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Case Report

Acute Haemorrhagic Oedema of Infancy

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Abstract
Acute haemorrhagic oedema of infancy is a small vessel leucocytoclastic vasculitis seen in young children less than 2 years of age. it is e benign self-limiting condition which is greatly under diagnosed and under reported. its aetiology is not known but it is associated with upper respiratory tract infections, vaccination and drug intake in most of the cases.
Keywords: *Acute Haemorrhagic Oedema of Infancy, Leucocytoclastic, Benign, Idiopathic, Purpuric Rash, Oedema, Henoch Schonlein Purpura.*

Introduction

Acute haemorrhagic oedema of infancy is an isolated leucocytoclastic vasculitis that affects infants and young children less than 2 years of age. It manifests as purpuric target like rash with swelling of the face and the extremities. Usually there is no visceral involvement. It may be idiopathic and sometimes prior vaccinations, viral infections and drugs may be considered as the inciting agents.

Clinical Case

An 8 months old infant presented to us with complaints of purpuric rash over the face and extremities associated with swelling of the hand and feet. There was preceding history of cough and runny nose. There was no fever, no prior vaccination and no history of any drug intake, no history of any bowel or bladder complaints. On examination, child was alert, active, afebrile and playful. There were erythematous, purpuric, nonpalpable target like lesions over face, extremities and in the buttocks, of varying sizes from 0.5-5cm in diameter (fig1-2). There was oedema of both upper and lower limbs(fig.3-4), no tenderness was elicited. The systemic examination was normal. The laboratory investigations including complete haemogram, C3, C4 levels, coagulation profile, urine analysis and liver and kidney function tests were within the normal range. The histological examination of skin biopsy showed mild hyperkeratosis with spongiosis in epidermis. Dermis showed oedema and periadenexal inflammatory infiltrate. Small blood

vessels showed focal neutrophilic infiltrate and concentric thickening of thin walls, with nuclear dust and fibrinoid changes; the features were compatible with the diagnosis of leucocytoclastic vasculitis. The immunofluorescence studies could not be done due to financial constraints. Child was managed with oral azithromycin and antihistaminic and calamine lotion for local application. The child showed an uneventful recovery over next 3-4 days. Child was discharged on D7 of admission without any squelae.

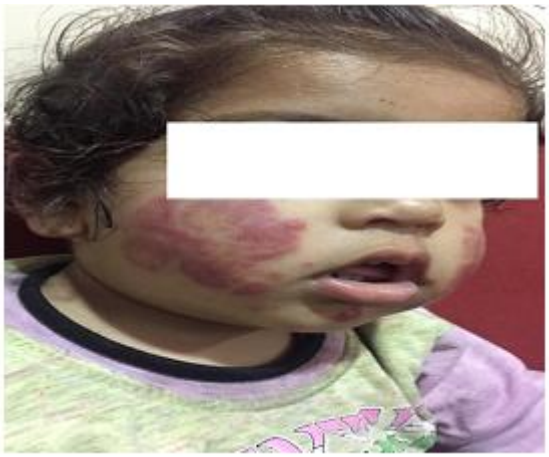

Fig.1

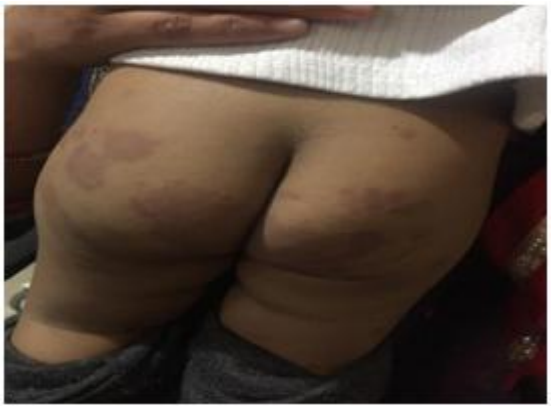

Fig.2

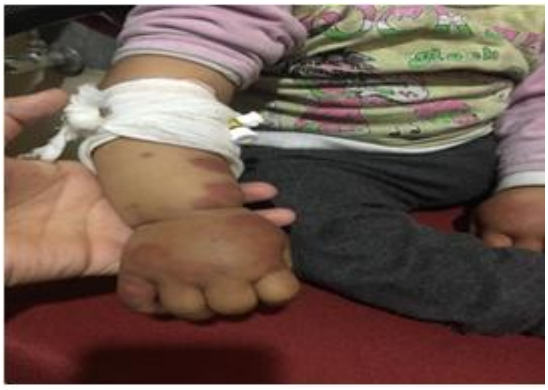

Fig.3

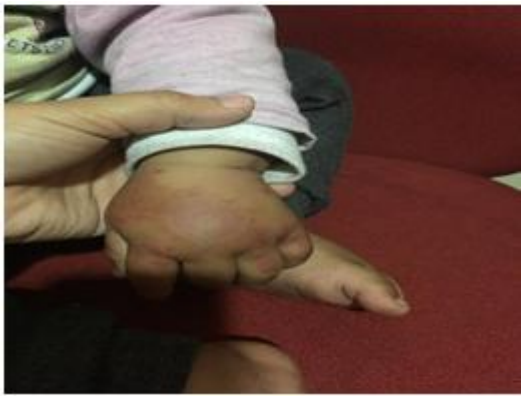

Fig.4

Discussion

The clinical features of our case, characterized by a dramatic onset of typical erythematous purpuric lesions with histology showing leukocytoclastic vasculitis and a spontaneous recovery, were consistent with the diagnosis of AHEI. AHEI was first described by Snow in 1913[1]. AHEI is also known as Finkelstein disease[2] or Seidlmayer syndromeor medallion-like purpura or infantile post infectious iris-like purpura and edema. It is a benign, small-vessel leukocytoclastic vasculitis, which typically affects children below 2 years of age and is characterised by erythematous purpuric and ecchymotic, target like lesions over the extremities, face, and ears with relative sparing of trunk. Fever and oedema of the distal extremities, ears, and eyelids are associated features of AHEI[3]. Mucosa is rarely involved but has been reported[4]. Visceral involvement is rare but it has been reported involving the kidneys and intestines, causing symptoms such as haematuria, mild proteinuria, and bloody diarrhoea[5]. Despite the explosive onset of the skin lesions, the general condition of the patient remains good. The exact aetiology of the disease is not known. But preceding viral or bacterial infections, vaccination and drug intake has been considered the triggering factors in many cases[6]. Histopathologicl examination is suggestive of small vessels leucocytoclastic vasculitis. AHEI needs complete investigations to differentiate it from other conditions like HSP, Meningococcemia, Septicemia, Kawasaki Disease, And Erythema Multiforme and Purpura Fulminans. HSP remains

close differential for AHEI and some authors consider AHEI variant of HSP. AHEI differs from HSP by its occurrence in children below 2 years of age, more rapid resolution of the lesions, negative staining for IgA on direct immunofluorescence (DIF), and lack of internal organ involvement and AHEI has better prognosis. Children with AHEI are to be managed symptomatically with no specific treatment. The role of corticosteroids in the management of AHEI is controversial. The disease runs a benign course and spontaneous resolution of lesions without any sequelae occurs within 2-3 weeks. Rarely, episodes of AHEI may recur.

To summarize, we have described typical case of AHEI associated with upper respiratory tract infection in male child who presented with purpuric/ecchymotic lesions and oedema of the extremities. Awareness about this uncommon but benign form of leucocytoclastic vasculitis will help to avoid unnecessary treatments, investigations, and anxiety for parents as well as clinicians.

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