Case Report

Acute hemorrhagic edema of the infant (Finkelstein's disease): case report

Abstract

Acute hemorrhagic edema of the infant (AHIE) is a benign small vessel vasculitis that occurs in children under 2 years of age associated with infections, drug use and/or vaccinations. The indicated treatment is symptomatic and expectant, unlike other vasculitides with systemic involvement. Case report: 10-month-old female infant presented to the emergency department with generalized purpuric lesions of spontaneous onset and rapid progression.

Keywords: acute hemorrhagic edema of the infant, vasculitis

Volume 11 Issue 1 - 2023

Ángel Siero, Estuardo Morales, Raúl Estuardo Marchena, Otto Peláez, Ely Fletcher
Department of Pediatrics, Hospital Infantil de Infectología y Rehabilitación, Guatemala City, Guatemala

Correspondence: Ángel Siero, Department of Pediatrics, Hospital Infantil de Infectología y Rehabilitación, Guatemala City, Guatemala, Email angeljsiero@gmail.com

Received: March 09, 2023| **Published:** March 31, 2023

Introduction

EAHL is a disease that has been reported only 100 times in the literature, but its incidence is believed to be higher because it can be confused with other vasculitides, such as Henoch-Schölein purpura (HSP), which is why it is important to make an early diagnosis in order to provide the appropriate treatment and improve the prognosis. The following is the case of a patient with EAHL.

Case report

Female patient, 10 months old, previously healthy, with a history of skin lesions of 3 days of evolution, associated with mild edema in the lower limbs. Mother reports sudden and spontaneous onset on both hands; with spread to forearms, arms and face; 24 hours prior to consultation, lesions reach the thorax and lower limbs. Patient with a history of self-limited upper respiratory infection two weeks ago. In emergency, febrile patient, with purpuric papules with well-defined borders that converge in plaques; present on the face, auricular pavilions, neck, anterior and posterior thorax, upper and lower limbs (Figure 1); without mucosal involvement. Rest of physical examination without important findings. During her hospital stay she presented febrile symptoms associated with convulsive crisis.

Labs were performed with a white blood cell count of 7,020/ul, hemoglobin 10.9 g/dl, platelet count at 445,000/ul; coagulation times, renal function tests and urinalysis within normal parameters for age. C-ANCA, P-ANCA, C3, C4 and immunoglobulin levels were normal, including IgA at 108.97 mg/dl.

Dermatology performed a biopsy of the chest lesion with a 2 mm punch and indicated to start treatment with antihistamine and intravenous corticosteroid. The pathology report evidences "inflammatory infiltrate of mononuclear predominance, mainly lymphocytes alternating with polymorphonuclear neutrophils and erythrocyte extravasation in small blood vessels; findings compatible with leukocytoclastic vasculitis". The diagnosis of hemorrhagic edema of the infant is considered based on clinical and pathologic findings. Patient receives intravenous treatment with evident improvement, disappearing lesions on the 7th day, with persistence of ecchymotic shadow in affected areas. The patient was referred to an outpatient clinic for follow-up (Figure 2).

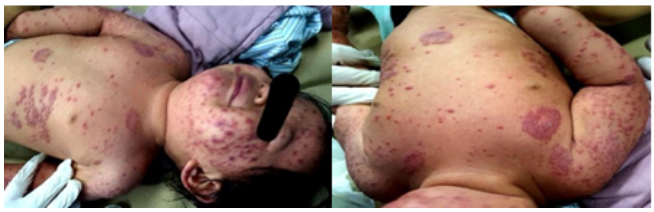

Figure 1 Distribution of purpuric lesions on the face and thorax of the patient on admission.

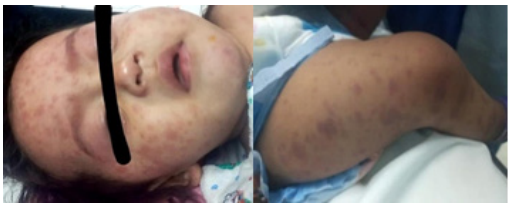

Figure 2 Resolving lesions on face and right lower limb at discharge.

Discussion

EAHL is a benign small vessel leukocytoclastic vasculitis that occurs in children between the ages of 3 months and 2 years. This is an immune complex-mediated vasculitis mainly associated with infections of bacterial and viral origin, but also described with drug use (mainly antibiotics and NSAIDs) and in some cases associated with vaccination. Other names as this entity is also known are: Seildmayer syndrome or Finkelstein's disease.[1]

The diagnosis is mainly clinical, the history of the disease is characterized by skin lesions of rapid onset and progression, usually within 24 to 48 hours; the lesions are characterized by onset as macules, papules or urticariform plaques that progress to palpable purpuric lesions, which may then evolve from red or purplish to yellowish-brown as the lesions resolve. The distribution is symmetrical, affecting mainly the face (especially the auricles, cheeks and eyelids) and extremities, although they may appear anywhere on the skin. Rarely it affects mucous membranes. It can also be associated with edema, which is non-pitting, soft, involving auricular pavilions, face and extremities (especially the dorsum of hands and feet). In male patients there may be scrotal edema. It may present with low-grade fever.[2]

Kraus et al proposed the following diagnostic criteria for EAHL: age less than two years; purpuric or ecchymotic lesions with edema of the face, joints or extremities with or without mucosal involvement; absence of systemic or visceral involvement; spontaneous recovery within days to weeks. The patient in the case meets all the criteria.[3] It is important to consider and rule out differential diagnoses, especially other types of vasculitis with systemic involvement. One of the entities with which EAHL is often confused is HSP. In the latter entity, the age of presentation is older (3 and 8 years); the distribution of the lesions is different (flexor surface of legs and buttocks); it is common to find systemic complications, such as arthralgias, gastrointestinal bleeding, abdominal pain and renal involvement; the resolution time is 4 weeks, although it can be prolonged if there are systemic complications.[4]

The histopathological findings in both vasculitides are similar; the difference is that in PHS the perivascular deposits are usually IgA, whereas in EAHL such deposits are found in 10-35% of cases. Some authors consider EAHL and PHS to be two variants of the same entity, while other authors consider that there are sufficient clinical and prognostic differences to consider them different entities.[3,5]

Other differential diagnoses that should be considered are: hemorrhagic urticaria, erythema multiforme, immune thrombocytopenia, meningococcemia, Kawasaki disease, drug reactions, sepsis, other vasculitides. These diagnoses are important since some of these pathologies require emergency treatment, while EAHL is usually self-limiting and resolves in one to three weeks. Regarding treatment, there is debate as to whether antihistamines and topical and/or systemic steroids have any benefit; some authors suggest benefit in their use, while others prefer conservative treatment as it is debated whether these medications modify the course of the disease. Regardless of whether medical or expectorative treatment is given, EAHL resolves completely without sequelae most of the time, although it has been reported that it may leave hyperpigmented lesions or mild skin atrophy at the site of the purpuric lesions.[3]

Acknowledgments

None.

Conflicts of interest

The authors declare no conflict of interest.

References

1. Siqueira Cunha DF, Fornazieri Darcie AL, Benevides GN, et al. Acute Hemorrhagic Edema of Infancy: an unusual diagnosis for the general pediatrician. *Autops Case Rep*. 2015;5(3):37–41.
2. Yarmuch P, Chaparro X, Fischer C, et al. Acute hemorrhagic edema of the infant: report of an atypical case presentation. *Rev Chil Pediatr.* 2012;83(5):462–467.
3. Serra C, Garcia M, Sokolova A, et al. Acute Hemorrhagic Edema of Infancy. *Eur Ann Allergy Clin Immunol*. 2015;48(1):22–26.
4. Riola A, López D, Serrano N. Acute hemorrhagic edema of the infant. *Rev Pediatr Aten Primaria*. 2019;21:397–399.
5. Roy K, Madke B, Kar S. Acute Hemorrhagic Edema of Infancy. *Indian J Dermatol.* 2015;60(6):624–625.