CASE REPORT

A RARE ETIOLOGY IN AN INFANT PRESENTİNG WITH ECCHYMOSIS: ACUTE INFANTILE HEMORRHAGIC EDEMA

Ekimoz İle Başvuran Bir İnfantta Nadir Bir Etyolojisi: Akut İnfantil Hemorajik Ödem

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ABSTRACT

Acute Infantile Hemorrhagic Edema (AIHE) is a rare leukocytoclastic vasculitis that primarily affects small vessels in the skin, commonly seen between the ages of 6 and 24 months. It is characterized by ecchymosis, purpura, and edema on the skin. The condition presents a benign course and typically resolves within 1-3 weeks. This article presents the case of a 9-month-old girl who presented with widespread ecchymosis and edema in her legs, was diagnosed with Acute Infantile Hemorrhagic Edema, and received intravenous (IV) methylprednisolone treatment. AIHE is an important differential diagnosis to consider in young children presenting with ecchymosis. Awareness about this condition is of utmost importance.

Keywords: *Ecchymosis; İnfant; Edema; Vasculitis; Steroids*

ÖZET

Akut infantil hemorajik ödem (AİHÖ), çoğunlukla 6–24 aylar arasında görülen, ciltteki küçük damarları tutan, nadir görülen lökositoklastik bir vaskülittir. En sık görülen klinik bulgusu ciltte ekimoz, purpura ve ekstremite ödemidir. Ekimozlar ani başlangıçlı, simetrik, daha çok yüz, kulak kepçesi, kol ve bacak yerleşimli, sınırları belirgin, olup çapları 1–5 cm arasında değişen palpabl purpurik, ekimotik plaklar şeklindedir. Lezyonlar ağrılı ve ödemli olabilirken genellikle birleşme eğilimi gösterir. Gövde genellikle korunurken genital bölgede de döküntü gelişebilir. Bazı olgularda ateş eşlik edebilmektedir.

Bu yazıda Akut İnfantil Hemorajik Ödem tanısı ile izlenen ve intravenöz (IV) metilprednisolon tedavisi verilen 9 aylık kız hasta sunulacaktır.

Anahtar Kelimeler: *Ekimoz; İnfant; Ödem; Vaskülit; Steroid*

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INTRODUCTION

Acute Infantile Hemorrhagic Edema (AIHE) is a rare leukocytoclastic vasculitis that predominantly affects small vessels in the skin, commonly observed between the ages of 6 and 24 months (1). The most frequent clinical manifestations include ecchymosis, purpura, and edema of the extremities (1,2). The ecchymoses are characterized by their sudden onset, symmetric distribution, predominantly on the face, auricles, arms, and legs, with well-defined borders, appearing as palpable purpuric, ecchymotic plaques ranging from 1 to 5 cm in diameter. Lesions can be painful and edematous, often tending to merge. While the trunk is usually spared, eruptions may occur in the genital area. Some cases may present with accompanying fever (2).

This article will present the case of a 9-month-old girl diagnosed with Acute Infantile Hemorrhagic Edema and treated with intravenous (IV) methylprednisolone.

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CASE REPORT

A 9-month-old female patient with a history of term birth presented to the emergency department due to bilateral bruising and swelling in the lower extremities. It was noted that the complaints had a sudden onset and these lesions on her legs increased from proximal to distal. There was no fever, no history of trauma, no recent vaccinations within the last month, and no medication usage. The patient had no other accompanying complaints.

Initial evaluation revealed a temperature of 36.7°C, respiratory rate of 34/minute, heart rate of 130/minute, and blood pressure of 80/50 mmHg. Physical examination revealed widespread palpable ecchymosis, the largest being 5-6 cm in size bilaterally in the lower extremities, some tending to merge, with irregular borders and non-blanching features. Edema was present in both lower extremities and on the dorsum of the feet (Image-1,2). No warmth or pallor was detected. Other system examinations were within normal limits.

Laboratory investigations showed Hemoglobin: 10.7 g/dL, MCV: 69.5 fL, Leukocytes: 11,400/µL, Platelets: 541,000/µL, INR: 0.9, CRP: 3.8 mg/L, ESR: 44 mm/h. Liver and kidney function tests and urinalysis were normal. Arterial and venous Doppler imaging of the lower extremities was unremarkable.

The patient was diagnosed with acute infantile hemorrhagic edema and was admitted for observation. Intravenous hydration was initiated. No additional treatment was administered initially as the patient had no fever and her general condition was stable. On the 3rd day of observation, there was a significant increase in ecchymosis and edema in both extremities. Coagulation tests were repeated, showing Hemoglobin: 9.9 g/dL, MCV: 69.4 fL, Leukocytes: 11,800/µL, Platelets: 501,000/µL, INR: 0.8. Due to increased ecchymosis and extremity edema, IV methylprednisolone was initiated at a dose of 2 mg/kg/day. Subsequently, regression of ecchymosis and extremity edema was observed (Image-3). On the 7th day of treatment, no ecchymosis or extremity edema was observed, and the patient was discharged.

Consent for the case report was obtained from the patient's family.

DISCUSSION

Acute Infantile Hemorrhagic Edema (AIHE) is an isolated cutaneous leukocytoclastic vasculitis observed in children under the age of 2 (3). It is more frequently seen in males (2,3). While there are no definitive data on its incidence, a case series by Parker et al. reported an incidence of 0.7 among 1000 patients presenting (4).

Previously, AIHE was considered a subtype of Henoch-Schönlein Purpura (HSP) (2,3). However, due to less frequent occurrence of immunoglobulin A (IgA) deposition in vessel walls in skin biopsies, it is currently regarded as a separate entity (2-4). Histopathological analysis, conducted in approximately 50% of AIHE cases reported in the literature, demonstrates leukocytoclastic vasculitis of the dermal vessels with fibrinoid necrosis, extravasation of red blood cells, and leukocytoclasia. Direct immunofluorescence examination, performed in approximately one third of the cases with biopsy studies, shows vascular deposits of immunoglobulin A in no more than one quarter of the cases (5).

The etiology of the disease remains uncertain (1-3). It is believed to be associated with prior bacterial and viral infections, vaccination, and the use of antibiotics and non-steroidal anti-inflammatory drugs (3,4). In our case, no specific cause was identified.

The diagnosis of AIHE is generally made based on history and typical clinical findings (1,2). There is no specific laboratory test, and laboratory examinations are usually within normal ranges. However, elevated erythrocyte sedimentation rate, C-reactive protein, leukocytosis, lymphocytosis, thrombocytosis, eosinophilia, and transient abnormalities in liver function tests may be observed (3-5). Our patient also exhibited leukocytosis, elevated CRP, and sedimentation rate.

The disease has a benign course and usually resolves within 1-3 weeks, with recurrences being uncommon. It is crucial to reassure parents about the benign nature of the lesions as ecchymosis can be distressing for them (1-3). While systemic involvement is not expected, cases with systemic involvement have been reported in the literature (4,6).

In a case series by Parker et al., arthritis was reported in 50% of cases, gastrointestinal bleeding in 15.3%, hematuria in 3.8%, and compartment syndrome in 3.8%.

Bozok Tıp Derg 2024;14(4):286-289
Bozok Med J 2024;14(4):286-289

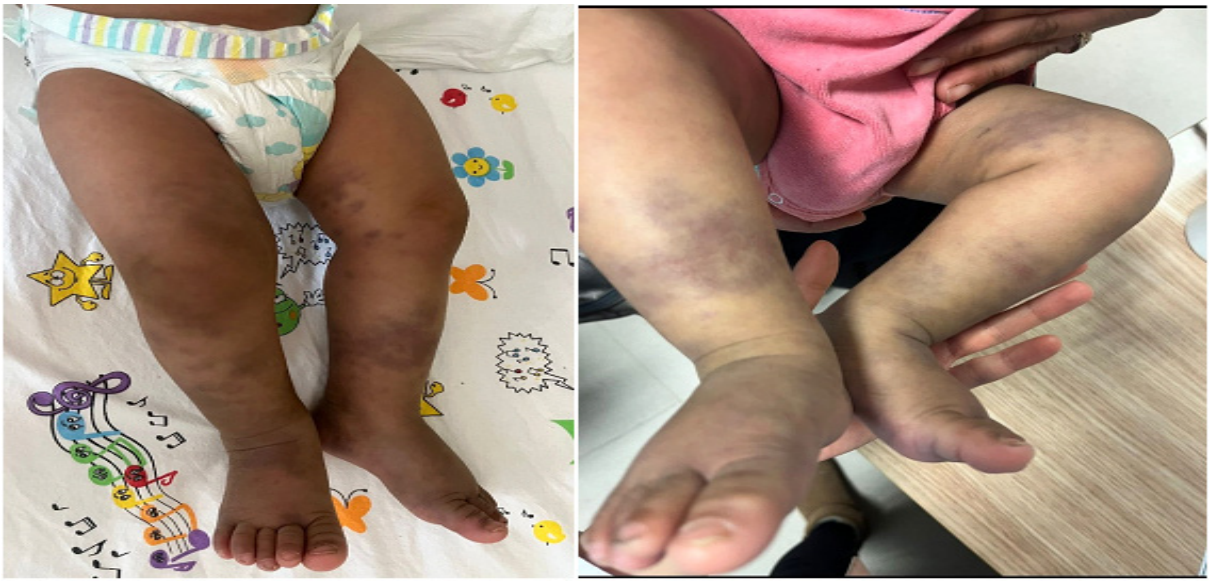

Figure 1,2. Ecchymosis and skin edema in both lower extremities

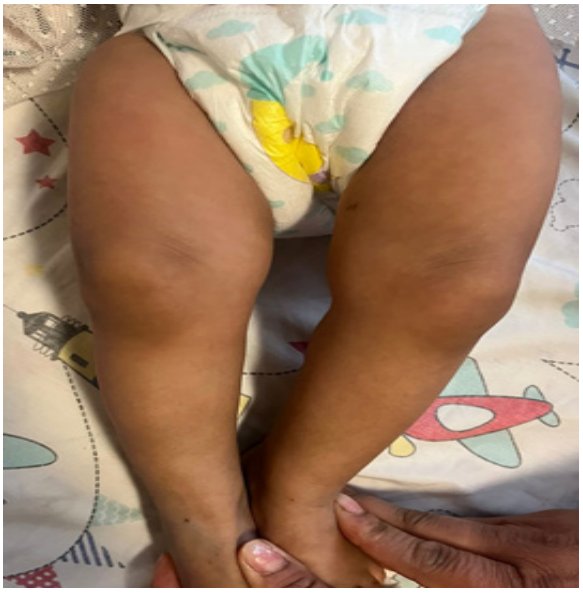

Figure 3. 7th day of IV steroid treatment

No systemic involvement was observed in our patient (4).

It is important to differentiate between AIHE and HSP with HSP for diagnosis (2-4). Although some authors consider AIHE as a variant of HSP in young children due to similarities in etiology and histopathology, most publications support that AIHE should be recognized as a separate pathology (3-5). The most significant features that distinguish AIHE from HSP include its occurrence in children under two years of age, the nature of skin lesions, the absence of renal and gastrointestinal involvement, and the rarity of recurrence (3,4).

Considering other differential diagnoses, meningococcemia, erythema multiforme, urticariawith a hemorrhagic component, Kawasaki disease,

Bozok Tıp Derg 2024;14(4):286-289
Bozok Med J 2024;14(4):286-289

skin manifestations of sepsis, drug eruption, Sweet syndrome, insect bites, and abuse should be considered (3,4). AIHE presentation can be easily distinguished clinically from these conditions. Due to hospital limitations, complement levels for differential diagnosis could not be assessed in our patient. Similarly, skin biopsy could not be performed due to hospital constraints. It is not necessary to demonstrate leukocytoclastic vasculitis in a skin biopsy to establish the diagnosis of AIHE (8).

Clinically, palpable purpura and ecchymosis without accompanying systemic symptoms led to the consideration of AIHE in our patient.

There is no specific treatment for AIHE (2-4). The use of steroids and antihistamines in treatment remains controversial (2-4). Due to the benign course of the disease, some authors do not recommend steroid use (5,6). However, due to its potential to expedite recovery, some authors suggest the use of steroids and antihistamines (9). In our case, intravenous (IV) methylprednisolone therapy was initiated due to bilateral increase in ecchymosis and marked increase in lower extremity edema observed on the 3rd day of observation.

CONCLUSION

Acute Infantile Hemorrhagic Edema is an important differential diagnosis to consider in young children presenting with ecchymosis. Awareness of this condition is crucial. In our case, while initially observed without treatment, the diagnosis of AIHE led to the administration of intravenous methylprednisolone due to increased ecchymosis and pronounced lower extremity edema. On the 7th day of treatment, the absence of extremity edema and significant regression of ecchymosis were observed, indicating the perceived benefit of IV methylprednisolone therapy. As there is insufficient evidence in the literature regarding AIHE treatment, this case has been shared

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