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Erythematous-purpuric lesions in a one-year-old child revealing a case of acute hemorrhagic edema of childhood

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To the Editor:

Acute hemorrhagic edema of infancy (AHEI) is a rare skin disease that almost exclusively affects children younger than two years old. It presents as a purpuric/ecchymotic rash and swelling of the limbs, ear, and face [1]. Histologically it exhibits an immune complex-mediated leukocytoclastic vasculitis [2].

A one-year-old boy, previously healthy, presented with erythematous plaques on the face and pinnae that became erythematous-violaceous and progressed to involve the upper and lower limbs, associated with symmetrical edema of the feet and necrosis of the right pinna. There were reports of flu-like symptoms with fever peaks ranging from 38ºC to 39ºC during the 8 days before the onset of skin lesions. On physical examination, purpuric, coalescing, symmetric plaques were observed, some in the form of iris and medallions, measuring 0.5cm to 4 cm on the cheeks and pinnae and the distal region of the upper and lower limbs (**Figure 1**). On the right helix, an area of necrosis measuring 0.4cm was visible (**Figure 1C**). The condition was accompanied by symmetrical swelling of the feet.

Hospital admission for investigation was indicated and intravenous corticosteroid therapy (methylprednisolone 2mg/kg/day) was initiated in view of the necrosis of the pinna. Complementary investigation showed: blood count with leukocytosis, normal liver and kidney function, and elevated inflammatory tests (erythrocyte sedimentation rate, C-reactive protein, and D-dimer).

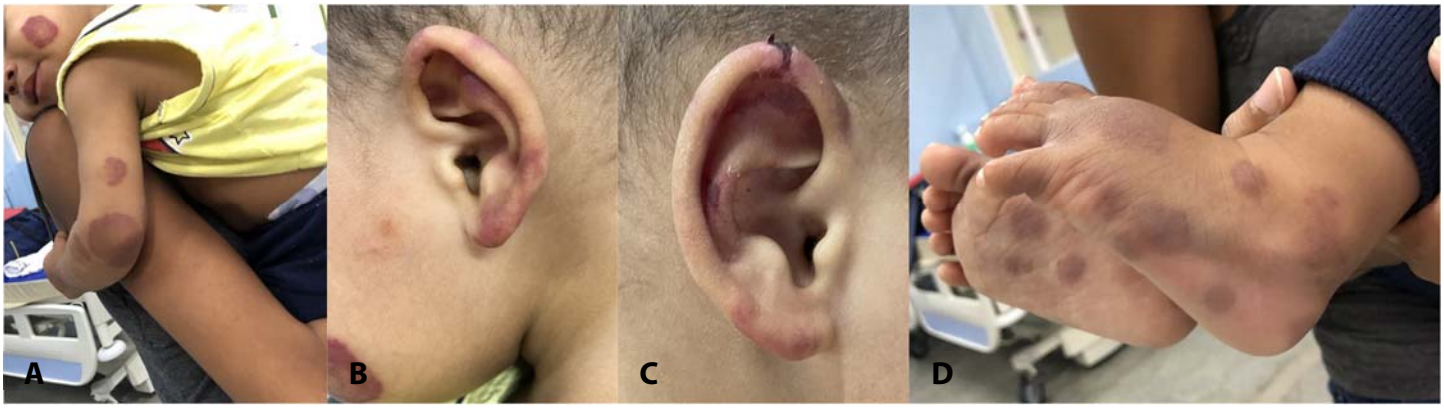

Figure 1. ***A)*** *Purpuric macules and plaques on the face, left pinna and distal region of the left upper limb. Iris patch on left cheek (consistent with the diagnosis).* ***B)*** *Purpuric macules on the left pinna.* ***C)*** *Purpuric macules on the right pinna associated with an area of necrosis on the ipsilateral helix (consistent with the diagnosis).* ***D)*** *Coalescing purpuric plaques on the lower limbs.*

Urinalysis, electrolytes, and coagulogram were within normal limits. Serologies were negative for Epstein-Barr, rubella, herpes one and 2, HIV, HTLV, toxoplasmosis, and COVID-19. IgG antibodies against cytomegalovirus was positive. Abdominal ultrasonography and transthoracic echocardiogram showed no alterations. Owing to the absence of systemic involvement and the benign course of the disease, corticosteroid taper was started on the fifth day of hospitalization to one mg/kg/day for three days when he was discharged from the hospital. During outpatient treatment, the corticosteroid was gradually reduced with a final dosage of 0.16mg/kg/day in one month and the corticosteroid was subsequently suspended. The child's clinical condition resolved without sequelae during hospitalization. No relapse was observed within 6 months of follow-up.

Acute hemorrhagic edema of infancy is characterized by the initial manifestation of palpable hemorrhagic skin lesions and petechiae that can progress to medallion-like plaques measuring one to 6cm in diameter, associated with local edema. The extremities are usually the most affected areas, as well as the ears, eyelids, and malar region. The trunk is usually spared [1,3]. Lesions on the ears have a greater chance of becoming necrotic [1]. The etiology is uncertain, some authors consider the disease to be a cutaneous form of Henoch-Schonlein purpura whereas others believe it to be an entity distinct within the spectrum of leukocytoclastic vasculitides [1]. Upper respiratory tract infections, including COVID-19, vaccines, and antibiotics are recognized as triggers of AHEI [4].

The treatment of AHEI is considered controversial. Some authors advocate the use of corticosteroids and antihistamines to accelerate the resolution of the disease. One study has reported an improvement in a case of AHEI accompanied by necrosis after the administration of intravenous corticosteroid therapy [2]. On the other hand, other studies have pointed out that the improvement is expected in the natural course of the disease, with sufficient supportive treatment; spontaneous resolution without sequelae is expected to occur in most cases [4,5].

With the advent of upper respiratory tract infections, including COVID-19, we will possibly face an increase in the number of cases of AHEI. Therefore, it is of fundamental importance to think about this entity in children with cutaneous manifestations.

Potential conflicts of interest

The authors declare no conflicts of interest.

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