

Full Length Research Paper

An unusual case of Henoch-Schönlein purpura associated with varicella zoster infection

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Henoch-Schönlein purpura (HSP) is one of the most common vasculitis of childhood which is characterized by nonthrombocytopenic palpable purpura, arthritis, renal and gastrointestinal system (GIS) involvement. The etiology of HSP is mainly unknown and antecedent upper respiratory tract infection, usually viral and Streptococcal origin, drugs, cold, insect bite or some foods are the known triggering factors. HSP is rarely triggered by varicella zoster infection. In this study, we present a case with HSP following varicella infection who was admitted to the hospital because of edema and erythema in the right ankle, palpable purpura in the lower extremities and crusted lesions of varicella in his back. As a result of the case, we believe that consideration of varicella zoster as a causative agent for HSP necessitates the vaccination against it which will prevent the development of many complications of varicella zoster infection including HSP.

Key words: Henoch-Schönlein purpura, varicella zoster infection, nonthrombocytopenic purpura.

INTRODUCTION

Henoch-Schönlein purpura (HSP) is one of the most common vasculitis of childhood (Kalman et al., 2005). Its incidence is approximately 10 per 100,000 children per year and is slightly more common among boys (60%) than girls (40%) (Kalman et al., 2005; Sohagia et al., 2010). It is characterized by nonthrombocytopenic palpable purpura, arthritis, renal and gastrointestinal system (GIS) involvement (Kalman et al., 2005; Sohagia et al., 2010; Kalyoncu et al., 2003). It is a vasculitis of small vessels manifested by a characteristic rash. The course of HSP is typically a benign one, but may be accompanied by varying degrees of abdominal pain, arthritis or arthralgia, gastrointestinal bleeding, and nephritis. The various manifestations of HSP may be present at any stage during the illness and mimic other

disease processes (Sohagia et al., 2010; Reamy et al., 2009).

Most cases are self-limiting and require no treatment apart from symptomatic relief, but recurrence of symptoms occurs in about 33% of the cases (Sohagia et al., 2010). It seems that relapse often occurs between 2 weeks and 18 months after initial resolution of symptoms and children with kidney involvement are more likely to have recurrences. In some patients, nephritis occurs due to immunoglobulin A (IgA) deposition in the renal mesangium (Kalyoncu et al., 2003; Leonardi et al., 1992; Mayer et al., 2005). More serious complications such as the involvement of central nervous system, renal failure, and intussusception may also occur (Sohagia et al., 2010). There is no consensus on the optimal treatment in the case of significant renal or other organs involvement where treatment may have a significant impact on the long-term outcome (Sohagia et al., 2010; Reamy et al., 2009).

The etiology of HSP is mainly unknown and antecedent

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Figure 1. The lesion of varicella zoster infection in the dorsum.

upper respiratory tract infection, usually viral, has been reported and streptococcal infection has been emphasized as an important triggering factor. It is also shown to be triggered by drugs, cold, insect bite or some foods (Kalman et al., 2005; Sohagia et al., 2010; Kalyoncu et al., 2003; Reamy et al., 2009). On the other hand, it is known that IgA plays an important role in the pathogenesis of the disease. The diagnosis is based on the palpable purpura in the presence of one of either diffuse abdominal pain, a biopsy showing predominant IgA deposition, arthritis, or arthralgia and/or renal involvement (hematuria and/or proteinuria) (Kalyoncu et al., 2003; Leonardi et al., 1992; Mayer et al., 2005). HSP is rarely triggered by varicella zoster infection (Kalman et al., 2005; Sohagia et al., 2010; Kalyoncu et al., 2003; Kalyoncu et al., 2003; Leonardi et al., 1992). In this study, we presented a case with HSP following varicella infection.

DESCRIPTION OF CASE

A 5 year-old-male child was presented with rash and pain in both legs with swollen ankles. He was diagnosed to have varicella zoster infection 10 days ago. His physical examination revealed edema and erythema in the right ankle, palpable purpura in the lower extremities and crusted lesions of varicella in his back (Figures 1 and 2). Review of the other systems was unremarkable. Laboratory tests showed white blood cell (WBC), 5800 mm³; hemoglobin (Hb), 12.7 g/dl; platelets, 268.000/mm³; erythrocyte sedimentation rate, 63 mm/h. His varicella zoster IgM was positive and his tests were otherwise normal. His throat and urine cultures were normal. The

patient was diagnosed with HSP as per European League Against Rheumatism (EuLAR) criteria and admitted to the hospital. According to EuLAR criteria (2006) and Sohagia et al. (2010), the mandatory criterion (Palpable purpura) together with one of the following criteria (Arthritis/arthralgias) was present. He was treated with oral Ibuprofen (10 mg/kg) for 3 days. He was discharged from the hospital with resolution of right ankle pain and swelling. Then, he was re-scheduled for a follow-up program in the out-patient clinics. During a follow-up period of 10 months, the child's physical examination did not reveal any abnormality and his laboratory tests including urinalysis and occult blood in stool were negative.

RESULTS AND DISCUSSION

HSP is a self-limited, systemic, nongranulomatous, autoimmune complex, small vessel vasculitis, with multiorgan involvement (Sohagia et al., 2010; Reamy et al., 2009). Its etiology is unclear but may be associated with bacterial, viral and parasitic infections. Antigen and antibody complexes, mostly IgA, form as a result of bacterial and viral infections and these antigen antibody complexes deposit in the small vessel walls and activate the alternate complement pathway which leads to neutrophil accumulation resulting in inflammation and vasculitis without a granulomatous reaction. This can involve multiple systems including skin, gastrointestinal tract, kidney, and joints but it can involve any organ system. Vasculitis causes extravasation of blood and its components into the interstitial spaces resulting in edema and hemorrhage (Sohagia et al., 2010; Reamy et al., 2009). In our case with positive varicella zoster IgM,



Figure 2. Palpable purpura Of Henoch-Schölein disease in the lower extremities.

varicella zoster infection might possibly have played a role in initiating the HSP cascade. Cutaneous manifestations include nonthrombocytopenic rash which evolves from erythematous to urticarial and macular wheels to nonblanching palpable purpura with petechiae and ecchymoses. Palpable purpura is seen in 50% of the cases as the presenting sign. Classical HSP is symmetrical in distribution involving dependent areas such as the lower extremities and buttocks but it can also be seen in the upper extremities (Sohagia et al., 2010; Reamy et al., 2009). Our case presented with classical HSP rash symmetrically was distributed in the lower extremities (Figure 1).

On histopathology leukocytoclastic vasculitis, characterized by neutrophilic infiltration and prominent nuclear fragmentation, involving the upper and middle layers of the dermis with IgA deposition on immunofluorescence, is seen. Consequently, angioedema (nonpitting edema) can be seen in the scalp, back, and extremities (Sohagia et al., 2010; Reamy et al., 2009; Mayer et al., 2005). Our case showed edema as well as erythema in the right ankle.

As a result of multiorgan involvement, varying degrees of abdominal pain, gastrointestinal bleeding and nephritis can be seen in patients with HSP (Sohagia et al., 2010; Reamy et al., 2009; Kalyoncu et al., 2003; Leonardi et al., 1992). But our case did not show any symptom or laboratory finding relevant to these complications. Diagnosis of HSP was made by EuLAR criteria as the

mandatory criterion together with one of the following criteria were present (Sohagia et al., 2010).

Treatment of HSP usually involves symptomatic treatment which will be sufficient for symptoms such as rash and arthritis. Acetaminophen and nonsteroidal anti-inflammatory drugs can be used. Aspirin should be avoided in children. Oral steroids are indicated in patients with severe rash, edema, severe abdominal pain (without nausea, vomiting), renal, scrotal, and testicular involvement (Sohagia et al., 2010; Reamy et al., 2009). As our case was free of any of these severe symptoms, we managed the treatment with nonsteroidal anti-inflammatory drugs.

In the pathophysiology of HSP antigen and antibody complexes, mostly IgA, form as a result of bacterial and viral infections, vaccinations, drugs, and autoimmune mechanisms (Sohagia et al., 2010; Kalyoncu et al., 2003; Kalyoncu et al., 2003). Although there is not enough information about the prevention of the development of HSP following varicella zoster infection by varicella zoster immunoglobulin (VZIG), we would like to debate this issue concerning our case. In light of the information about the pathophysiology of HSP developed after varicella zoster infection, we assume that VZIG would only be beneficial in immunodeficient patients. Besides, VZIG is only effective when it is given within 72 h following infection.

Our case of HSP which was presented 10 days after a varicella zoster infection is a rare case proving the evidence that varicella zoster infection may be an

etiologic agent for HSP. In the literature, there are very rare cases of HSP presented after varicella zoster infection. So, we believe that our case is an exemplary one for the literature. The prevention of varicella zoster infection is possible with vaccination.

Conclusion

We believe that preventive medicine facilities should be carefully managed for all kinds of preventable infections including varicella zoster. The vaccination against varicella zoster will prevent not only the development of the infection but also the development of many complications after varicella zoster infection by immunological mechanisms, including HSP. For this purpose, we suggest that varicella vaccination should take place in routine schedules all over the world including developing and underdeveloped countries.

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