Case Report

Unraveling acute hemorrhagic edema of infancy in the COVID-19 era: Insights from a tribal area in Jharkhand

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Abstract

Acute hemorrhagic edema of infancy (AHEI) is a benign and rare presentation of leukocytoclastic vasculitis that usually affects children between 4 months and 24 months of age. It is characterized by purpuric and ecchymotic lesions that mainly involve the distal extremities, the face, and the ears. It usually follows some viral or bacterial infection. Here, we report a case of a 25-month-old male child who presented with mild grade fever and upper respiratory tract infection symptoms. Subsequently, he developed progressive purpuric and ecchymotic lesions over his body, mainly on his lower limbs and face. Laboratory tests were done, showing elevated C-reactive protein and erythrocyte sedimentation rate with rest of normal results. In view of patients having respiratory symptoms, reverse-transcriptase polymerase chain reaction of a nasopharyngeal swab for coronavirus disease 2019 (COVID-19) was done, which came out to be positive. The baby received supportive care only. He gradually improved and was discharged successfully. AHEI may be a possible after effect of COVID-19 infection.

Keywords: AHEI, COVID-19, Finkelstein disease, Seidlmayer disease

Introduction

Primary care providers are the first line of contact for the detection, prevention, and treatment of many diseases. Therefore, it is important for primary care providers and family medicine practitioners to diagnose acute hemorrhagic edema of infancy (AHEI) at the earliest and provide adequate treatment to patients. This will also ease the undue anxiety of parents.

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Acute hemorrhagic edema of infancy is a rare form of benign leukocytoclastic vasculitis affecting children between 4 months and 24 months of age.[1] It was first described by Snow in 1913. It is characterized by purpuric and ecchymotic rashes with edema of the face and extremities. AHEI is most commonly misdiagnosed as HSP (Henoch Scholein purpura).[2] However, these two entities must be distinguished because AHEI is a rare benign disease, whereas HSP typically has renal and gastrointestinal co-morbidities.[3] History and physical examination differentiate between two entities; however, histopathology and immunofluorescence add further diagnostic confidence.

Case Report

A 25-month-old boy, who had previously been in his usual state

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of health, was brought to the out-patient department (OPD) with the complaint of multiple purpuric and ecchymosis rashes over his lower limbs, genital area, and face, as shown in Figures 1 and 2. The patient had edema of the lower extremity and genital area along with rashes, as shown in Figures 3 and 4. Six days before the development of rashes, the patient had a fever with cough and rhinorrhea. The patient was given symptomatic treatment by a local doctor before being referred to us for purpuric rashes all over the body and lower extremity edema. Immunizations were all up-to-date.

Pertinent laboratory findings include raised C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) with a normal complete blood count (CBC), RFT (renal function test), LFT (liver function test), PT/INR, and urine analysis. A coronavirus disease 2019 (COVID-19) reverse-transcriptase polymerase chain reaction (RT-PCR) was conducted in light of respiratory symptoms and was positive. Three weeks later, a serology test for COVID-19 was done, which was positive. After consulting with the dermatological team, the patient was clinically diagnosed with AHEI, and a skin biopsy was sent. Skin biopsy findings were consistent with leukocytoclastic vasculitis, showing perivascular neutrophilic infiltration with numerous nuclear fragments in the vascular wall, culminating in fibroid necrosis. The patient was discharged after diagnosis, and skin lesions disappear in 3 weeks without any sequelae. The patient was followed up for 6 months during that time; urine samples were analyzed monthly and found to be normal.

Discussion

AHEI is a rare form of benign leukocytoclastic vasculitis, also known by other names such as Finkelstein disease, Seidlmayer disease, and post-infectious cockade purpura.[4] It has a usually non-toxic presentation and typically presents with fever, palpable purpura, and edema. The skin lesion is characterized by well-demarcated, medallion-like, annular purpuric rashes that appear on the face and extremities. Sometimes, skin lesions can present as ecchymotic and hemorrhagic blisters as well. It is associated with edema of the face, auricles, genitals, and extremities.[5]

Although the cause of AHEI is uncertain, it is thought to be an immune-mediated vasculitis and is frequently brought

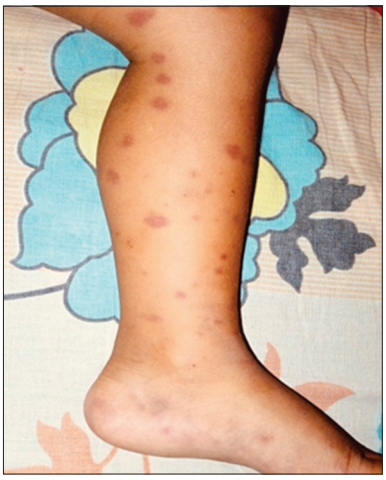

Figure 1: Purpuric and ecchymotic rash over the lower limb

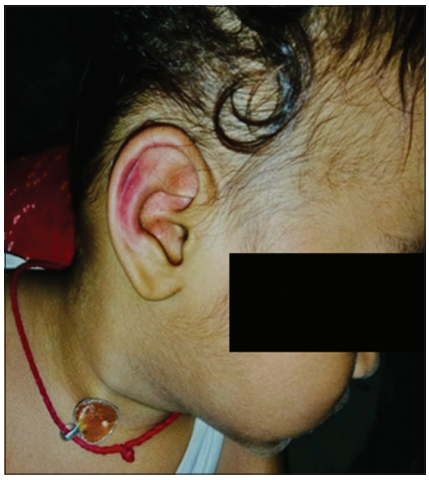

Figure 2: Rash over the ear

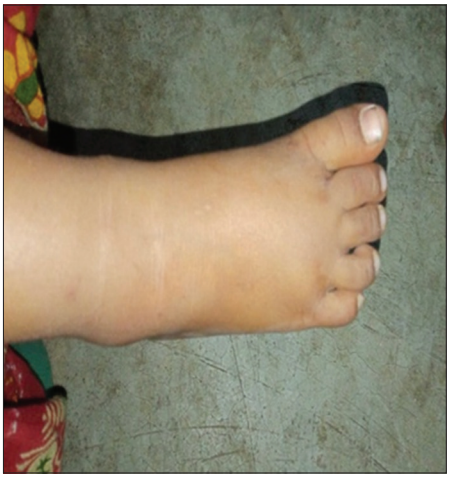

Figure 3: Lower limb edema

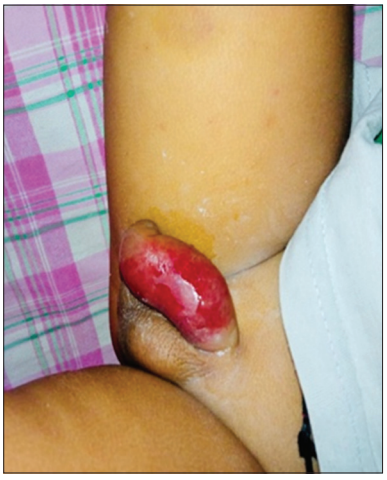

Figure 4: Genital rash and edema

on by predisposing factors such as viral infections, bacterial infections (such as upper respiratory tract infections or urinary tract infections), and a history of drug intake (cephalosporine, paracetamol, thiazides).[6]

Typically, blood laboratory tests are non-specific. There could be thrombocytosis, leucocytosis with eosinophilia, or lymphocytosis in the CBC. Furthermore, CRP and ESRs may be elevated. AHEI may present with hypocomplementemia affecting complement C1q, complement C4, and complement CH50.[7]

If the diagnosis is ambiguous, a skin biopsy for histology and immunofluorescence testing is recommended. The typical histology in an AHEI patient consists of perivascular neutrophilic infiltration with many nuclear fragments in the vascular wall, culminating in fibrinoid necrosis.

Immunofluorescence analysis of skin biopsy reveals that C3 and IgM deposit and less frequently IgA and IgE.[8]

The differential diagnosis includes HSP, Kawasaki, meningococcemia, and erythema multiforme's because of their striking similarity. There is still much disagreement as some authors view the condition as a solely cutaneous version of HSP, while others see it as a distinct entity. Table 1 lists some of the distinctions between the two diseases.

It has a self-limiting course. Skin lesions mostly resolve within 1–3 weeks. Treatment is still debatable; usually, patients are treated conservatively, but some experts support corticosteroid therapy.[9]

Conclusion

AHEI is a self-limiting disease that normally resolves on its own. It is critical to make an accurate diagnosis of AHEI by primary care practitioners to minimize unnecessary work-up and therapy. AHEI may be a possible aftereffect of COVID-19 infection.

Ethical approval

This article does not contain any studies on human participants or animals performed by the authors.

Table 1: Differences between AHEI and HSP

Features	AHEI	HSP
Age of Presentation	6 months to 24 months	3 to 8 years
Skin Lesions	Face, genital area, and extremities	Lower extremities, buttocks, thigh
Renal Involvement	Rare	Frequent
Gastrointestinal Involvement	Rare	Frequent
IgA deposits	Rare	Frequent
Resolution	2–3 weeks	4 weeks

Informed consent

Informed consent of parents of patients concerned was taken in an appropriate format.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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