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RASHES, EDEMA, AND THE DIAGNOSTIC PUZZLE: UNRAVELING ACUTE HEMORRHAGIC EDEMA OF INFANCY IN A 9-MONTH-OLD

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ABSTRACT

Acute Haemorrhagic Edema of Infancy (AHEI) is a rare and benign small vessel vasculitis that typically affects children aged 4 to 24 months. Despite its self-limiting nature, AHEI often poses a diagnostic challenge due to its clinical similarity to more severe conditions such as Henoch-Schönlein purpura (HSP), erythema multiforme, Kawasaki disease, and meningococcaemia. The report on the case of a 9-month-old girl who presented with localized swelling and purpuric skin lesions, initially misdiagnosed as cellulitis. The diagnostic challenge was compounded by the rarity of AHEI and its resemblance to other vasculitis and infectious conditions. Following a series of clinical evaluations and a skin biopsy, the diagnosis of AHEI was confirmed. The patient was treated with a short course of oral corticosteroids, resulting in complete resolution of symptoms. The accurate diagnosis of AHEI not only facilitated appropriate management but also prevented unnecessary investigations and alleviated parental anxiety. This case underscores the importance of considering AHEI in the differential diagnosis of infants presenting with purpuric lesions and localized edema. Early recognition and accurate diagnosis are crucial in preventing over-treatment and ensuring optimal patient care.

1.0 INTRODUCTION

Acute Haemorrhagic Edema of Infancy (AHEI), also known as postinfectious cockade purpura, Finkelstein's disease, or Seidlmayer purpura, is an uncommon small-vessel vasculitis that typically presents with transient haemorrhagic oedema and ecchymosis in young children [1]. AHEI remains a poorly understood condition, and its exact prevalence is not well-established due to its infrequent occurrence and the difficulty in distinguishing it from other similar conditions. A case series and systematic review by Fiore et al. has reported approximately 300 patients with AHEI have been reported since it was first described in 1913 [2]. Given that AHEI is a benign, self-limiting disease, many cases may go unreported or undiagnosed, particularly if clinicians are unfamiliar with this entity. Consequently, the true prevalence and incidence of AHEI are not well documented in large-scale population studies.

Although AHEI is benign, its cutaneous manifestations can be dramatic in both appearance and rapid onset, often mimicking more concerning conditions such as Henoch-Schönlein purpura, erythema multiforme, Kawasaki disease, and meningococcaemia [3]. This can lead to significant anxiety for both parents and clinicians, potentially resulting in overzealous investigations and unnecessary treatments for a condition that is, in fact, self-limiting. It has been reported that rashes in AHEI were mistaken for lesions associated with child abuse, leading to potential false alarms [2]. We present this case to raise awareness of this uncommon yet benign form of leukocytoclastic vasculitis. Recognizing its characteristic presentation is critical in clinical practice, as it can help clinician to avoid unnecessary investigations and treatments.

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2.0 CASE PRESENTATION

A 9-month-old girl, born term and thriving with no previous medical illness presented with left lower limb swelling and purpuric skin rashes for 2 days prior to admission. The lower limb swelling was first noticed at the left knee and spread distally, causing pain and restricted her movement. Shortly afterwards she developed multiple dark purplish discrete purpuric rashes on the left leg (Figure 1). Prior to the appearance of the rashes, mother recalled the child felt feverish to touch but no temperature was documented. The feverish sensation resolved without antipyretics. Further history taking revealed no history of fall or insect bite. She has no history of bleeding tendency such as easy bruising. There is no family history of blood disorders. She remained active with good appetite.

On examinations, her left leg was swollen from knee to dorsum of left foot, it was slight erythematous, warm and tender to touch. There are multiple dark purplish rashes on the medial aspect of left thigh and calf. The rashes appeared circular with erythematous raised rim and central clearing, indurated and tender on palpation. Her full blood count revealed thrombocytosis and leucocytosis (Platelet 733, WBC: 19.1). Inflammatory markers were raised (CRP: 52.85 ESR: 37). Her coagulation profile was normal. She also had normocytic normochromic anaemia. Her left lower limb X-rays showed no facture or local bony lesions. She was admitted and treated as left leg cellulitis. The left lower limb swelling resolved spontaneously after 2 days. On day 3 of admission, she developed similar swelling on the right lower limb. Patellar tap was positive over the right knee. An ultrasound of the right lower limb showed no evidence of right knee effusion. Intravenous antibiotics were continued and the swelling gradually subsided. On Day 6 of admission, child developed right ear pinna swelling and redness but resolved spontaneously. She was discharged home with a course of oral antibiotic for a total of 10 days and was given appointment to review in 2 weeks.

In the next 10 days, the child continued to develop multiple episodes of alternating swelling with purpuric rashes involving different parts of her body. The swelling occurred with sudden onset including both her lower limbs, upper limbs, both ears and even face. The duration of the swelling varied between 1-3 days. These episodes of swelling were accompanied by varying sizes of purpuric skin lesions. She did not have fever and other constitutional symptoms. Although the swelling and skin lesions appeared worrisome to clinician and parents, the patient remained well and comfortable. She was readmitted for diagnostic work up. Darrier sign was negative and there was no lymphadenopathy or organomegaly. Repeated blood work-up revealed worsening thrombocytosis (Platelet: 733-> 828) but her CRP was reducing in trend (52.8-> 16.8). ESR levels were static and her connective tissue markers were not raised (C3: 1.41, C4: 0.35, Rheumatoid factor and ANA were negative). A skin biopsy was done. The finding revealed leukocytoclastic vasculitis with negative immunofluorescence test, consistent with AHEI. She was given a short course of oral prednisolone and achieved full recovery.

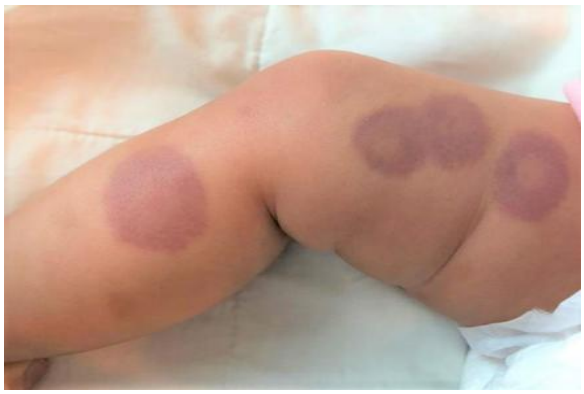

Figure 1. Rashes on the right lower limb shows the typical ‘cockade’ (medallion-like) pattern, with scalloped borders and central clearing

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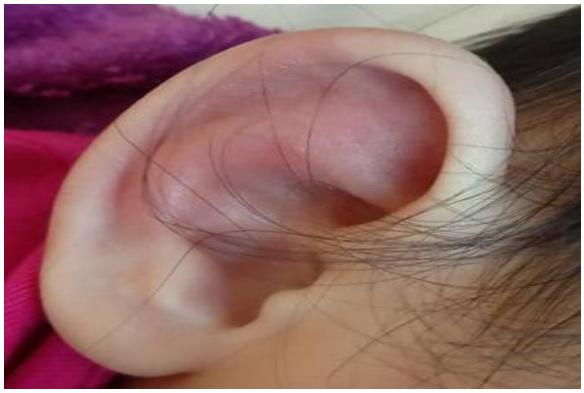

Figure 2. Oedema and purpuric rash on the right ear pinna

3.0 DISCUSSION

Although the number of reported cases of AHEI has been limited over the past century, studies consistently show that most affected children are between 4 and 24 months old. However, more recent literature has started to identify cases across a broader age spectrum, including children as old as 5 years [4] and as young as neonates [5, 6]. This raises the possibility that many cases may have been underreported in the past, possibly due to clinicians' unfamiliarity with this condition.

There are no specific laboratory tests to diagnose AHEI, so its recognition relies primarily on the patient's history and physical examination. Most of the available literature highlights a triad of symptoms—fever, oedema, and purpura—as key to diagnosing the disease [7,8]. However, a study by Krause et al. found that only 45% of patients with AHEI develop fever [9], which challenges the traditional diagnostic framework. Similarly, Serra et al. and Miconi et al. reported that only approximately 50% of children with AHEI present with fever [10,11]. The patient in our case did not have a documented fever throughout her illness. Based on their observations, Krause et al. proposed a diagnostic framework that does not require fever as a necessary symptom (Table 1); however, this framework may risk prematurely excluding the possibility of AHEI in patients outside the typical age range of 6-24 months [12]. Therefore, a more comprehensive set of diagnostic criteria is still needed.

Table 1. Proposed diagnostic criteria for AHEI [12]

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

The hallmark of AHEI is the characteristic skin lesions, which are typically described as erythematous, annular, medallion-shaped, or rosette-like purpuric plaques. The initial eruption may begin as a simple or popular wheal, or a macule that evolves into a nummular rash, and less commonly, directly into an annular pattern [13]. These lesions often cluster, coalesce, and have a rapid onset, primarily affecting the face and extremities, while sparing the trunk and mucosal membranes. Oedema commonly involves the feet, hands, face, and auricles. In most children with AHEI, the skin oedema is characterized by tenderness, induration, and an absence of indentation when pressure is applied [13]. In males, the scrotum may also be affected [14]. These distinctive skin lesions are typically the first clue that prompts clinicians to consider AHEI as a potential diagnosis.

Despite the dramatic onset of skin manifestations, children with AHEI generally appear well, and laboratory results are often non-diagnostic, which can further complicate the diagnosis. Common findings in laboratory workups may include increased erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), as well as mild thrombocytosis and leucocytosis—none of which are specific to AHEI [15]. Therefore, careful clinical observation and a strong knowledge of AHEI remain crucial for making the diagnosis and avoiding unnecessary invasive procedures, such as skin biopsy. If a skin biopsy is performed, it typically shows features consistent with leukocytoclastic vasculitis, such as perivascular neutrophilic infiltration with scattered nuclear fragments that invade the vascular wall, leading to fibrinoid necrosis. Many experts

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argue that invasive diagnostic procedures can be avoided in favour of clinical recognition and careful monitoring, particularly as clinicians become more familiar with the clinical features of AHEI. Cucinotta et al. propose that skin biopsy is not necessary for diagnosing AHEI and should be reserved only for uncertain cases [16].

The management of AHEI remains somewhat controversial. Some studies have suggested the use of corticosteroids (either orally or as high-dose methylprednisolone) [17-18] and antihistamines for treatment. However, there is growing consensus that a watchful waiting approach is more appropriate, as recent evidence indicates that most children recover completely within one to three weeks without long-term complications, regardless of corticosteroid use [19]. The recurrence of AHEI is extremely rare, further supporting conservative management. While rare complications may occasionally warrant the use of steroids, their efficacy in this context remains uncertain [20-21]. Given the rarity of the condition, current recommendations continue to favour a conservative approach, emphasizing close observation and supportive care over more aggressive treatments.

4.0 CONCLUSION

AHEI is a rare, self-limiting condition that, despite its alarming appearance, is generally benign. Accurate diagnosis is critical to avoid unnecessary investigations and to distinguish AHEI from more serious conditions. In most cases, conservative management is sufficient, and spontaneous recovery occurs in most patients. Increased clinician awareness of AHEI helps ensure timely diagnosis, reduces unnecessary testing, provides appropriate care, and facilitates effective parental counselling, all of which help lower healthcare costs and reduce patient anxiety. Correct diagnosis is essential to prevent unnecessary workup and treatment, while also allowing for monitoring of rare but potentially severe complications.

5.0 CONFLICT OF INTEREST

The authors declare no conflicts of interest.

6.0 AUTHORS CONTRIBUTION

Lim, P. P. (Conceptualization; Literature review; Writing - original draft; Writing - critical revision of the article for important intellectual content)
Sivanesan, R. (Clinical data collection; Writing- contributed to manuscript drafting)

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