

Imaging clinical case

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A 12-month-old, previously healthy boy presented at the Emergency Department with a seven-day history of nasal obstruction and cough, low-grade fever, and sudden appearance of a purpuric rash over the left lower limb, which progressed to the face, right lower limb, and buttocks. No recent history of vaccination or drug intake was reported and no hematuria was reported by the parents.

Clinical examination revealed a good general status, stable vital signs, palpable annular purpuric lesions with well-defined edges distributed over the face (Figure 1), limbs (Figure 2), ears, buttocks, and scrotum, with associated right tibiotarsal joint edema (Figure 3). Laboratory tests revealed slightly elevated platelet count (474.000/ μ L), C-reactive protein 0,79 mg/dL, and IgE 139 UI/mL. Hemoglobin level, leukocyte count, blood urea

nitrogen, serum creatinine, serum electrolytes, transaminase levels, IgA/G/M, prothrombin time, and activated partial thromboplastin were all within the normal age reference values. Urinalysis was unremarkable.

Given joint involvement, empiric treatment with oral betamethasone 0.1 mg/kg/daily was initiated on outpatient setting for two days.

Forty-eight hours after the onset of purpura, child's rash and edema began to dissipate. Three weeks later, complete recover was observed.

What is your diagnosis?



Figure 1 – Purple, annular plaques over the face.



Figure 2 – Purple, annular plaques over the lower limbs.



Figure 3 – Right tibiotarsal joint edema.

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DIAGNOSIS

Acute hemorrhagic edema of infancy (AHEI)

DISCUSSION

Acute hemorrhagic edema of infancy is a rare and benign small vessel leukocytoclastic vasculitis, first described by Snow in 1913, characterized by the sudden onset of purpuric lesions on the face, ears, and extremities in a healthy child. It is associated with edema of the extremities and, sometimes, low-grade fever.^{1,2} Visceral involvement is rare.¹

Despite its unknown etiology, 75% of cases are preceded by a viral or bacterial infection, immunization, or drug exposure.¹

It typically affects children aged 4 to 24 months, with a slight male predominance, and is more frequent during winter time.¹

The main differential diagnoses include *Henoch-Schönlein* purpura, meningococemia, erythema multiform, Kawasaki disease, drug reaction, urticaria, Sweet's syndrome, neonatal Lupus, and battered-child syndrome.^{2,3}

The clinical triad (typical annular purpuric rash on the face, ears and extremities, edema of the extremities, and mild fever), no specific laboratory findings, and a rapid improvement, usually allow for a correct diagnosis.¹ Skin biopsy is generally not required.⁴

The disease is self-limiting and a spontaneous and complete resolution usually occurs within one to three weeks.^{2,3} Treatment with oral corticosteroids can be considered for patients with severe presentation or complications (arthralgia/arthritis, gastrointestinal bleeding, renal involvement).¹

ABSTRACT

Introduction: Acute hemorrhagic edema of infancy (AHEI) is a rare and benign leukocytoclastic vasculitis, frequently misdiagnosed.

Case report: A twelve-month-old boy presented with a seven-day history of coryza and cough associated with low-grade fever and the sudden appearance of a purpuric rash over the left lower limb. It progressed to an annular purpuric rash, with well-defined edges, on the face, limbs, ears, buttocks, and scrotum, with right tibiotarsal joint edema. Mild thrombocytosis was detected on laboratorial investigation. Patient was treated with a two-day course of oral betamethasone, with clinical improvement. No complications were reported within three weeks of follow-up.

Discussion: AHEI has a sudden onset and a benign, self-limiting course. This report may be a helpful reminder to discriminate AHEI from other, more serious diseases, avoiding exhaustive and unnecessary investigation.

Keywords: Hemorrhagic edema; Purpuric rash

RESUMO

Introdução: O Edema Hemorrágico Agudo da Infância (EHA) é uma vasculite leucocitoclástica rara e benigna, por vezes subdiagnosticada por não ser correctamente reconhecida.

Caso clínico: É descrito o caso de um lactente de 12 meses, do sexo masculino, com obstrução nasal e tosse com

sete dias de evolução associado a aparecimento súbito de exantema purpúrico no membro inferior esquerdo e febre baixa. O exantema progrediu com placas purpúricas palpáveis, anulares, com bordos bem definidos na face, extremidades, pavilhões auriculares, nádegas e escroto e edema na articulação tibiotársica direita. A investigação laboratorial evidenciou trombocitose ligeira. O doente foi medicado com betamatesona durante dois dias, com melhoria clínica. A resolução completa do quadro clínico foi observada após três semanas, sem sequelas.

Discussão: O EHA tem início súbito e um curso benigno e autolimitado. Pretende-se com este caso alertar para o reconhecimento precoce desta entidade, a fim de evitar uma investigação exaustiva e desnecessária.

Palavras-chave: Edema hemorrágico; Rash purpúrico

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REFERENCES

1. Parker L, Nissan K, Hoffung L, Harel L, Amir J, Bilavsky E, *et al.* Acute hemorrhagic edema of infancy: the experience of a large tertiary pediatric center in Israel. *World Journal of Pediatrics.* 2017,13:341-5.
2. Ceci M, Conrieri M, Raffaldi I, Pagliardini V, Urbino AF. Acute hemorrhagic edema of infancy - Still a challenge for the pediatrician. *Pediatr Emerg Care.* 2018; 34:e28-9. doi: 10.1097/PEC.0000000000000749.
3. Glamann J, Morrison A, Mychaliska K. On target: a case of acute hemorrhagic edema of infancy. *Hospital Pediatrics.* 2014, 4:106-8.
4. Chesser H, Chambliss J, Zwemer E. Acute hemorrhagic edema of infancy after coronavirus infection with recurrent rash. *Case Reports in Pediatrics.* 2017. Article ID 5637503.

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