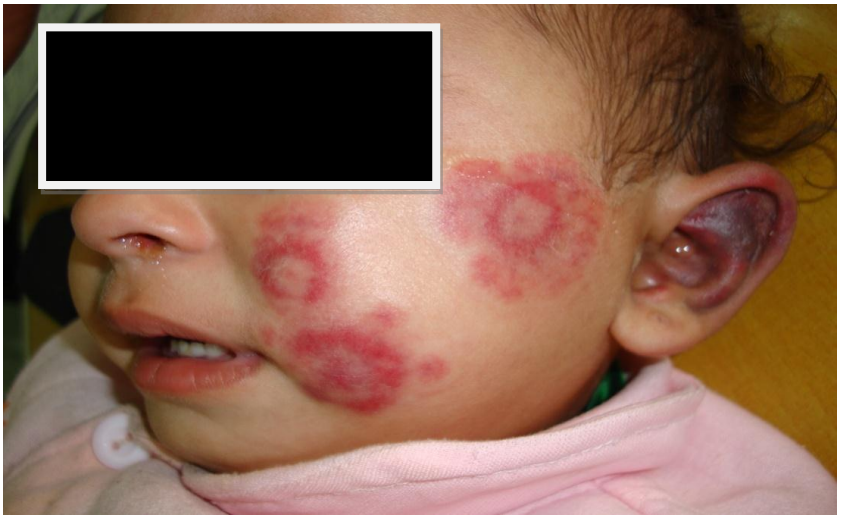

Figure-1 Target like purpuric lesion involving the cheeks and ears.

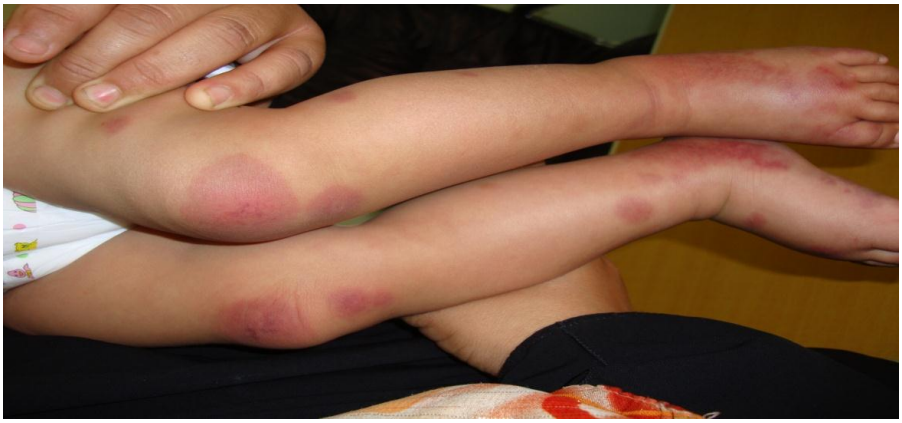

Figure-2 purpuric lesion involving the knee and ankle joints with edema of foots

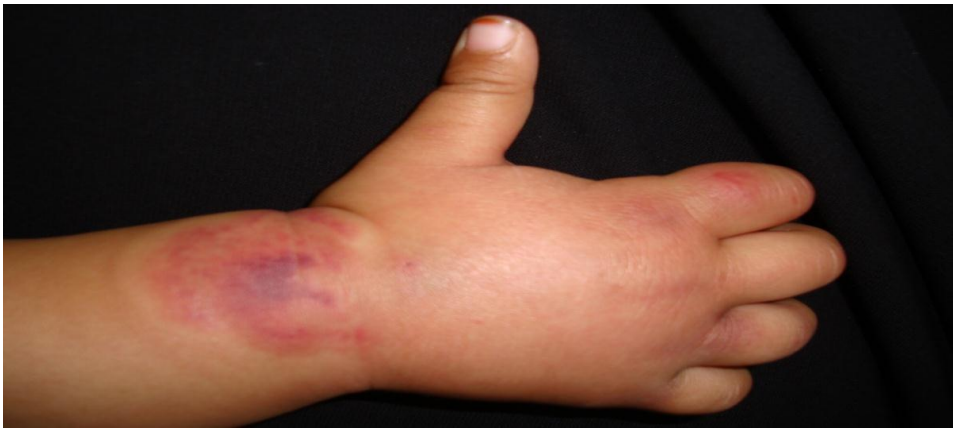

Figure -3 Purpuric target lesions involving the wrist joints with edema of the hands

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

1. Snow IM. Purpura, urticaria and angioneurotic edema of the hands and feet in a nursing baby. JAMA.1913; 61:18-19.
2. Finkelstein H. Lehrbuch der Sauglingskrankheiten. 4th ed. Amsterdam: 1938:814-30.
3. Poyrazoglu H M, Per H, Gunduz Z, et al. Acute hemorrhagic edema of infancy. Pediatr Int 2003; 45: 697-700.
4. Dubin BA, Bronson DM, Eug AM. Acute hemorrhagic edema of childhood: an unusual variant of leukocytoclastic vasculitis. J Am AcadDermatol 1990; 23: 347-350.
5. Ballinger S. Henoch-Schönlein purpura. Curr Opin Rheumatol 2003; 15: 591-594.
6. Goraya JS, Kaur S. Acute infantile hemorrhagic edema and Henoch-Schönlein purpura: is IgA the missing link? J Am Acad Dermatol 2002; 47: 801-802.
7. Garty BZ, Ofer I, Finkelstein Y. Acute hemorrhagic edema of infancy. Isr Med Assoc J 2002 Mar; 4(3):228-9.
8. Poyrazoglu HM, Per H, Gunduz Z,et al. Acute hemorrhagic edema of infancy. Pediatr Int 2003 Dec; 45(6):697-700.
9. da Silva Manzoni AP, Viecili JB, de Andrade CB, Kruse RL, Bakos L, Cestari TF. Acute hemorrhagic edema of infancy: a case report. Int J Dermatol 2004 Jan; 43(1):48-51.
10. Long D, Helm KF. Acute hemorrhagic edema of infancy: Finkelstein's disease. Cutis 1998; 62:65-6.
11. McDougall CM, Ismail SK, Osmerod A. Acute hemorrhagic oedema of infancy. Arch Dis Child 2005 Mar; 90(3):316
12. Gattorno M, Picco P, Gambini C, et al. Erythema multiforme-like manifestations and arthritis in a 3-year-old child with leukocytoclastic vasculitis. Clin Exp Rheumatol 1997 May-Jun; 15(3):329-32.
13. Gattorno M, Picco P, Gambini C, et al. Erythema multiforme-like manifestations and

arthritis in a 3-year-old child with leukocytoclastic vasculitis. Clin Exp Rheumatol 1997 May-Jun; 15(3):329-32.
14. Legrain V, Lejean S, Taieb A, et al. Infantile acute hemorrhagic edema of the skin: studies of ten cases. J Am Acad Dermatol 1991; 24:17–22.
15. Gonggrypp LA, Todd G. Acute hemorrhagic edema of childhood (AHE). Pediatr Dermatol 1998; 15:91–96.
16. Dubin BA, Bronson DM, Eng AM. Acute hemorrhagic edema of childhood: an unusual variant of leukocytoclastic vasculitis. J Am Acad Dermatol 1990; 23: 347-350.
17. Saraçlar Y, Tinaztepe K, Adalıoğlu G, Tuncer A. Acute hemorrhagic oedema of infancy (AHEI) - a variant of Henoch-Schönlein purpura or a distinct clinical entity?. J Allergy Clin Immunol 1990; 86: 473-483.
18. Tomaç N, Saraçlar Y, Türktaş I, Kalaycı Ö. Acute hemorrhagic oedema of infancy: a case report. Clin Exp Dermatol 1996; 21: 217-219.