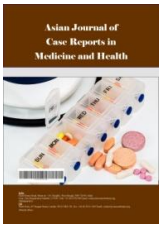

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Corticosteroid Treatment for Recurrent Henoch-Schönlein Purpura with Gastrointestinal Manifestation: A Case Report

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Authors' contributions

This work was carried out in collaboration between both authors. Author EM designed the study, wrote the first draft of manuscript, managed the analyses of the study and managed the literature searches. Author FNT designed the study and managed the analyses of the study. Both authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Aims: Henoch-Schönlein Purpura (HSP) is the most common vasculitis during childhood yet sometimes is misdiagnosed. Gastrointestinal symptom in HSP is a frequent finding which causes patient to seek medical care. Recurrent episode of HSP occurs in 2.7–30% cases, most of which has predisposing factors. HSP is usually a self-limiting disease which is managed with supportive treatment. Limited evidence has been reported about the use of corticosteroid for HSP.

Presentation of Case: A 10-year-old girl came with chief complaints of abdominal pain, nausea and vomiting, and palpable purpura in lower extremities a day prior to admission, preceded by upper respiratory tract infection. Patient had history of similar symptoms before, she was diagnosed with recurrent HSP with skin and gastrointestinal manifestations. She was treated with corticosteroid for two weeks and showed clinical improvement during follow-up.

Discussion: Recurrent episode of HSP occurs more frequently in patients with predisposing factors. Although there is no specific recommendation about the use of corticosteroid for HSP, several studies reported benefits of corticosteroid for patients with organ involvement such as gastrointestinal to relieve the pain. We considered giving short-term corticosteroid and observed clinical improvement in our patient.

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Conclusion: Identifying predisposing factors for HSP in each patient may help to prevent the recurrence. Despite limited evidence regarding the use of corticosteroid, short-term use of corticosteroid could be considered in HSP with gastrointestinal involvements to relieve the symptoms.

Keywords: Henoch-Schönlein Purpura; gastrointestinal; corticosteroid; recurrent; pediatrics; case report.

1. INTRODUCTION

Henoch-Schönlein Purpura (HSP), also called IgA vasculitis, is a leukocytoclastic vasculitis with predominant IgA deposition in small vessel, mostly in the skin, joint, gastrointestinal tract, and kidney [1]. It is characterized by clinical triad of palpable purpura (without thrombocytopenia), abdominal pain, and arthritis, or tetrad with renal involvement. HSP is the most common vasculitis in childhood. The incidence of HSP in children is approximately 14–20 per 100,000 person-years. About 90% of HSP cases occur during childhood (3–10 age group) with peak incidence at the age of 6 [2]. Considered as a rare disease, HSP is sometimes misdiagnosed as other entities, for example insect bite. HSP in adult population is associated with more severe features and chronic complication, mainly kidney disease [1,2].

The pathogenesis of HSP is still yet to be determined. However, HSP is usually preceded by episode of upper respiratory tract infection (URTI), mostly occurs in seasonal pattern during winter and spring [1,3,4]. Infection by certain microorganisms, such as *Streptococcus β-hemolyticus group A, Staphylococcus aureus, Mycoplasma,* and *Adenovirus,* is considered as potential trigger. Furthermore, HSP is characterized by deposition of IgA immune complex and C3 in small vessel walls as well as polymorphonuclear neutrophil infiltration. Deposition of immune complex further triggers inflammatory mediators in target organ, especially skin, joint, gastrointestinal, and kidney. Certain alleles, such as HLA-B34 and HLA-DRB1*01, are associated with HSP nephritis. Hence HSP is regarded as an immune-mediated disease associated with genetic and environmental factors [1].

In this report, we aim to present a case of recurrent HSP with gastrointestinal manifestation treated with short-term corticosteroid.

2. CASE PRESENTATION

A 10-year-old girl came to the hospital with chief complaints of abdominal pain, nausea and vomiting since a day prior to admission. Abdominal pain was diffuse, colicky, mostly happened postprandial. Patient had watery stools, no mucus or blood. Alongside with gastrointestinal complaints, patient had painless and nonpruritic palpable purpura in both lower extremities. Patient had no history of joint swelling or pain and urinary problem. She had history of upper respiratory tract infection four days prior to admission. Patient and caregiver denied any history of allergy, recent vaccination, and drug consumption. Patient had similar symptoms three months ago, she had been diagnosed with Henoch-Schönlein Purpura and given steroid therapy but did not adhere for routine control.

Vital signs were within normal limits, patient had normotension and afebrile. Physical examination showed diffuse abdominal pain and palpable purpura in lower extremities (Fig. 1). Her body weight was 22 kilograms. Laboratory results showed leukocytosis (22,150) and thrombocytosis (523,000) with normal creatinine level (0.59 mg/dL). Urinalysis test showed leukocyte 10-15 per high power field, ketone 1+, no proteinuria or hematuria. Abdominal ultrasonography was unremarkable.

Patient was diagnosed with recurrent Henoch-Schönlein Purpura with skin and gastrointestinal manifestations. She was given steroid therapy with intravenous methylprednisolone injection 2x20 mg, Ranitidine injection 2x25 mg, and oral Sucralfate 3x5 ml. Patient had clinical improvement and then discharged after three days of hospitalization with consumption of oral methylprednisolone (0-16 mg-24 mg). After five days, patient came to the outpatient clinic with no gastrointestinal symptoms and the purpura had been faded away (Fig. 2). Patient continued to take steroid therapy for a week.

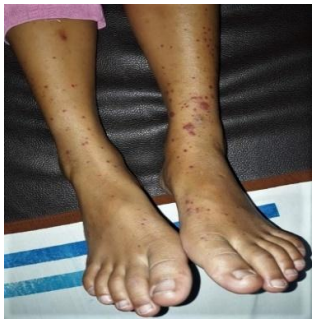

Fig. 1. Palpable purpura in lower extremities

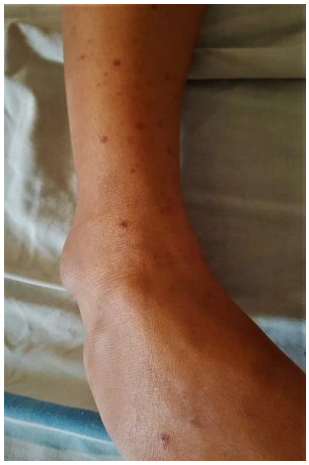

Fig. 2. Faded purpura seen after 8 days of follow-up and treatment

3. DISCUSSION

Diagnosis of HSP is usually made clinically using diagnostic criteria for its typical presentations. First proposed criteria were made by The American College of Rheumatology (ACR) in 1990 (sensitivity 87.1%, specificity 87.7%). More recent diagnostic criteria by European League Against Rheumatism/Paediatric Rheumatology International Trials Organization/Paediatric Rheumatology European Society (EULAR/ PRINTO/PRES) with better sensitivity and specificity (100% and 87% respectively) was developed in 2010 for pediatric population [4]. Gastrointestinal involvement is common, occurs in about 80% cases of HSP in children, with clinical manifestations such as abdominal pain, nausea and vomiting, diarrhea, melena or hematochezia, and paralytic ileus [1,2]. In our case, the diagnosis was made clinically by the presence of typical palpable purpura not thrombocytopenic in predilection area of lower extremities and abdominal pain. More advanced procedure such as kidney or skin biopsy is usually performed in patients with uncertain diagnosis, atypical presentation, or more severe organ involvement.

Supporting examinations are sometimes needed to confirm the diagnosis or detect complication. For patient with gastrointestinal symptoms, abdominal ultrasound can be used to exclude other differential diagnosis and detect complication such as intussusception or bowel perforation [1]. In this case, the abdominal ultrasound detected no abnormality. Fecal occult blood test could be considered for our patient who had experienced 2 episodes of HSP with the same gastrointestinal manifestation, especially in resource-limited setting. Study by George et al confirmed that a simple yet inexpensive biomarker for inflammatory condition, such as neutrophil-to-lymphocyte ratio (NLR), is significantly higher in adult patients with GI involvements. NLR is also proven to be the strongest predictive value for systemic involvement in IgA vasculitis [5]. More sophisticated examination such as fecal

calprotectin which marks neutrophil influx in certain gastrointestinal inflammatory conditions might be performed in tertiary health care for several cases. Fecal calprotectin may be useful for predicting gastrointestinal involvement and disease severity in HSP [6].

Relapse or recurrence is defined as the appearance of cutaneous lesion or other systemic vasculitis manifestations in a previously diagnosed patient with HSP who has been asymptomatic for at least 2 weeks [3]. Recurrence mostly happens within 2–3 months after first episode of HSP and has similar manifestations with previous episode [7]. Recurrence rate of HSP is 2.7–30% [8], it might recur in one-third of affected children [9,10]. Several studies reported that infection is the most common predisposing factor for HSP. Study by Xu et al. reported relapse of HSP in 10.25% subjects with predisposing factors such as infection (39.68%), allergy (6.88%), injury/surgery (0.53%), while the rests had no clear predisposing factors [3]. In our case, history of URTI is considered as predisposing factor for recurrent HSP. During the first and second episode of HSP, patient experienced the same symptoms, which were palpable purpura and abdominal pain.

Lei et al. reported recurrent HSP occur more frequently in patients with renal involvement, were receiving steroid therapy for >10 days, and had allergic rhinitis [8]. Multivariate analysis by Calvo-Rio et al reported the best predictive factors for relapse were joint and gastrointestinal manifestations at HSP diagnosis [11]. In accordance with previous studies, we identified that our patient had higher risk for relapse regarding the gastrointestinal manifestations at time of diagnosis and history of steroid therapy for >10 days during previous episode of HSP.

Management for mild HSP is supportive treatments, include adequate hydration, nutrition, and analgesics. Acetaminophen is usually used for analgesic and antipyretic while NSAIDs is avoided, especially if there is gastrointestinal and/or renal involvement(s) [1]. Corticosteroid has been postulated to be effective for HSP treatment owing to its inhibitory effect on inflammatory process and its usefulness in treating other childhood vasculitides. However, corticosteroid use as universal treatment for HSP remains controversial [9,10]. In case of organ involvement (gastrointestinal or renal), corticosteroid therapy (oral prednisone 1–2 mg/kg body weight per day or intravenous methylprednisolone for severe case) for 1–2 weeks then tapering off should be considered [1,10]. Steroid may relieve symptoms, mainly abdominal pain, arthralgia, and purpura [10]. A systematic review suggests that early corticosteroid treatment is associated with increased odds of abdominal pain resolution within 24 hours [9], which is beneficial for the patient. However, steroid use for more than 10 days is possibly associated with recurrence of HSP [8], thus we decided to give short-term corticosteroid for this patient. Good response, marked by clinical improvement, was seen during hospitalization and follow-up in outpatient clinic. Until now, there is no specific recommendation or consensus regarding optimal duration and steroid use in HSP treatment [8]. In fact, the use of corticosteroid for nephritis prevention is not recommended [1].

Overall prognosis for HSP is favorable, the disease is usually benign and self-limiting. The symptoms usually resolve after 4 weeks. Nevertheless, 15–60% children have recurrent episode of HSP for one or more, usually with milder symptoms, within 4–6 months after diagnosis. Children with more severe clinical manifestations tend to have higher risk for relapse. Long-term prognosis depends on severity of the disease and organ involvement, mainly gastrointestinal and renal. Chronic kidney disease occurs in 1–2% HSP cases. About 5% of HSP nephritis progress into end-stage renal failure [1]. Long term follow-up is important, especially in patients with predisposing factors for relapse. Patients with second episode of HSP should be monitored at least 6 months, particularly to detect another potential episode of relapse [8]. Also, renal complication may occur in 1–2% patients with normal urinalysis at time of diagnosis within 6 months. Therefore, prolonged observation for blood pressure and urinalysis is needed [1].

4. CONCLUSION

HSP is the most common vasculitis in childhood. Gastrointestinal involvement is one of the most frequent presenting symptoms, therefore, clinicians should be more aware in considering HSP as differential diagnosis. Short-term use of corticosteroid could be considered in HSP with gastrointestinal involvements to relieve the symptoms, indubitably along with other supportive treatments. Identifying predisposing

factors for HSP in each patient may help to prevent the recurrence. As recurrence rate and possible long-term complication is quite high, follow up is important as a part of comprehensive management for HSP.

CONSENT

Parental written consent has been collected and preserved by the authors.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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