Acute hemorrhagic edema of infancy (Finkelstein's disease): favorable outcome with systemic steroids in a female patient *

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Dear Editor,

Acute hemorrhagic edema of infancy was initially described in 1913 by Snow. It is a benign, self-limited leukocytoclastic vasculitis that occurs most commonly in males. It evolves favorably with conservative medical management and treatment with steroids is not usually administered.

We present a case of a 2-year-old girl, non-toxemic, with sudden onset of asymptomatic large purpuric non-blanching plaques, with a targetoid and annular appearance, predominantly on the limbs and trunk, sparing the genital area, and edema and tenderness of the left ankle 2 days before the initial medical evaluation. A history of diarrhea from the previous week that resolved with oral antibiotics (cephalosporin) for 5 days was the only relevant medical history. The patient's immunizations were up to date; she had no other clinical conditions. Her temperature on arrival was 36.5°C, and she presented no other signs or symptoms. She was prescribed loratadine by a general practitioner (Figure 1).

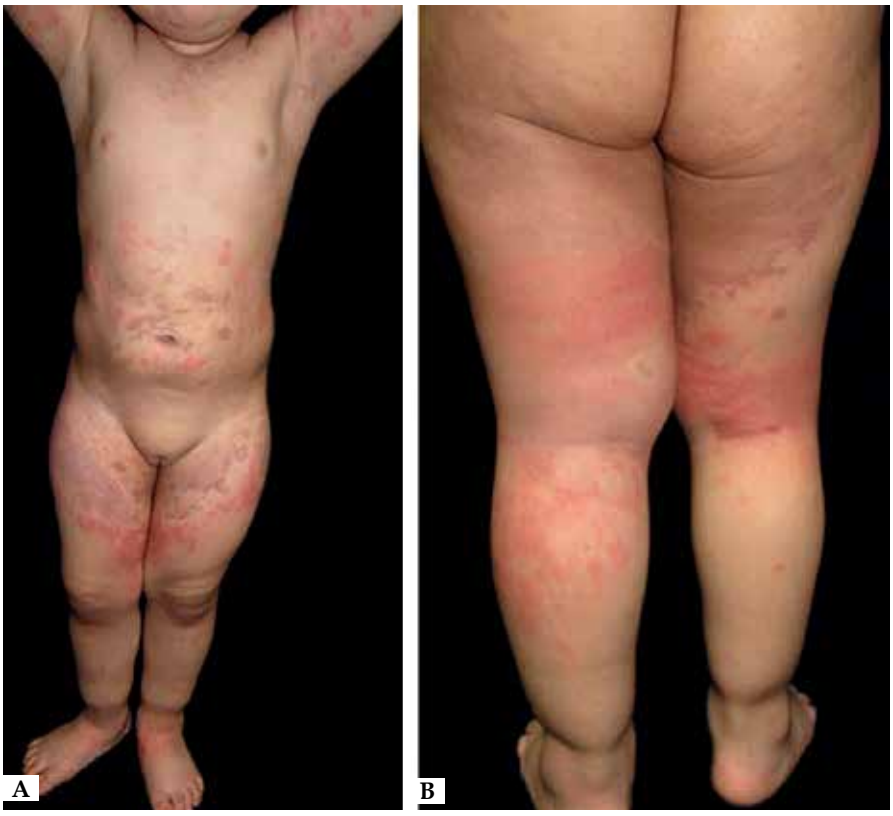

Figure 1: A and B: Erythematous-purpuric annular and targetoid lesions on the limbs and trunk

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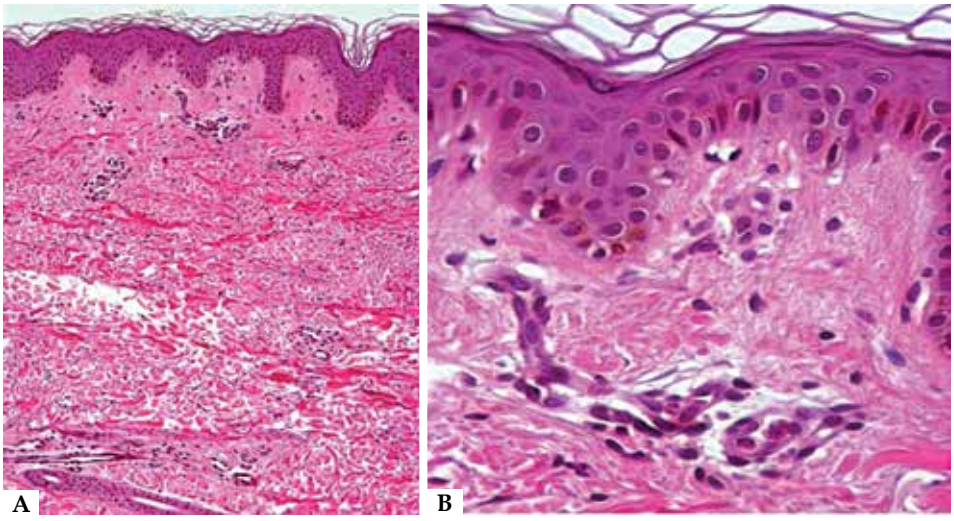

Figure 2: A and B: Dermal vessel leukocytoclastic vasculitis (H&E, 10 x) **(A)** Close-up showing leukocytoclastic vasculitis (H&E) **(B)**

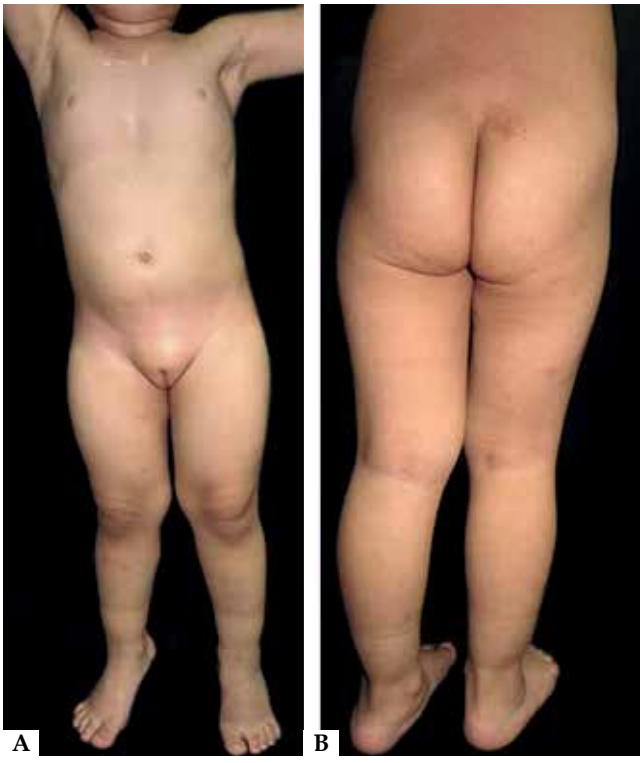

Figure 3: A and B: Patient with complete resolution of lesions 4 days after steroid treatment

Parents were reassured of the benign nature of this condition. The laboratory investigation showed hemoglobin: 12.9 g/dL, hematocrit 38.2%, leucocytes 11.48 x 10^9 cells per L, platelets 401 x 10^9 cells per L, C reactive protein 1:320, ESR: 18 mm/hr (0-20) and a normal urinalysis. A biopsy of the forearm was performed, which showed evidence of leukocytoclastic vasculitis in the dermal vessels with nuclear dust (Figure 2). The patient was managed as an outpatient with a tapered dose of betamethasone 1 mg/kg. At follow-up three days later, the patient had complete clearing of the lesions (Figure 3).

Acute hemorrhagic edema of infancy occurs mainly in male patients aged between 4 and 24 months. It has a worrisome, sudden presentation, but it is self-limited and lasts from 1 to 3 weeks.[1] Additional rare systemic symptoms are abdominal pain, gastrointestinal bleeding, arthritis or nephritis.[2] It is considered a variant of Henoch-Schönlein purpura but differs from this condition with the low prevalence of renal involvement, which is uncommon and mild if present.[1]

The differential diagnosis includes Henoch-Schönlein purpura, meningococcemia, Kawasaki disease, erythema multiforme, urticarial vasculitis, malignancies and child abuse.[1] There are some criteria proposed to correctly identify this disease:

I. Age less than 2 years;
II. Purpuric or ecchymotic target-like lesions, with edema of the face, auricles, and extremities, with or without mucosal involvement;
III. Lack of systemic disease or visceral involvement;
IV. Spontaneous recovery within a few days or weeks.[3]

Immunofluorescence usually does not identify IgA deposits on the patients' skin.[1]

This disease is usually inconsequential. There are only 2 reports in which secondary complications were presented. In one case, visual impairment occurred due to massive periorbital edema, which resolved after 1 day of treatment with prednisolone. In another case, foot edema occurred, which led to a compartment syndrome that responded to fasciotomy. [4,5] In this patient, there was a rapid clearing of lesions with no secondary complications.

Complete spontaneous recovery usually takes from 6 to 21 days.[1] It is of paramount importance to reassure the patient´s family of the benign nature of the lesions because its presentation is usually alarming. Systemic steroids may help accelerate the resolution of the disease, calm parents, alleviate the patient's symptoms, and spare the patient from complications.❑

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