



Cerebral Vasculitis in Henoch-Schönlein Purpura

Henoch-Schönlein Purpurasında Serebral Vaskülit

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Summary

Henoch-Schönlein purpura (HSP) is the most common systemic vasculitis of childhood, affecting the small blood vessels. The disease is a clinical syndrome characterized by multiple organ involvement including the skin, joints, gastrointestinal tract, and the kidneys. Headache and behavioral changes can be frequently seen during the course of the disease, but severe neurological signs such as seizures, focal neurologic deficits, intracerebral hematoma, mononeuropathies, and polyradiculoneuropathies are the rare complications of HSP. In this report, we described a 17-year-old girl with HSP who presented with seizures and disturbance of consciousness. The subsequent magnetic resonance imaging of the brain demonstrated findings consistent with cerebral vasculitis. The pulse steroid therapy was performed immediately after the diagnosis and clinical improvement was achieved rapidly. In conclusion, we suggest that cerebral vasculitis should be considered in all HSP patients with neurological symptoms and signs.

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Key Words: Henoch-Schönlein purpura; cerebral vasculitis; magnetic resonance imaging

Özet

Henoch-Schönlein purpurası (HSP) küçük damarları etkileyen çocukluk döneminin en sık sistemik vaskülitidir. Hastalık, cilt, eklemler, gastrointestinal sistem ve böbrekleri içeren multiple organ tutulumu ile karakterizedir. Baş ağrısı ve davranış değişiklikleri hastalık süresince sıklıkla görülebilir, ancak nöbet, fokal nörolojik defisit, intraserebral hematoma, mononöropatiler ve poliradikülönöropatiler gibi ciddi nörolojik bulgular HSP'nin nadir nörolojik komplikasyonlarıdır. Bu yazıda 17 yaşında nöbet ve bilinç bozukluğu olan HSP si olan bir olgu sunduk. Beyin manyetik rezonans görüntüleme serebral vaskülit ile uyumlu bulgular gösterildi. Tanıdan sonra pulse steroid tedavisi verildi ve hızlı bir klinik yanıt elde edildi. Sonuç olarak serebral vaskülitin nörolojik semptom ve bulguları olan tüm HSPli hastalarda düşünülmesi gerektiğini hatırlatmak istedik. *Türk Fiz Tıp Rehab Derg 2012;58:329-31.*

Anahtar Kelimeler: Henoch-Schönlein purpura; serebral vasculit; manyetik rezonans görüntüleme

Introduction

Henoch-Schönlein purpura (HSP) is a small vessel vasculitis that affects predominantly the skin, joints, gastrointestinal tract, and the kidneys (1,2). HSP is seen most frequently in early childhood, although it can occur at any age (3,4). HSP is a self-limited disease in most cases. The great majority of patients recover completely from HSP (3-5). However, it may be associated with several complications. Neurological symptoms and signs

occur in a significant proportion of cases with HSP (2,6-11). Although headache and behavioral changes are seen in many patients, severe neurological manifestations such as seizures, intracerebral hematoma, hemiplegia, and encephalopathy are the rare but potentially serious complications (1,2,7,8,10,12,13). The neurologic manifestations in HSP are usually observed in younger children. Neurologic symptoms usually develop within less than 2 weeks after the onset of HSP (1). Recent reports suggest that the magnetic resonance imaging (MRI) of the brain is the modality of

choice for the diagnosis of cerebral vasculitis because of its high sensitivity (1,2,7,11,12,14). In this report, we described a 17-year-old girl with HSP who presented with seizures and disturbance of consciousness. The MRI findings were consistent with cerebral vasculitis.

Case Report

A 17-year-old girl was admitted to the emergency department of our hospital with seizures, ataxia, confusion and disturbance of consciousness. One week prior to admission, she had developed sudden onset of purpuric rash on her lower extremities, arthralgia, and abdominal pain following upper respiratory tract infection. She presented with sudden onset of seizures, vertigo, ataxic gait, confusion and disturbance of consciousness a day before admission. Her medical and family history were nonspecific. There was no history of medication and immunization. The patient was transferred to the neuro-intensive care unit (NICU) of the neurology department.

Physical examination showed a body temperature of 36.8^o C, blood pressure of 115/70 mmHg and pulse rate of 90/min. The heart sounds were normal and lungs were clear in auscultation. Abdominal palpation was painful but without rebound phenomenon. Positive physical examination findings included multiple erythematous purpura on her lower extremities. The wrists and ankles were swollen and tender. Her lower extremities were edematous.

Initial laboratory findings were as follows: hemoglobin - 11,4 g/dl, hematocrit-40%, white blood cell count (WBC)-12000/mm³, platelet count-310000/mm³, erythrocyte sedimentation rate - 60 mm/h, C-reactive protein - 1,2 g/dl, blood urea - 55 mg/dl, serum creatinine - 1,7 mg/dl. Serum lipid profile, serum C3 and C4 complement levels, electrolytes, liver function tests, coagulation parameters, serum immunoglobulins were determined to be within normal ranges.

Serological tests were negative for antinuclear antibody (ANA), anti-dsDNA, antineutrophil cytoplasmic antibody (ANCA), and hepatitis markers. The anti-streptolysin O antibody titer was 800 Todd unit but throat culture was negative. The occult blood in the stool was found to be negative. Chest X-ray and electrocardiogram were normal. At the time of seizures, serum electrolytes, blood glucose, and calcium levels were found to be within normal limits. Urinalysis determined hematuria and proteinuria with a follow up of 24-h urinary excretion. Proteinuria was 50 mg/kg/day and creatinine clearance rate was 75 ml/min. Examination of the cranial nerves and fundus was normal. Based on these clinical and laboratory findings, the patient was diagnosed as having HSP with skin, renal, joints and neurologic involvements. The skin biopsy showed leukocytoclastic vasculitis. Brain computed tomography was negative. The magnetic resonance imaging (MRI) of the brain demonstrated multiple small areas of high signal intensity in the white matter, interpreted as ischemic lesion of vasculitis (Figure 1).

She was treated with phenytoin for seizures. Initially,

intravenous methylprednisolone therapy (1000 mg/day, three consecutive days) was given for cerebral vasculitis. Later, oral prednisolone was given over the following 3 months. However, the clinical symptoms of HSP improved dramatically after pulse methylprednisolone therapy, the neurological findings were successfully treated with long-term proceeded (3 months) oral prednisolone. A follow-up MRI study was performed 3 months later which demonstrated improvement in cerebral lesions.

Discussion

HSP is the most common childhood vasculitis, and is characterized by the classic tetrad of nonthrombocytopenic palpable purpura, arthritis or arthralgias, gastrointestinal and renal involvement (15). Its etiology is unclear but is associated with infections (bacterial, viral, parasitic), medications, vaccination, tumors (non-small cell lung cancer, prostate cancer and hematological malignancies), alpha-1-antitrypsin deficiency, and familial Mediterranean fever. The peak age incidence is 4-6 years and 90% of HSP cases occur before the age of 10 years (16-18). HSP is often preceded by an upper respiratory tract infection, with Group A beta-hemolytic streptococcus responsible for up to 50% of the occurrences (3).

Neurological involvement in HSP has rarely been reported (1-3,6,9,10,12,13). Belman et al. (10) have reported that headache and mental status changes are the most frequent neurological complications of HSP, followed by seizures, focal neurologic deficits, mononeuropathies and polyradiculoneuropathies. However, severe neurological manifestations such as seizures, intracerebral hematoma, hemiplegia, coma, blindness and encephalopathy are rare but potentially serious complications (1-13). It has been reported that hypertensive encephalopathy, uremic encephalopathy, electrolyte imbalance, and cerebral vasculitis might contribute to the neurologic manifestations (9,10,13). The diagnosis of cerebral vasculitis may be difficult because the underlying neurologic abnormalities in HSP may have a multifactorial etiology (13).

HSP is a clinical diagnosis but when the presentation is atypical, tissue biopsy may be helpful (19). New criteria are

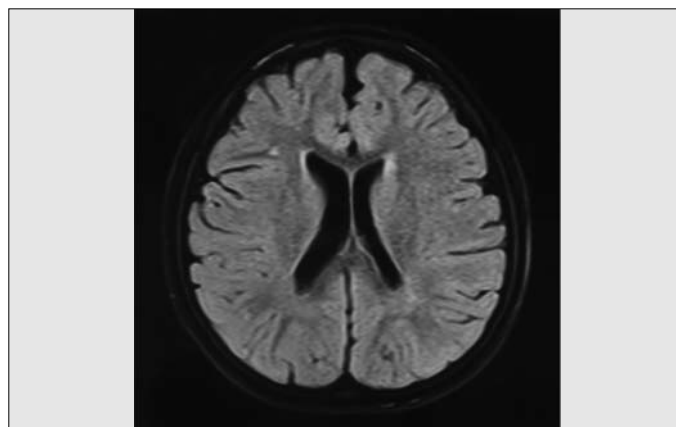


Figure 1. Flair axial images demonstrates multiple small areas of high signal intensity in the white matter.

proposed by the European League Against Rheumatism (EuLAR) and Pediatric Rheumatology Society (PReS) for HSP diagnosis (11,17). Our patient was diagnosed with HSP according to the EuLAR/PReS criteria. The diagnostic criteria of EuLAR/PReS consensus report for HSP are the following: palpable purpura (mandatory criterion) in the presence of at least one of the following four features: diffuse abdominal pain; any biopsy showing predominant IgA deposition; arthritis (acute, any joint) or arthralgia; renal involvement (any hematuria and/or proteinuria) (11,17,19). The skin biopsy showed a leukocytoclastic vasculitis in our patient. She had developed upper respiratory tract infection 15 day before HSP onset. There was no history of drug exposure and immunization. However, her blood pressure and electrolytes were within the normal range, she had neurological symptoms such as seizures, confusion and disturbance of consciousness. Therefore, we suspected cerebral vasculitis in our patient with severe neurological manifestations. The magnetic resonance imaging of the brain is the modality of choice for the diagnosis and evaluation of cerebral vasculitis in HSP patients (1,2,7,11,12,14). However, MRI of the brain was performed immediately and demonstrated the findings consistent with cerebral vasculitis.

Mild disease resolves spontaneously, and symptomatic treatment alone is sufficient. Systemic steroids are recommended for moderate to severe HSP (11). In most cases, it is a self-limiting disorder and tends to resolve within 1 month of presentation (3-5). According to the previous literature on cerebral vasculitis in HSP, pulse methylprednisolone therapy provides effective relief of cerebral vasculitis (2,7,11,12,14). However, intravenous methylprednisolone followed by oral prednisolone was given for cerebral vasculitis in our patient. The clinical symptoms of HSP dramatically improved after pulse steroid therapy. The neurological findings were successfully treated with long-term oral prednisolone. A follow-up MRI was performed 3 months later, demonstrated the improvement of cerebral lesions. The prognosis depends on the age of onset, extent of renal involvement and its course, extent of skin involvement, particularly above the waist line, immunoglobulin imbalance and neurological involvement (16,20).

In conclusion, early diagnosis of multiorgan involvement and appropriate therapy can mitigate the disease. We suggest that cerebral vasculitis should be considered in all HSP patients with neurological symptoms and signs.

Conflict of Interest:

Authors reported no conflicts of interest.

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