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Infantile Acute Hemorrhagic Edema: Four Attacks in Two Years

İnfantil Akut Hemorajik Ödem: İki Yılda Dört Atak

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Özet

İnfantil akut hemorajik ödem (İAHÖ), ani başlangıç ve ekimotik purpura ile karakterize hedef ve rozet şeklinde inflamatuar ödemle seyreden benign bir hastalıktır. Genellikle 2 yaşın altındaki çocuklarda görülür ve neredeyse hiç rekürrens gözlenmez. Bu hastalık kutanöz vaskülitin nadir bir formu olarak düşünülür. Burada biz kliniğimize başvuran ve İAHÖ tanısı alan bir kız çocuğunu bildirdik. Döküntü ilk kez 2 yaşın altında başlamış ve 2 yıl içerisinde 6 ay arlıklarla 4 kez tekrar etmiş. Literatürde relaps çok nadir olarak bildirilmiştir, biz bu vakayı hastanın 2 yaşının üzerine olması ve toplamda 4 atak geçirmesi üzerine seçtik.

Anahtar Kelimeler: İnfantil hemorajik ödem, purpura, relaps.

Abstract

Infantile acute hemorrhagic edema (IAHE) is a benign disease which is characterized by target and rosette-shaped inflammatory edema which has a dramatic beginning and characterized by ecchymotic purpura. It is generally seen in infants under 2 years of age and recurrence is hardly observed. It is thought that this disease is a rare form of cutaneous vasculitis. Here we reported a 4-year-old girl who applied to our clinic and was diagnosed IAHE. Serpigo first started at the age of 2 and repeated 4 times in 2 years with 6-month intervals. As the relapse was seldom reported in the literature, we chose to report this case owing to the fact that the patient was over 2 years old and had 4 attacks in total.

Keywords: Infantile acute hemorrhagic edema, purpura, relapse.

Introduction

Infantile acute hemorrhagic edema (IAHE) is a skin disease seen in infants between 4 months and 2 years of age and also seen in face, legs and acral areas following high fever and characterized by large rosette shaped purpuric lesions. Internal organ involvement is not observed and it spontaneously regresses in 3 weeks. Some authors consider the disease as a purely cutaneous form of Henoch-Schönlein purpura, and others believe that IAHE should be regarded as a distinct clinicobiologic entity within the spectrum of leukocytoclastic vasculitis (1-3).

Although it was first described by Snow et al. in the USA in 1913, this disease was then called the infantile acute hemorrhagic edema of the skin or post-infectious rosette pattern. However the Eurpoeans described it like Finkelstein disease. It has not been classified in European countries yet (4,6). Its histopathological examination shows leukocytoclasis vasculitis.

Case Report

Having bleariness on face, arms and legs and swollen lesions in the skin, a four-year-old girl from pediatric outpatient department was consulted. Her complaints had started 2 days ago and red swellings occurred on face, arms and legs following high fever. She had the same disease first when she was 2 years old. Following high fever, there had been bleariness and swellings on cheeks, ears, and legs. Her complaints regressed in one week and purple spots disappeared in 10 days. She has had serpigo 4 times in total since she was 2 with 6-month intervals. Before serpigos, she had no medication and upper respiratory infection story.

During her dermatologic examination, there were erythema, pale centered, swollen and rosette shaped ecchymotic plaques and they were multiple plaques on the left cheek, a few plaques on the right cheek and multiple plaques upper and lower extremities (Figure 1, Figure 2 and Figure 3).

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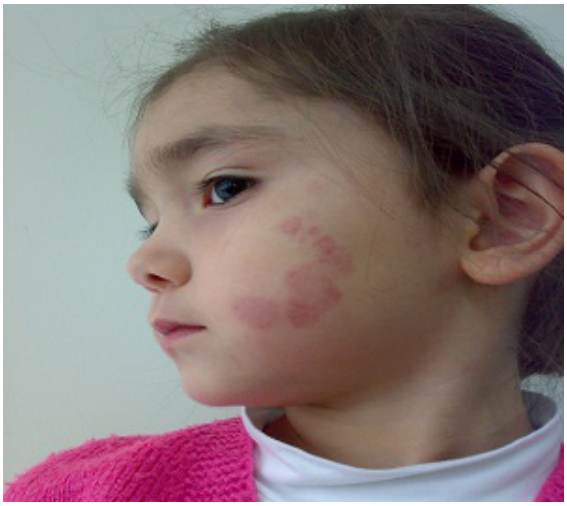

Figure 1: Multiple rosette-shaped purpuric plaques on left cheek.

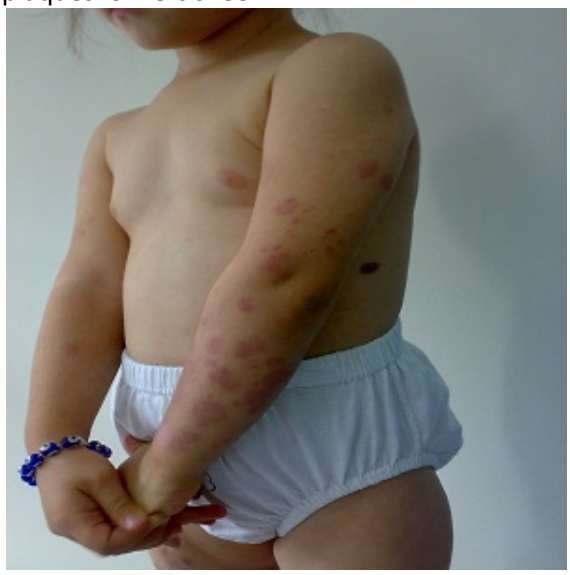

Figure 2: Multiple rosette-shaped purpuric plaques on upper extremities.

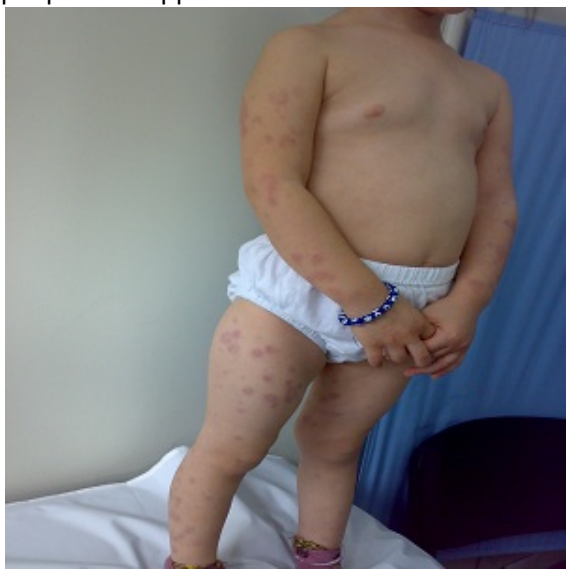

Figure 3: Multiple rosette-shaped purpuric plaques lower extremities.

With infantile acute hemorrhagic edema and erythema marginatum pre-diagnosis, skin biopsy was taken from the patient. In the histopathologic examination; a small venule show striking fibrinoid necrosis and nuclear dust around the venul. Also so many extravase red blood cells can be seen easily (Figure 4a,b).

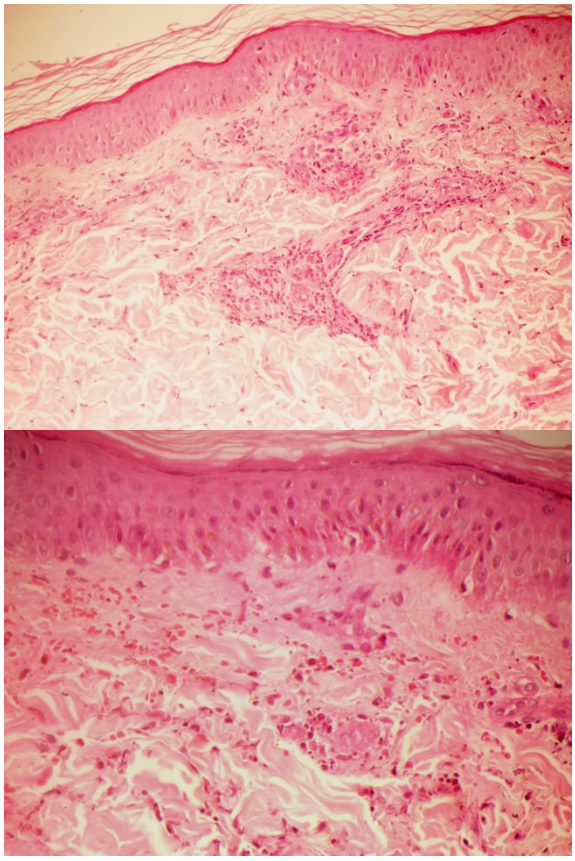

Figure 4a (HEx100): A small venule show strinking fibrinoid necrosis and nuclear dust around the venul. Also so many extravase red blood cells can be seen easily.

Figure 4b (HEx 200): High power view of leukocytoclastic vasculitis.

With clinical histopathologic findings, infantile acute hemorrhagic edema was diagnosed in the patient. As it was a benign course disease and the lesions regressed spontaneously in 1-2 weeks, the treatment was not launched. In the control after 10 days, it was observed that the lesion regressed completely. The patient had no clinical findings such as nuisance and nausea-vomiting except cutaneous finding. Detailed examination was carried out in terms of internal organ involvement but a systematic finding was not detected.

Discussion

IAHE is regarded as an uncommon benign form of cutaneous vasculitis which forms in infants less than 2 years of age. The disease frequently occurs in winter months. It has little male domination. Etiology is still not known although 75% of the patients have upper-respiratory infection, urinary system infection, medication or immunization story. Rotavirus infection is the most important cause of nosocomial infections of childhood period and one of the most significant pathogens of this period in winter months. Therefore it is difficult to distinguish that the primary reason inducing the disease is resulted from this infection (1-5).

IAHE clinically; urticarial lesions and maculopapular rash are seen on chin,ears and extremities with fever and it is followed by target and medallion like purpura. Similar lesions were also reported in external genital (5,6). It is generally observed in infants under two years. The disease regressed in weeks without internal organ involvement.

The histopathologic features of IAHE is concordant with small vessel vasculitis holding middle and upper derme of capillary and postcapillary venules. The results of direct immunofluorescent studies show the presence of IgM, C3 and fibrinogen. These are not clear for the diagnosis. In addition, IgA storage was found in 1/3 of the cases (1,7).

In the differential diagnosis, Sweet syndrome, erythema multiforme, Kawasaki disease, purpura fulminans, purpura induced by trauma and granuloma faciale must be considered. The diagnosis is generally made by personal background, physical examination, laboratory studies and histological inspection.

It is still being discussed whether the IAHE is a benign variant of HSP or a separate clinical table in leukocytoclasis vasculitis spectrum. Though IAHE and HSP overlap cases are reported, most authorities think that these two diseases have different clinic and pathology. The outset age of IAHE is lower than HSP (2-24 months) and when compared with HSP, it is limited to the skin and there is less rash in the skin according to HSP. Mostly relapse does not happen in this disease. In our patients, the lesions were seen at the age of 2 and relapsed four times with 6-month intervals until 4 years of age. HSP is often seen at 3-7 ages, and papulopetechial and urticarial lesions are seen on the extensor side of legs and hips (1,4). While systemic complications such as arthralgia, gastrointestinal hemorrhage and nephrite are common in HSP, these are rare in IAHE. General course of the disease is 12-20 days and relapse rate is much lower than HSP (4).

In 10-case serial paper with IAHE by Legrain et al., they reported 2 attacks in 2 patients, 4 attacks in 2 patients and no relapse in the other 6 patients. Legrain et al. think that these two diseases are overlap (5).

Consequently, IAHE is benign coursed form of leukocytoclastic vasculitis which is limited to the skin. Although frequent relapse was not seen in the patients, total 4 attacks were seen between 2-4 years of age in our patient.

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