

Case Report

Henoch-Schonlein Purpura Successfully Treated with Dexamethasone: A Case Report of Six-year-old Female

Purushottam Adhikari

Department of Medicine, Gandaki Medical College, Nepal

Address for correspondence: Purushottam Adhikari, Department of Medicine, Gandaki Medical College, Nepal. E-mail: puru.ad12@gmail.com



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Abstract

Henoch-Schonlein Purpura (HSP) is one of the most common causes of small vessel vasculitis in children. A 6-year-old female presented with abdominal pain, swelling, and rashes over both the legs associated with multiple episodes of vomiting for around 10 days. Stool for the occult blood test was positive, but there was an absence of hematuria and albuminuria. The case was diagnosed as HSP and treated with dexamethasone for 10 days. The patient was finally discharged on the resolution of her symptoms. Early diagnosis and treatment favor the better outcome in cases without any renal complications.

Keywords: Henoch-Schonlein Purpura, Vasculitis, Steroids, Dexamethasone

Introduction

Henoch-Schonlein Purpura (HSP), also known as IgA vasculitis, is a small vessel vasculitis with IgA1-dominant immune deposits predominantly on capillaries, venules, or arterioles. It often involves skin and gastrointestinal system and may also cause arthritis.^[1] It is commonly seen in children and characterized by palpable purpura more commonly located in the dependent body parts such as lower extremities and buttocks, arthritis/arthralgia, and bowel angina along with hematuria/proteinuria.^[2] The various etiologies have been suggested such as varieties of pathogens, drugs, and environmental exposure among which Group A beta-hemolytic *Streptococcus* has been much studied.^[3]

The natural history of the disease has self-limiting course in most of the cases except that those of renal complications associated with it. Symptomatic treatment with non-steroidal anti-inflammatory drugs along with the steroids will have the joint and abdominal pain relief.^[4] Corticosteroids if given in the early course of disease can help to provide a better clinical outcome.^[5] The prognosis is good, with exception of renal involvement that may need the follow-up till 6 months or longer.^[6,7]

The exact prevalence of HSP in our settings has yet to be discovered, but various studies revealed it to be one of the important causes of childhood renal disease.^[8,9]

Case Report

The case is reported after taking informed written consent from the patient's mother. 6-year-old female presented with the complaints of pain in abdomen, swelling, and rashes over the lower limbs associated with multiple episodes of vomiting for 10 days. Pain in the abdomen was localized around the umbilicus and was sudden in onset, intermittent in nature, non-radiating, and alleviated on lying flat on the bed. She developed swelling in both legs subsequently a day after abdominal pain along with the appearance of rashes starting from feet and progressing to thigh and buttocks. She also developed three to four episodes of vomiting in last 5 days. The vomiting was non-projectile and non-blood stained.

On examination, the general condition of the patient was fair and vitals were stable. The abdomen was soft and non-tender. Mild bilateral

non-pitting edema was present over both the legs. There was the presence of non-tender, non-blanching, purpuric rashes over both lower limbs, and extending up to the buttocks.

Laboratory tests showed leukocytosis with WBC count of 12,000/mm³; neutrophils: 8800/mm³; lymphocyte: 3200/mm³; platelets: 4,22,000/mm³; hemoglobin: 14 g/dl; ESR: 12 mm/1st h; serum urea: 33 mg/dl; serum creatinine: 0.6 mg/dl; sodium: 139 mmol/l; potassium: 4.6 mmol/l; CRP: 0.92 mg/l; urinalysis: No hematuria or proteinuria, and stool analysis showed the presence of occult blood. The plain X-ray abdomen and ultrasound abdomen/pelvis revealed no abnormalities.

Diagnosis of HSP was made in accordance with the American College of Rheumatology and European League Against Rheumatism (EuLAR) and Pediatric Rheumatology Society (PREs) criteria.^[10,11]

She was treated with dexamethasone 0.14 mg/kg/dose 4 times a day intravenously for 5 days continuously and then tapered to 3 times a day for 3 days following by 2 times a day for next 2 days.

The recovery from purpura and bilateral swelling of the legs was observed after treatment in the 2nd and 3rd week [Figures 1-3].

Discussion

Henoch-Schonlein Purpura (HSP) was first described by William Heberden in 1801. Later, Schonlein recognized the association between purpura and arthritis, whereas Henoch reported a case that also included gastrointestinal symptoms along with the renal involvement.^[12] HSP is the most common vasculitis of the children. Half among all the cases occur before the age of five and males are affected twice as common as females.^[13]

The exact etiology and pathogenesis of HSP are yet to be determined. Seasonal variation has been related with a high prevalence rate in autumn and winter.^[2] However, the case described here was diagnosed in the spring season. It has also been proposed that various triggers such as bacterial and viral infections, vaccinations, drugs, and autoimmune mechanisms may result in the formation of an antigen and antibody complex and the deposition of such formed immune complex in the small vessels may activate the alternate complement pathway leading to neutrophil aggregation which results in inflammation and vasculitis.^[14] Among all, the preceding infection of β -hemolytic *Streptococcus* has been one of the most studied cases. The positive throat cultures, as well as increased titers of anti-streptolysin O, have been often found in many patients.^[3] The evidence of prior infection has not been recorded in the present case.

The patient generally presents with the classic tetrad of rashes, polyarthralgia, abdominal pain, and renal disease. The non-



Figure 1: Purpura and swelling of bilateral lower limbs in the 1st week



Figure 2: Progress of purpura in the 2nd week after the admission of patient in hospital



Figure 3: 3rd week before the discharge from hospital

blanching rashes clinically appear as a palpable purpura on the lower legs and arms.^[13] The joint involvement is generally characterized by the pain and swelling of the joints, most

Table 1: The American College of Rheumatology - 1990 criteria for the diagnosis of HSP^[10]

Criterion	Definition
Palpable purpura	Slightly raised “palpable” hemorrhagic skin lesions, not related to thrombocytopenia
Age < 20 at disease onset	Patient 20 years or younger at onset of first symptoms
Bowel angina	Diffuse abdominal pain, worse after meals, or the diagnosis of bowel ischemia, usually including bloody diarrhea
Wall granulocytes on biopsy	Histologic changes showing granulocytes in the walls of arterioles or venules

Table 2: EuLAR/PReS – 2006 criteria^[11]

Criteria	Definition
Mandatory criteria	Palpable purpura
Additional criteria	Diffuse abdominal pain
	Any biopsy showing predominant IgA deposition
	Arthritis or arthralgia
	Renal involvement (any hematuria and/or proteinuria)

affecting the knees and ankles. The abdominal pain followed by vomiting and intestinal bleeding is the dominant features involving the gastrointestinal system. Microscopic hematuria and albuminuria are the prominent renal findings.^[5,15]

Our case had the symptoms of rashes over both lower legs, pain in abdomen, and vomiting. Joint involvement was absent. Significant laboratory finding was occult blood test positive in stool. However, there were no signs of hematuria and albuminuria.

The diagnosis of HSP was made by American College of Rheumatology-1990 criteria [Table 1] and EuLAR/PReS - 2006 criteria [Table 2].

The patient is said to have HSP if at least two of the four above criteria is present. In our case, there was the presence of palpable purpura, age <20 at disease onset and bowel angina, and thus was diagnosed as HSP.

The patient is said to have HSP if mandatory criteria are present along with at least one of the additional criteria. Our case fulfilled the mandatory criteria along with diffuse abdominal pain as the additional criteria.

There is a predominantly spontaneous resolution of all symptoms except that of the renal disease in the majority of the cases. Steroids are more often used for the relief of abdominal pain, joint pain, and skin disease. Alternatively, methotrexate and dapsone have been quite effective (steroid-sparing agent) for the treatment of chronic abdominal pain and skin involvement.^[4] The role of the corticosteroids in preventing the long-term outcome of renal complications is controversial. A meta-analysis in the Medline database and the Cochrane

Controlled Trials Register based on a comprehensive review of the literature by Weiss *et al.* stated that early treatment with corticosteroid significantly reduces the odds of developing persistent renal disease along with surgical intervention and recurrence.^[5] In contrary, other trials and updates in the literature have mentioned that there is no long-term renal protective outcome on the early treatment with prednisolone.^[16,17]

In general, prednisolone is the commonly used steroid for the treatment of HSP. Although in our case, we used dexamethasone, there is no evidence in literature to prove the superiority of one over another. Several cases and studies have been reported resulting in better outcomes on treatment with dexamethasone.^[18,19]

The renal involvement has a high morbidity and mortality; otherwise, the disease has better prognosis.^[7] A systematic review by Narchi stated that even if urinalysis is normal at the presentation, follow-up urine testing should be continued for at least 6 months as 97% children will develop abnormal urine findings by that time.^[6]

Rarely, few cases of complicated HSP as intussusceptions, gastrointestinal bleeding and with cardiac involvement have been reported.^[18,20,21]

Conclusion

HSP being one of the most common vasculitides of the children and its classic presentation of palpable purpura, arthritis, abdominal involvement, and renal features makes the diagnosis quite easier. Early initiation of treatment with steroids will help in symptomatic relief and bring a positive outcome. The renal disease may need long-term follow-up; otherwise, the diseases have favorable prognosis.

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