Acute hemorrhagic edema of infancy

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ABSTRACT

Acute hemorrhagic edema of infancy is a rare form of leukocytoclastic vasculitis. Mostly it appears under three years of age and is characterized by purpuric skin lesions, fever and edema. A three years-old boy, who has cough and coryzea was admitted to our clinic for fever and red spots on legs and arms. In physical examination; ecimotic skin lesions on right ear, face, arms, dorsal of the hands, buttocks, legs and dorsal of the feet were found. In the laboratory tests acute phase reactants were elevated and blood coagulation tests were in normal range. Hepatit A,B,C and TORCH markers were negative. Punch biopsy obtained from gluteal area showed leukositoclastic vasculity. Focal fibrinogen accumulation was detected by immun fluorescent microscopy. Regression on lesions was not observed despite supportive therapy, so prednisolone (1 mg/kg/day) therapy was started. On the third day of the steroid therapy, complete recovery was achived.

Key words: Hemorrhagic edema of infancy , leukocytoclastic vasculitis, steroid therapy

Bebeklik dönemi akut hemorajik ödemi

ÖZET

Akut infantil hemorajik ödem lökositoklastik vaskülitin bir formu olup, daha çok üç yaş altı çocuklarda ateş, ödem ve purpurik deri lezyonları ile karakterizedir. Üç yaşında öksürük ve soğuk algınlığı olan erkek çocuk kliniğimize bacak ve kollarında kırmızı renkli döküntü, ateş nedeni ile başvurdu. Fizik incelemede sağ kulak üstünde, yüzünde, kollarında ve ekstremitelerinde ekimotik lezyonlar bulundu. Laboratuvar sonuçlarında akut faz reaktanları pozitif, koagulasyon testleri ise normal idi. Hepatit A,B,C ve TORCH belirteçleri negatif idi. Gluteal bölgeden alınan punch biyopsi sonucu lökositoklastik vaskülit ile uyumlu ve immünfloresan incelemede lokal fibrinojen birikimi tespit edildi. Destek tedavisine rağmen lezyonlarında gerileme gözlenmeyen hastaya bu nedenle 1mg/kg/gün prednizolon tedavisi başlandı. Tedavinin üçüncü gününde lezyonlar tamamen düzeldi.

Anahtar kelimeler: infantil hemorajik ödem, lökositoklastik vaskülit, steroid tedavisi

INTRODUCTION

Acute hemorrhagic edema of infancy (AHEI) is a leukocytoclastic vasculitis; clinically characterized by purpuric skin lesions, fever and edema[1]. There is an abrupt onset of the large cockade, annular, or targetoid lesions involving the face, ears, and extremities. Scrotal lesions have been reported rarely. The edema is nontender and mostly symetric[2].The disease must be considered on discrimination of meningococcal infection, septicemia, purpura fulminans and other purpuric skin eruptions especially Henoch schönlein purpura[3,4]. In this case, three years old, asymetric skin lesions mostly on right and edema, was presented.

CASE REPORT

A three years old boy, had complaint of fever and red spots on legs and arms. The complaints of cough and coryzea had started three days ago, then red spots on the legs were realised in the evening of the next day by his parents. This situation became widespread on face, legs and arms in the morning of the third day. It was reported that the patient had not

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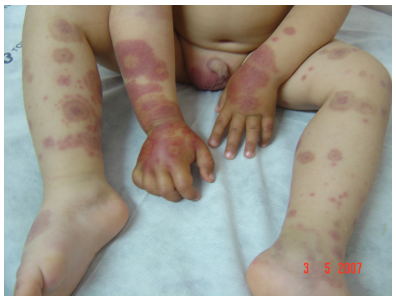
been vaccinated or drugged in earlier time. In physical examination; weight:14 kg, height:96 cm, temp:37.5ºC, blood pressure:100/70 mmHg, there were ecimotic skin lesions on right ear, face, arms, dorsal of the hands, buttocks, legs and dorsal of the feet (Figure 1). The laboratory resulted values: hemoglobin: 10gr/dl, amount of leukocytes in blood: 9.700/mm³, eriythrocytes: 5.1million/mm³, trombocytes: 360.000/mm³, eriythrocyte sedimentation rate (ESR): 41mm/h, C-Reactive Protein (CRP): 90mg/dl, ASO 25 todd/U, Romatoid factor: 20 IU/ml, fibrinogen 392mg/dl. Periferic blood smear: 56% polimorf nuvel leukocytes (PNL) 44% lymphocyte, activated partial tromboplastin time (aPTT) 26.1sec, prothrombin time (PT) 12.6sec, INR 1.05, serum biochemical parameters, immunglobulins, C3, C4, Antitrombin III, protein C, protein S, eozinofil katyonic protein (ECP) in normal values, salmonella and brucella antibodies were negative in serology study. Hepatit A,B,C parameters were negative in elisa test other than depending on vacine antiHbS 1000 IU/L, TORCH markers negative, direct urine examination normal. Abdomen and scrotal ultrasonography, bilateral lower extremity deep vena-arteria system colored doppler research were in normal limits. Punch biopsy resulted from gluteal area cross-section. Under the multi storey epitel, near the vascular or on vascular walls mixed type inflammation cell infiltery includes neutrophils were detected and accommodated with leukositoclastic vasculity. Focal fibrinogen accumulating was detected by immun fluorescent microscopy. AHEI was diagnosed by all these clinical studies and symptoms. Despite the supportive therapy regression on lesions was not observed. On the eighth day, the patient still had not recovered completely and the laboratory studies showed; leukocytes: 10.500/mm³, eriythrocytes: 4.99million/mm³ thrombocytes: 354.000/mm³ , CRP: 8.65mg/dl, ESR: 32mm/h. Steroid (prednisolone 1mg/kg/day) therapy was given. On the third day of the steroid therapy, complete recovery was reached. In the follow up no complication was detected.

Figure 1: The asymmetric purpuric lesions and edema which located dominantly on the right side of the body are seen.



DISCUSSION

The etiology of the AHEI is not known exactly today and 12% of leukocytoclastic vasculitis are AHEI, many of them are seen between four month to two years old, not depending on gender and especially the patients who have a past with having cold before[5]. This disease is defined as an immunologic vasculity which progresses against different antigenic stimulants. 75% of the cases have a past of having an infection, using drugs or immunization background[3]. Patient had not been vaccinated or drugged in earlier time. Especially purpuric skin lesions on face, buttock and extremities medallion like ecimotic purpura with inflammation edema on extremities and face are two important characteristic about AHEI[6,7]. In our case, there is an ecimotic purpura on scrotal area which is unusual. Beside this, the disease was not in symetric character in clinically, the right side was dominant in spite of its characteristic specialitiy of symetrical progression. There are some data support that immun complexies pathogenesis of leukocytoclastic vasculitis cause derms to be poisoned by settling on vessel walls. The starters of the immun complex producing on leukocytoclastic vasculitis are not known well. Furthermore viral and bacterial infection, drugs and chemicals with other proteins are put forward as etiologic factors. The pathogens from

infections that have the best known connection with leukocytoclastic vasculity are: group A or B hemolytic streptococci, Staphylococcus aureus, Mycobacterium leprae, hepatitis B and C viruses, HIV, and cytomegalovirus. The drugs which cause leukocytoclastic vasculity such as various antibiotics (penicilin), thiazids and some nonsteroid antiinflammatories are reported[3]. Several reports have described cases that systemic corticosteroids and antihistaminics are ineffective on therapy of cutaneous lesions[5]. Our patient who had cutaneous lesions which had not been retreating with supportive therapy for eight days has recovered with steroid therapy for three days. In this study we emphasize that steroid might be need on therapy of AHEI and seperative diagnosis must be claimed.

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