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CASE REPORT

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Acute hemorrhagic edema of infancy (AHEI): Alarming cutaneous presentation of a benign and self-limited disease

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Abstract

Acute hemorrhagic edema of infancy (AHEI) is a cutaneous leukocytoclastic vasculitis. We report on an 11-month-old boy, presenting the classical feature of AHEI with alarming cutaneous presentation, but good clinical condition. Early recognition is crucial to avoid unnecessary medical investigations or therapies, as well as to identify potentially severe complications.

KEYWORDS

acute haemorrhagic edema of infancy, case report, Finkelstein disease, leukocytoclastic vasculitis, Seidlmayer disease

1 | INTRODUCTION

Acute hemorrhagic edema of infancy (AHEI), also known as Finkelstein or Seidlmayer disease, is a rare cutaneous leukocytoclastic vasculitis. It was first described by Snow, in 1913.[1] It is characterized by a triad of manifestations: fever, edema, and large purpuric skin lesions. It mainly occurs in infants and young children aged 6–24 months. Despite its alarming presentation, it has a benign course with spontaneous resolution within few weeks.[2] It was previously considered as a variant of Henoch–Schönlein purpura (HSP). Currently, it is recognized as a distinct clinical picture, also in light of lack of visceral involvement and less frequent occurrence of IgA skin deposition.[3] Here, we report on an 11-month-old male infant, coming to our observation in good general clinical conditions, and with oval purpuric lesions on the face and extremities.

2 | PATIENT PRESENTATION

An 11-month-old male infant presented with rapidly appearing oval purpuric lesions, on the face and extremities. In the previous week, the patient had fever (maximum ear temperature 38.5°C) for 5 days, associated with upper respiratory infection symptoms. At admission, his general conditions were good, with normal vital signs. History of trauma, recent vaccination, and/or previous use of drugs were ruled out. On physical examination, a rash characterized by purpuric confluent elements with cockade pattern was observed on the right cheek, ears (Figure 1), upper and lower limbs, sparing trunk and back. The diameter of hemorrhagic lesions varied from few millimeters to several centimeters. Edema of the hands and feet, joint swelling (Figure 2), and pain completed his clinical profile. The articular manifestations were responsive to analgesic therapy with paracetamol and resolved spontaneously in a week. Blood counts, coagulation, hepatic, and renal function tests, C3 and C4 complement fractions and immunoglobulins, as well as urine analysis were normal. C-reactive protein and erythrocyte sedimentation rate were increased, with values of 1.5 mg/dl (normal <0.5 mg/dl) and 36 mm/h (normal <15 mm/h), respectively. Ultrasound examination of the abdomen and the urinary tract excluded visceral involvement. During the hospitalization the infant maintained good clinical condition, with stable vital parameters. Skin lesions showed a worsening clinical outcome, with increased diameter and color intensity during the following 2 days from admission. Based on the history and clinical and laboratory findings, an AHEI diagnosis was made. Then, cutaneous manifestations and edema gradually decreased, and the patient was discharged after 5 days. The infant was included in a clinical follow-up. The purpuric lesions completely disappeared after 2 weeks, without treatment. After 6 months, he is in good clinical conditions without any complications of disease.

3 | DISCUSSION

AHEI, also known as Finkelstein or Seidlmayer disease, is a rare cutaneous leukocytoclastic small vessel vasculitis, which mainly affects male infants and young children aged 6–24 months.[4,5] The etiology is unclear, although viral and bacterial infections (cytomegalovirus, herpes simplex virus, varicella zoster virus, adenovirus, rotavirus, streptococci, staphylococci, and tuberculosis) of respiratory, urinary, and gastrointestinal tracts may be observed in some affected patients.[6–10] Vaccinations and medications (penicillin, cephalosporin, trimethoprim–sulfamethoxazole, paracetamol, thiazides, and nonsteroidal anti-inflammatory drugs) may also be involved in its pathogenesis.[11] In our case, the infant had a 5-day history of upper respiratory tract infection, suggesting an immune complex-mediated pathogenesis. A classical feature of AHEI is the contrast between alarming cutaneous presentation and good general conditions of patients. The diagnosis is based on the history and typical clinical findings, which are fever, purpura, and edema.[12] Low-grade fever is observed in 50% of patients. The skin lesions have rapid onset, develop over 24–48 h, and typically include large nummular red or purpuric plaques or cockade-like lesions. Cutaneous manifestations usually involve extremities, cheeks, and ears, sparing the trunk. Lesions of the mucosal membranes are rarely observed.[13] Edema is typically asymmetrical, tender, nonpitting, and painful, as shown in our case. It may involve ears, face, extremities, and mainly the back of hands and feet.[14] The involvement of other organs and systems, in addition to skin, is rarely described, with few reported cases of abdominal pain, arthralgia, renal involvement, and scrotal swelling.[15]

FIGURE 1 Purpuric confluent elements with cockade pattern on face (mainly on right cheek) and ears.

The proposed criteria for disease identification are the following: (1) age less than 2 years; (2) purpura or ecchymosis with typical skin lesions, edema of face, ears, and extremities with or without mucosal involvement; (3) lack of systemic disease or visceral involvement; and (4) spontaneous recovery within few days or weeks.[16] Our infant fulfilled all these criteria.

Routine blood tests are typically normal in affected subjects. High levels of erythrocyte sedimentation rate and C-reactive protein, as in our infant, as well as leukocytosis, thrombocytosis, eosinophilia, and mild increase of hepatic function tests, may be observed in AHEI. In some cases, patients may have hypocomplementemia affecting C4, C1q, and CH50 complement.[15] Although urine analysis is normal in most children, in some instances it may reveal microscopic hematuria or proteinuria, markers of

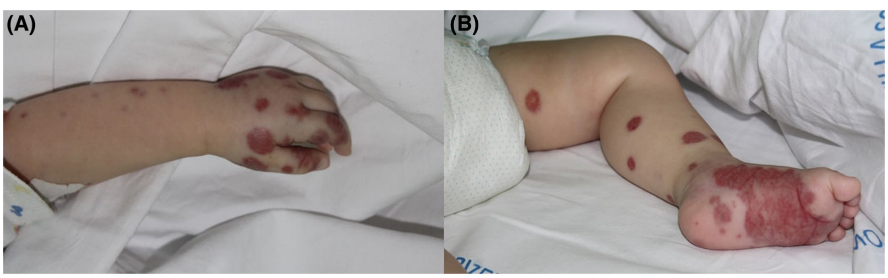

FIGURE 2 (A/B). Purpuric skin lesions on limbs. Note edema of hand (A) and foot (B).

renal involvement.[17] A skin biopsy may be performed in selected cases, in which a clinical diagnosis was not possible to be performed. Histopathological examination shows a leukocytoclastic vasculitis of dermal vessels, characterized by perivascular neutrophilic infiltration, with several nuclear fragments in the vascular wall, resulting in fibrinoid necrosis.[4] Direct immunofluorescence test, performed in approximately one third of cases with biopsy studies, reveals vascular deposits of immunoglobulin A in less than a quarter of patients.[18] AHEI should be differentiated by HSP, Sweet's syndrome, acute febrile neutrophilic dermatosis, erythema multiforme, hemorrhagic urticaria, Kawasaki disease, trauma-induced purpura, drug-induced vasculitis, and child abuse.[19]

Despite the rapid progression of lesions, AHEI is a self-limited disease with spontaneous and complete resolution, often occurring within 1–3 weeks.[20] Affected patients rarely have recurrent attacks and complications, like scarring lesions as well as renal, gastrointestinal, or joint involvement. AHEI has no specific treatment, and conservative management is the most common approach.[5] Systemic corticosteroids and antihistamines do not improve the course of disease. Antibiotics may be used in case of bacterial infection.[18] In the present, patient no treatment was started, and the cutaneous lesions completely disappeared after 2 weeks, without leading to *sequelae*.

4 | CONCLUSIONS

The present report highlights the classical feature of AHEI, with the contrast between its alarming cutaneous presentation and the good clinical conditions of affected patients. AHEI is an uncommon benign and self-limited disease, which pediatricians should consider in infants and young children (mainly aged 6–24 months) with purpuric lesions and tender non-pitting edema. Early recognition is crucial to avoid unnecessary medical investigations or therapies, as well as to identify the rare, although potentially severe, complications.

AUTHOR CONTRIBUTIONS

MT: conceptualized the study and revised the manuscript. LLS: collected and analyzed clinical data and drafted the manuscript. DT: contributed in drafting the manuscript. FM: contributed to clinical management of the patient. AG: contributed to clinical management of the patient and in drafting the manuscript. MG: collected and analyzed clinical data. GP: contributed to clinical management of the patient. BLG: contributed to clinical management of the patient and in drafting the manuscript. GC: conceptualized the study, revised the manuscript, and gave final approval of the version to be submitted. All authors approved the final manuscript as submitted.

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CONFLICT OF INTEREST

The authors declare that they have no competing interests.

DATA AVAILABILITY STATEMENT

Data sharing not applicable to this article as no datasets were generated or analysed during the current study

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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