Henoch-Schönlein Purpura Secondary to a Vaccine Preventable Disease: Varicella

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✔ Henoch-Schönlein purpura (HSP) is one of the most common small vessel vasculitis of children. In this study, a 14-year old girl with HSP secondary to varicella infection with gastrointestinal and mild renal involvement is presented.
Key words: *Henoch-Schönlein purpura, varicella, vasculitis*

✔ **Aşı ile Önlenebilir Bir Hastalık Olan Su Çiçeğine Bağlı Henoch-Schönlein Purpurası**
Henoch-Schönlein Purpurası (HSP) çocukların en sık görülen vaskülitlerinden biridir. Bu olgu bildiriminde Varisella enfeksiyonuna ikincil olarak gelişen, hafif gastrointestinal ve renal tutulumu olan, 14 yaşında bir kız HSP olgusu sunuldu.
Anahtar kelimeler: *Henoch-Schönlein purpurası, varisella, vaskülit*

INTRODUCTION

Henoch-Schönlein purpura (HSP) is one of the most common small vessel vasculitis of children characterized by palpable purpuric rash, arthritis, renal and gastrointestinal involvement. Many causes have been proposed to account for the etiology and pathogenesis of HSP including upper respiratory tract infections, food allergens, insect bites, exposure to cold and trauma[(1)]. However none of these factors have been confirmed to be of pathogenetic importance. Varicella is a highly communicable but preventable childhood disease with considerable complications. Herein we present a patient who developed HSP following varicella zoster infection.

CASE REPORT

A 14-year old girl was admitted with abdominal pain, arthralgia and palpable purpuric rash over her legs for three days. It was learned that she had varicella zoster infection 15 days ago. On physical examination, she had a few hyperpigmented macules and crusted papules typical for varicella over the trunk. She had also newly appeared palpable purpuric vasculitic lesions on the lower extremities typical for HSP (Figure). The left knee was slightly swollen and tender. The child had not been immunized against varicella. Laboratory results were as follows; Hemoglobin: 10.6 g/dL, leukocytes: 13900/mm^3, platelets: 346000/mm^3, BUN: 24 mg/dL, creatinine:0.8

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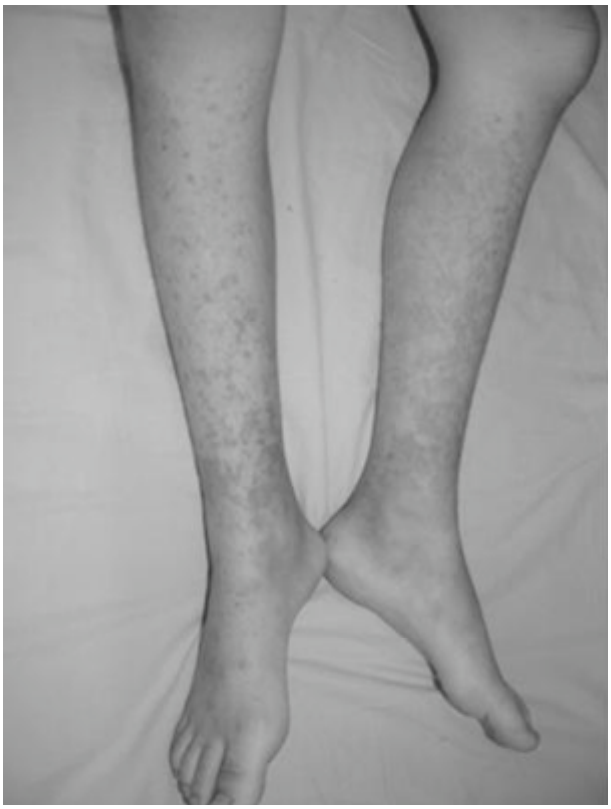

Figure. Typical palpable purpura over the legs of the patient.

mg/dL, Na: 137 mEq/L, K: 4.01 mEq/L, total protein: 7.2 gr/dL, albumin: 3.9 gr/dL, erythrocyte sedimentation rate: 150 mm/h and occult blood in stool: positive. In urinalysis, there were 10-15 erythrocytes per high power field on microscopy without proteinuria or any other abnormality. Varicella zoster virus IgM was 4.6 U/ml (0.0-1.0) and IgG was 3.2U/ml (0.0-0.6) reflecting the convalescence period of acute varicella infection. Abdominal ultrasound examination was normal. The patient was diagnosed as Henoch Schönlein purpura developed secondary to varicella zoster infection. Prednisolon (1 mg/kg/day) was started for the gastrointestinal involvement and tapered within two weeks. Renal biopsy was not considered since the patient had only microscopic hematuria without proteinuria.

DISCUSSION

The case presented in this report, had HSP following varicella infection. The exact cause of HSP is unknown and several theories concerning the pathogenesis have been proposed. Circulating immune complexes containing IgA are detected in the serum of patients as well as immune complexes containing C_3 and IgA in skin, intestines and kidneys[2]. Since Ig A is the prominent immunoglobulin in mucosal secretions acting as a defense against viral and bacterial agents, several authors indicated that HSP may be a post-infectious immune-mediated vasculitis[3,4]. Many bacteria and viruses such as *Streptococcus* β, *Yersinia*, *Mycoplasma*, *Toxoplasma*, measles, rubella, parvovirus, Epstein-Barr Virus, adenovirus, HIV and several others have been reported to be associated with HSP without direct evidence of causality. Viral antigens are thought to be responsible for vasculitic process through immune complex formation activating inflammatory pathways and cytokine release[5]. In our case, the triggering factor may be the varicella zoster specific viral antigens leading to development of HSP. Association of HSP with varicella zoster infections in the literature has been previously reported. In these reports HSP has been recorded to occur either prior to or following varicella infection[6–9]. In our patient, appearance of palpable purpuric rash following varicella infection strongly suggests a causal relation between varicella infection and HSP. Serological confirmation with varicella zoster specific IgM and IgG positivity also supports this association.

The majority of children with HSP nephritis who present only with hematuria and/or low-grade proteinuria as in our case have a good long term prognosis[10,11]. However, patients with massive proteinuria at onset frequently show a progressive course and some patients exhibit persistent urinary abnormalities and progress to terminal renal failure[11,12]. In many series evaluating the HSP nephritis, varying frequencies some of which are not neglectable at all for progression to chronic renal disease have been reported[11,13,14]. Therefore HSP, when its frequency among children is considered, does not seem such a

benign condition as once believed. Likewise, as an infectious problem, varicella is also quite common in pediatric age which is a highly communicable infection due to varicella–zoster virus. In normal host it is generally a benign self-limited disease. However, it may result in life-threatening complications such as purpura fulminans, pneumonia and encephalitis even in healthy children[15]. Although varicella is preventable by vaccination it is still a public health issue in low vaccination areas. After the implementation of routine immunization of children against varicella in the USA, the number of varicella cases and the associated morbidity and mortality have clearly declined[15,16]. Therefore, HSP seems to be another relatively rare complication of varicella infection justifying the recommendations for routine vaccination against varicella in developing countries.

In conclusion, together with the previous reported cases[6–9], we believe that HSP should be added to the list of complications of varicella infection.

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