Case Report

Acute Hemorrhagic Edema of Infancy

Susheera Chatproedprai MD*,
Siriwan Wananukul MD*

** Department of Pediatrics, Faculty of Medicine, Chulalongkorn University*

The authors report a case of acute hemorrhagic edema of infancy in an 8-month-old boy with a history of recent vaccination prior to the illness. He was well in general, apart from mild fever, diarrhea, and the rash compatible with acute hemorrhagic edema of infancy; though, he had unusual trunk and mucosal involvement. The diagnosis was confirmed by the histopathological findings and direct immunofluorescence of the lesion. Complete resolution spontaneously occurred within 3 weeks.

Keywords: *Hemorrhagic, Edema, Papule*

J Med Assoc Thai 2007; 90 (6): 1205-7
Full text. e-Journal: *http://www.medassocthai.org/journal*

Acute hemorrhagic edema of infancy (AHEI) is an uncommon form of leukocytoclastic vasculitis that was first described in 1913. The cutaneous findings in AHEI are dramatic in both appearance and rapidity of onset. It affects children between 4 and 24 months of age. The head and distal part of the extremities are favored sites and visceral involvement is rare[1]. The authors present a case of AHEI in an 8-month-old boy and review the characteristic features of this dermatosis.

Case Report

An 8-month-old boy presented with asymptomatic erythematous rash at the abdominal wall 3 days prior to admission. On the following day, the lesion gradually progressed to the extremities including palms, soles, and forehead. A day before admission, he developed low-grade fever, watery loose stool, and central vesicles ensued in the preceding rash. His parents denied using any medication except for H. influenza type B vaccine that was given a day before the onset of the skin lesion.

Physical examination revealed an active, mildly febrile (38°C) male infant with a few small ulcers at the soft palate. Multiple erythematous papules with central vesicles were present all over the body. Edema of the extremities as well as redness of the perianal area was included in attendance. Others were unremarkable.

Laboratory findings revealed a hemoglobin of 13.1 g%, a white blood cell count of 11,260/ l with a differential count of 33% neutrophils, 51% lymphocytes, 9% monocytes, 7% eosinophils, and a platelet count of 228,000/µl. The urine was negative for hemoglobin. Microscopic examination showed up to 1 red blood cell per high power field. Stool exam revealed up to 1 white blood cell and red blood cell per high power field. Stool culture was negative. Stool for Rotavirus antigen by ELISA was negative. Test for level of complement was normal.

A skin biopsy of the lesion showed dense superficial perivascular and interstitial infiltration. The infiltrate was composed of lymphocytes, a few neutrophils, nuclear dust, eosinophils, and extra-vasated red blood cells. Direct immunofluorescence study was negative.

The patient was treated with lactose free formula for his diarrhea and lidocaine viscus for his oral lesions. No specific treatment was given. His lesion gradually improved within 1 week and resolved in the third week of illness. He has had no recurrences after a 6-month period.

Discussion

Acute hemorrhagic edema of infancy affects infants between 4 to 24 months of age[2]. It is thought

Correspondence to : Chatproedprai S, Department of Pediatrics, Faculty of Medicine, Chulalongkorn University, Rama IV Rd, Pathumwan, Bangkok 10330, Thailand. Phone: 0-2256-4951, Fax: 0-2256-4911, E-mail: susheerac@hotmail.com

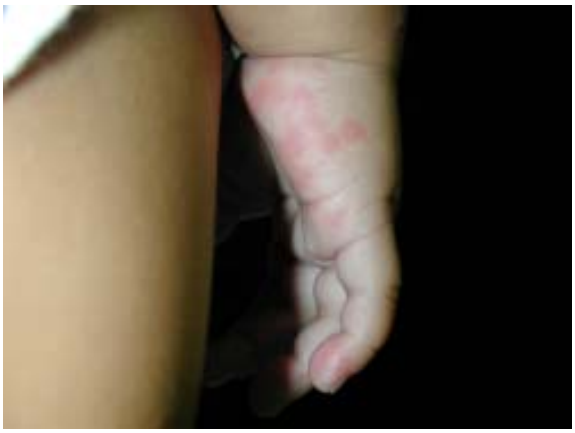

Fig. 1 Multiple erythematous papules in the edematous palms

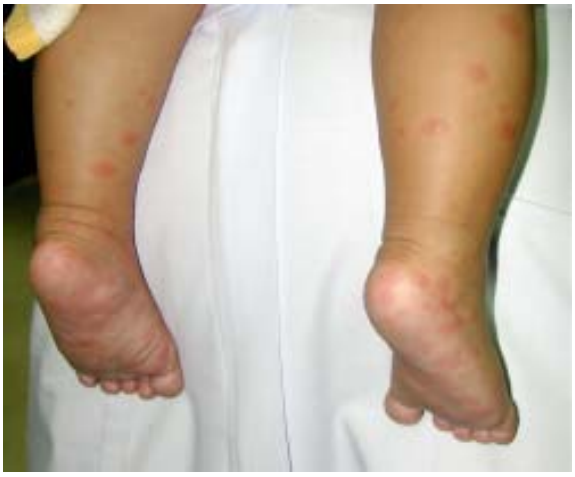

Fig. 2 Multiple erythematous papules with central vesicles in some papules both legs

to represent an immune-complex mediated disease, most probably precipitated by a preceding vaccination, respiratory infection, and medication. A history of recent infection and/or drug intake or immunization is found in 75% of cases[1,2]. Streptococci, staphylococci, and adenoviruses have been implicated as infective agents[3]. Tuberculosis correlated with two cases of AHEI[3]. Regarding immunization, there was a combined vaccine and measles vaccine reported to be associated with AHEI cases[1,2]. The presented patient received H. influenza type B vaccination a day before the onset of the rash.

Acute hemorrhagic edema of infancy presents with fever, peripheral edema, and large, purpuric, annular or targetoid plaques on the face and extremities[2,4-7]. Bullous and necrotic lesions infrequently exist. It has been reported that the trunk and mucosal involvement are usually spared[8]. Unlike Henoch-Schonlein purpura (HSP), visceral involvement is rare and it is one of the main characteristics of AHEI[2]. However, joint pain, gastrointestinal symptoms and renal involvement have been reported in AHEI. The most striking feature of AHEI is the contrast between the acuteness of the cutaneous signs and the general well appearance of the child, apart from mild fever[3].

The primary histopathological finding in AHEI is a leukocytoclastic vasculitis of the dermal vascular plexus that may rarely extend to the subcutaneous fat[9]. Since these findings are common to other forms of leukocytoclastic vasculitis, clinic pathological correlation is necessary for the diagnosis of AHEI[7]. Direct immunofluorescence studies in most cases of AHEI have been reported to give negative results[2,9]. Fibrinogen, C3, IgM, IgA, IgG are, nonetheless, able to be presented in vascular walls and surrounding blood vessels[7].

The clinical differential diagnosis of AHEI includes Henoch-Schonlein purpura, all other forms of cutaneous vasculitis, urticaria, acute febrile neutrophilic dermatosis (Sweet syndrome), erythema multiforme, and child abuse. Results of history, physical examination, and appropriate laboratory studies including examination of the skin biopsy specimen can differentiate these disorders from AHEI[1].

The present patient's presentation followed the typical history of rapid onset and preceding vaccination in AHEI. However, he encountered the trunk and mucosal involvement, which were usually spared[8]. Because the patient's oral lesions were not the ecchymotic ones, either atypical mucosal involvement in AHEI or other differential diagnosis such as erythema multiforme, concurrent infection should be considered. Biopsy of the lesion was performed to differentiate these diseases. He was finally diagnosed as "acute hemorrhagic edema of infancy" due to the history of rapidity of onset, the recent vaccination, a day prior to the illness, the clinical findings without visceral involvement, and the histopathological findings accompanied with negative direct immunofluorescence.

Treatment in AHEI is usually supportive. It is recommended that antibiotics should be given when there is concurrent infection[2]. Systemic coricosteroids and antihistamine have not been proved to hasten the resolution of cutaneous lesions[2,4].

Despite the dramatic clinical findings, the

course of disease is benign in general. Complete and spontaneous recovery occurs within 1-3 weeks. However, there is one report of a case of AHEI with gastrointestinal tract involvement that had a fatal outcome[10]. Recurrences during the weeks to months after presentation have been reported but are uncommon[9]. In one child, a recurrence was triggered by a group A -hemolytic streptococcal pharyngitis[5].

Conclusion

Acute hemorrhagic edema of infancy is an uncommon leukocytoclastic vasculitis affecting the child aged less than 24 months. Despite the dramatic and characteristic cutaneous findings, it had a benign course with complete resolution within 1-3 weeks.

References

1. Poyrazoglu HM, Per H, Gunduz Z, Dusunsel R, Arslan D, NarIn N, et al. Acute hemorrhagic edema of infancy. Pediatr Int 2003; 45: 697-700.
2. Legrain V, Lejean S, Taieb A, Guillard JM, Battin J, Maleville J. Infantile acute hemorrhagic edema of the skin: study of ten cases. J Am Acad Dermatol 1991; 24: 17-22.
3. Gonggryp LA, Todd G. Acute hemorrhagic edema of childhood (AHE). Pediatr Dermatol 1998; 15: 91-6.
4. Crowe MA, Jonas PP. Acute hemorrhagic edema of infancy. Cutis 1998; 62: 65-6.
5. Dubin BA, Bronson DM, Eng AM. Acute hemorrhagic edema of childhood: an unusual variant of leukocytoclastic vasculitis. J Am Acad Dermatol 1990; 23: 347-50.
6. Ince E, Mumcu Y, Suskan E, Yalcinkaya F, Tumer N, Cin S. Infantile acute hemorrhagic edema: a variant of leukocytoclastic vasculitis. Pediatr Dermatol 1995; 12: 224-7.
7. Long D, Helm KF. Acute hemorrhagic edema of infancy: Finkelstein's disease. Cutis 1998; 61: 283-4.
8. Smitt JH, Vermeer MH, Faber WR. Acute hemorrhagic edema of infancy (AHEI). Clin Dermatol 2002; 20: 2-3.
9. Cunningham BB, Eramo L, Caro W. Acute hemorrhagic edema of childhood present at birth. Pediatr Dermatol 1999; 16: 68.
10. Larregue M, Lorette G, Prigent F, Canuel C. Acute haemorrhagic edema of the skin of the new-born with lethal intestinal complication (author's transl). Ann Dermatol Venereol 1980; 107: 901-5.

โรค acute hemorrhagic edema of infancy

สุชีรา ฉัตรเพริดพราย, ศิริวรรณ วนานุกูล

รายงานผู้ป่วยเด็กอายุ 8 เดือนมารักษาด้วยอาการไข้ ถ่ายเหลว ผื่นแดงตามหน้าท้องและฝ่ามือ ฝ่าเท้า ผู้ป่วยมีประวัติได้รับวัคซีนป้องกันเยื่อหุ้มสมองอักเสบจากเชื้อฮิบ 1 วันก่อนมีอาการแต่ไม่มีประวัติได้รับยาอื่น ๆ ตรวจร่างกายพบว่าผู้ป่วยมีไข้ต่ำร่วมกับแผลในปาก ผิวหนังพบผื่นแดงกระจายทั้งตัวร่วมกับตุ่มน้ำตรงกลางในบางจุด การตรวจทางห้องปฏิบัติการไม่พบความผิดปกติ การตรวจพยาธิสภาพผิวหนังพบความผิดปกติที่เข้าได้กับโรค acute hemorrhagic edema of infancy ผู้ป่วยไม่ได้รับการรักษาจำเพาะใด ๆ สำหรับผื่นซึ่งจางลงภายใน 1 สัปดาห์ และหายไปในสัปดาห์ที่ 3
