VAN DER HEGGEN T¹, DHONT E¹, SCHELSTRAETE P², COLPAERT J³

[1] Department of Pediatric Intensive Care, University Hospital Ghent, Belgium [2] Department of Pediatric Pneumology and Infectious Diseases, University Hospital Ghent, Belgium [3] Department of Pediatrics, AZ Groeninge, Kortrijk, Belgium

ACUTE HEMORRHAGIC EDEMA OF INFANCY: A DRAMATIC PRESENTATION WITH A BENIGN COURSE

Introduction

Acute hemorrhagic edema of infancy (AHEI) is an uncommon type of leukocytoclastic vasculitis affecting young children under two years of age. It is a benign condition that usually resolves spontaneously in a few weeks, without any sequelae or a need for active therapy. Nevertheless, as the onset is often dramatic with an impressive purpuric rash on the extremities and face, its presentation can easily be confused with more serious conditions that require immediate treatment.

Case report

An 11-month-old male infant was admitted to a pediatric department with a viral bronchitis, and treated supportively with antipyretics and intravenous fluid therapy. On the second day of admission, he rapidly developed multiple annular hemorrhagic lesions on his hands, feet and both ears. Simultaneously , all extremities showed a progressive, nonpitting dorsal edema. Broad-spectrum antibiotics were initiated because of suspicion of meningococcal sepsis. However, the patient remained nontoxic and hemodynamically stable; he was alert but irritable.

Complete blood count showed a leukocytosis, mild thrombocytosis and a Creactive protein of 54 mg/L. Liver and kidney function tests, urine analysis and blood culture were normal.



Acute hemorrhagic edema of infancy



The patient was transferred to the pediatric ICU because of ongoing gradual increase in swelling and pain in both hands, with a prolonged capillary refill time, indicating compromised limb perfusion. An urgent fasciotomy of both hands was performed. Nasopharyngeal aspirate was positive for adeno-, boca- and parainfluenza virus. Diagnosis of AHEI was confirmed by a skin biopsy, which showed a leukocytoclastic vasculitis. Besides intravenous pain medication, no further therapy was initiated.

After two days of ongoing progression of the skin lesions in both size and number, and development of a bilateral conjunctivitis, a gradual improvement of the hemorrhagic rash and the patient's general condition was seen with complete recovery.

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Criteria for diagnosis "classic triad"	Low grade fever + a rapidly progressing purpuric rash on ears and lims + nonpitting acral edema Non-toxic child Skin biopsy is only nessecary in case of uncertain diagnosis
Cause	Believed to be immune-complex mediated: 75% of cases are preceded by infections, drug exposure or active immunization (here: adenovirus infection)
Evolution	Self-limiting in 10 – 20 days
Need for therapy	None (steroids in severe cases?) Need for a fasciotomy, as in our case, is rare and has only been reported once in literature

Conclusion

AHEI is an uncommon type of leukocytoclastic vasculitis, with an often dramatic with the good general condition of the patient, is the key feature to differentiate it from other worrying diagnoses, like meningococcemia.

Contact

tatjana.vanderheggen@ugent.be Department of paediatrics Ghent University Hospital, Belgium

f Universiteit Gent

in @ugent

¥ **Ghent University**



