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Acute Hemorrhagic Edema of Infancy in Saudi Infant-Case Report

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Abstract | **Case Report**

Acute hemorrhagic edema of infancy (AHEI) is a rare acute benign cutaneous leukocytoclastic vasculitis affecting children below 2 years of age. We present a case report of 1 year old Saudi infant presented with impressive purpuric rashes and edema of the hands and feet. He had benign courses with complete resolution of clinical findings within 72 hours. Early recognition of AHEI is important to avoid unnecessary procedures and tests, and help in counseling the patient's family.

Keywords: Hemorrhagic edema, Henoch-Schönlein purpura.

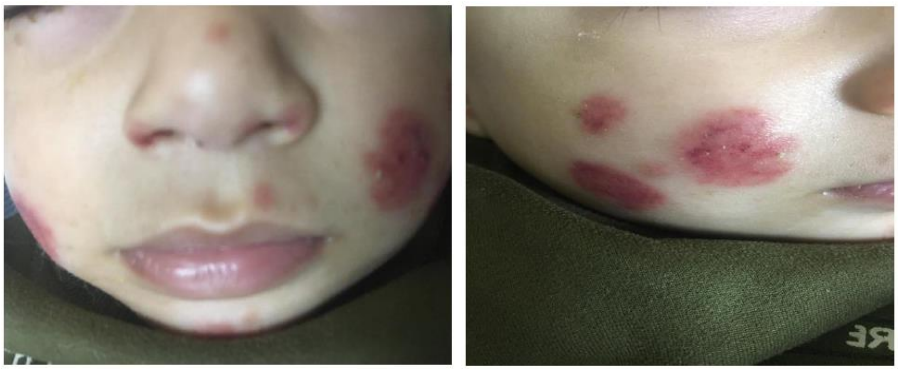
INTRODUCTION

Acute hemorrhagic edema of infancy (AHEI) is a rare acute benign cutaneous leukocytoclastic vasculitis affecting children below 2 years of age. Its presentation can be mimic urticaria, erythema multiforme, Henoch-Schönlein purpura, idiopathic thrombocytopenia, meningococcemia, Kawasaki disease, and drug rash [1].

CASE REPORT

Previously healthy one-year saudi old boy presented to emergency with a 2 days history of fever and Skin Rash associated with decrease activity and poor Feeding. Fever at home not documented, intermittent, responded to Antipyretic. Rash first appear as papules on both ears, then spread along the face, nose, cheeks, hands and feet in less than 24 hours. It was not itchy. Associated with redness swelling on the wrist and the ankle, not involve Abdomen and back or mucus membrane or genitalia .unremarkable past history.

On examination, the patient was febrile with normal blood pressure for age and a maximum heart rate of 156 beats per minutes. His skin was well demarcated, round typical targetoid redness lesion, non-blanching, or pruritic, symmetrical distribution in face, hand and leg, Center pale, Red rim, 1.5 cm diameter, blister developed in cheek, break to form ulcer. Additionally, there was non-pitting edema of wrists and ankles. Further physical exam was otherwise normal (figure 1a, 1b -2-3).



(a) **(b)**

Fig-1: Macular Erythema with few Crusted Patches

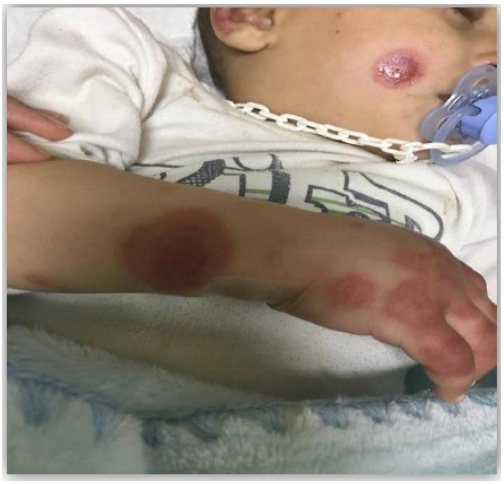
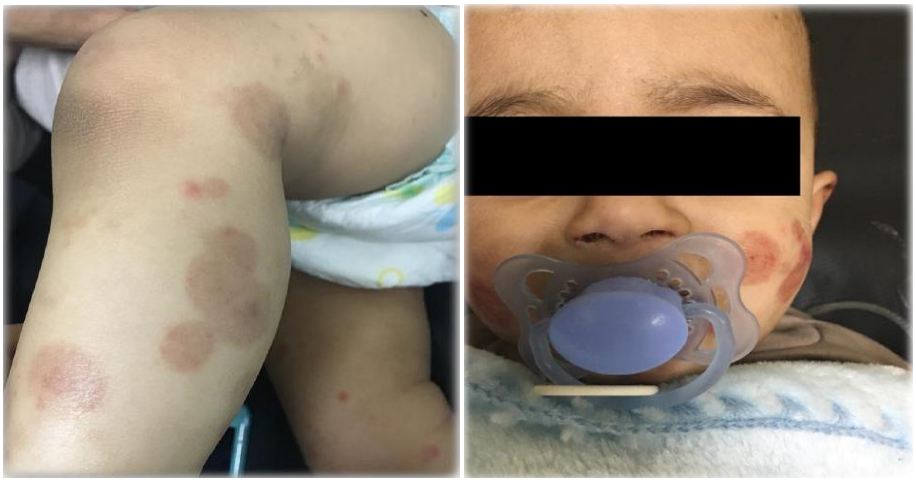


Fig-2: Well demarcated, round typical targetoid lesions Center pale, Red rim, 1.5 cm diameter

Fig-3: Erythematous rash with swelling of wrist

Laboratory investigations showed white cell count 13.78x10^9/liter, lymphocyte 48.28%, hemoglobin 8.7 g/dl, platelet 522x10^9/liter, reticulocyte count 11.18%, C-reactive protein 25.6 mg/dl, erythrocyte sedimentation rate 55 mm/hr. blood culture was negative. Patient had complete resolution in his condition within 72 hours (figure 4a, 4b).



(a) **(b)**

Fig-4: Three days after treatment: fading and shrink of rash

DISCUSSION

The full etiology of AHEI is still understood, however it is hypothesized to be a type of leukocytoclastic vasculitis induced by an immune complex hypersensitivity related to recent infection or drug [2].

The skin lesions are erythematous, annular, rosette, or targetoid patches that cluster and often coalesce. The face and extremities are predominately affected; however it can affect any cutaneous tissue. These self-limiting lesions can appear very suddenly, and many resolve into ecchymotic purpuric patches. Edema of the hands, feet, face, or extremities and absence of any visceral involvement is main featues as well [3]. Krause *et al.* [3] proposed the following criteria for diagnosing AHEI:

- Age b 2 years;
- Purpuric or ecchymotic target-like skin lesion with edema on the head and face, with or without mucosal involvement;
- Lack of systemic disease or visceral involvement; and
- Spontaneous recovery within few days or weeks.

There are currently no specific laboratory test for AHEI; however, a basic knowledge of the characteristics of AHEI and the other disorders in the differential of a purpuric rash help pediatrician to exclude serioud diagnoses and avoid unnecessary procedures and tests [4].

The possible differential diagnoses include urticaria, erythema multiforme, urticarial multiforme Henoch-Schönlein purpura (HSP), idiopathic thrombocytopenia, meningococcemia, Kawasaki disease, and drug rash [1].

Treatment for AHEI is mainly supportive with family reassurance of the self-limited benign course of the disease. Steroids are not changing the course of illness. Patients may receive some symptomatic relief from the swelling, and there may be some improvement in the appearance of rash [5-7].

CONCLUSION

Acute hemorrhagic edema of infancy (AHEI) is a rare acute benign Self-limited illness. High index of suspicion, exact diagnosis is critical to rule out more serious illness like sepsis and Kawasaki and to reassure the concerned family.

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