ACUTE HEMORRHAGIC EDEMA OF INFANCY – A CASE SERIES OF 10 PATIENTS

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Background/Aims: Acute hemorrhagic edema of infancy (AHEI) is a relative unknown leukocytoclastic vasculitis. It consists of symmetric edema of the face and extremities with acute onset, followed by rapid development of ecchymotic purpura, with or without fever. Our objective was to determine the clinical features and outcome of AHEI and to discuss the complexity of the diagnosis.
Methods: Retrospective review of clinical files of patients diagnosed with AHEI in the last 9 years.
Results: We report 10 cases of AHEI (6 females; 4 males; median age at diagnosis of 11 months). Most cases occurred in the winter months. All had purpuric lesions in the inferior limbs and all except one had purpura elsewhere. Peripheral edema was only present in 7 patients and fever in 4; 7 had a context of previous infection, mostly upper respiratory tract, and 4 of those were still on active treatment with antibiotics/ antihistaminics. 3 patients complained of joint pain and one presented with blood in the stools, none had renal involvement. 8 patients had a diagnostic work-up to exclude other diagnosis such as sepsis but none had a positive urine or blood culture. Histological exam was performed in only 1 patient and was consistent with the clinical diagnosis. All had a full recovery in few weeks without specific treatment and only one had recurrent episodes.
Conclusion: Although typical, the clinical presentation of AHEI can be variable and sometimes be associated with life-threatening infections, raising issues of differential diagnosis. While the cutaneous lesions are certainly impressive, a wait-and-see approach is, in most cases, the only treatment required as complete resolution is expected in the following 2 to 3 weeks. AHEI is an under-diagnosed entity that is still unknown to many pediatricians. The obligation for skin biopsies and full septic screen should be reserved to the atypical cases.