Acute Hemorrhagic Edema of Infancy

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Key words: purpura, edema, hemorrhage

IMAJ 2002;4:228–229

Henoch-Schönlein purpua is the most common leukocytoclastic vasculitis in children. Occasionally other forms of leukocytoclastic vasculitis may occur, as described here.

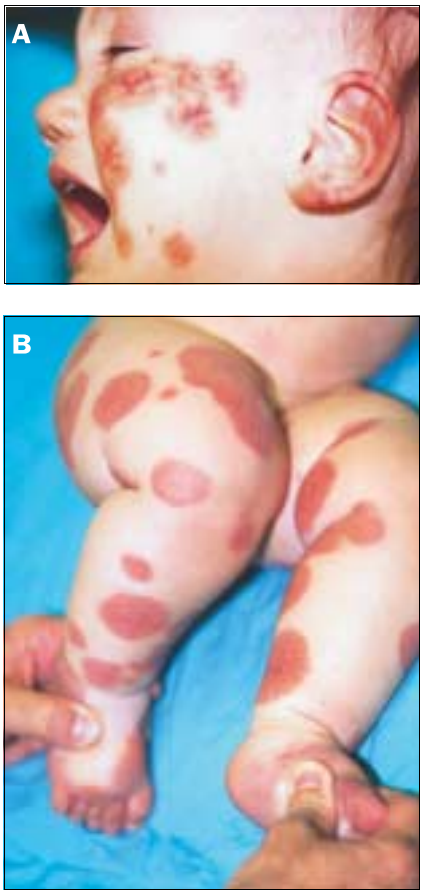
Patient Description

An 11 month old boy was admitted because of fever, edema and rash. His history and that of his family were unremarkable. He had diarrhea for 3 days and a rash appeared on his thighs on the day before admission. Examination revealed an ecchymotic rash over the face, auricles, arms, hands, gluteal area, legs and feet. The lesions, which were tender, initially resembled insect bites but gradually became larger, concentric and purpuric [Figure]. The child also had a fever of 39.5°C and non-pitting edema of the face and legs. Other vital signs and the rest of the physical examination were unremarkable.

Blood laboratory tests showed a white blood cell count of 18,000/mm^3, hemoglobin 12 g/dl, platelet count 532,000/mm^3 and erythrocyte sedimentation rate 35 mm/hour. Urinalysis showed 4–5 red blood cells/high power field. Blood chemistry (SMA-18), coagulation studies and sepsis workup, including lumbar puncture, were all within normal limits. Serum immunoglobulin and complement levels were normal. Blood urine and stool cultures were negative.

Skin biopsy showed leukocytoclastic vasculitis with nuclear dust mixed with leukocytic infiltrate in the vessel walls. Direct immunofluorescence revealed a mild perivascular deposit of C3 but no deposits of immunoglobulins. A diagnosis of acute hemorrhagic edema of infancy was made.

During hospitalization, bloody diarrhea was noted. Intravenous ceftriaxone (100 mg/kg/day) was administered for 3 days until the fever gradually normalized, and the blood disappeared from the stools. Five days after admission the skin lesions began to clear and the edema subsided. New purpuric lesions recurred during the following 3 weeks, but afterwards the skin cleared and there were no further recurrences for 12 months of follow-up.



[A] Edema and ecchymotic lesions of the legs.
[B] Ecchymotic lesions of the cheeks and auricles.

Comment

AHEI or Finkelstein's disease [1] is an acute leukocytoclastic vasculitis occurring in infants and young children between the ages of 3 months and 2 years. Since its first description in 1913, approximately 100 cases of AHEI have been reported [2,3]. This may reflect a low incidence, underdiagnosis, or confusion with other disorders. AHEI is considered a distinct clinical entity in the European (non-English) medical literature [4], whereas most American authors identify it as a variant of Henoch-Schönlein purpura [5]. AHEI is probably not rare since at least 12 children have been admitted to our medical center during the last 5 years with the diagnosis of AHEI.

The onset of AHEI is acute and may be associated with fever, tender symmetric edematous areas (on the face and extremities) and subsequent rapid development of ecchymotic purpura, especially in the areas of pre-existing edema. It has been suggested that the distribution of the purpura is gravity-dependent [2]. Visceral involvement is rare and may affect the kidneys, causing hematuria or mild proteinuria; or the intestine, causing bloody diarrhea [4] as observed in our patient. Complete recovery usually occurs within 2–3 weeks. Recurrent episodes have rarely been reported [3].

Most patients have a mildly elevated erythrocyte sedimentation rate and a few patients have elevated circulatory immune complexes. Histologic study demonstrates leukocytoclastic vasculitis. In contrast to Henoch-Schönlein purpura, perivascular deposits of immunoglobulin A are detected in only 10–35% of AHEI patients [4].

The differential diagnosis of AHEI includes Henoch-Schönlein purpura, meningococcemia, erythema multiforme and

AHEI = acute hemorrhagic edema of infancy

drug eruption. The main differences between AHEI and Henoch-Schönlein purpura are that AHEI patients are very young (usually not older than 2 years), fever is common, the distribution of the rash includes the face and auricles, edema is almost always present, perivascular deposits of immunoglobulin A are infrequent, and renal involvement and long-standing complica-tions are rare.

The pathogenesis of AHEI is unknown. The increased prevalence of the disease in winter has been attributed to its possible association with bacterial or viral infection [3]. Other factors thought to be associated with AHEI are vaccinations and drug therapy, mainly antibiotics. However, since most vaccinations are given to children during the first year of life, and upper respiratory tract and other viral infections are very common in this age group, these associations may be coincidental.

Recognizing AHEI is important because despite the dramatic initial clinical appearance that may cause parents and medical personnel significant anxiety, the outcome is favorable.

References

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