

## Case Report

# Henoch-Schonlein purpura in child: a case report and review of the literature

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### ABSTRACT

Henoch-Schonlein purpura (HSP) is the most common vasculitis of childhood and is characterized by leukocytoclastic vasculitis and immunoglobulin A deposition in the small vessels in the skin, joints, gastrointestinal tract and kidney. We report a 6 year old female child presented with rashes over both legs and buttocks along with joint pain and abdominal pain and multiple episodes of vomiting. Patient was diagnosed with HSP and treated with oral prednisolone followed by methylprednisolone and other supportive care.

**Keywords:** Henoch-Schonlein purpura, Leukocytoclastic vasculitis, Immunoglobulin A, Methylprednisolone

### INTRODUCTION

Henoch-Schonlein purpura (HSP), also referred to as immunoglobulin A (IgA) vasculitis, is a small vessel vasculitis that affects children of 3-10 year old age group. Incidence of HSP is estimated at 14-20 per 100,000 children per year and it affects males more than females with a 1.2-1.8:1 male/female ratio.<sup>1</sup>

It usually follows an upper respiratory infection caused by group A beta-hemolytic streptococcus, *Staphylococcus Aureus*, or *Mycoplasma*. Skin biopsies demonstrate leukocytoclastic vasculitis involving dermal capillaries and venules. It often involves skin and gastrointestinal system and may also cause arthralgia/arthritis. It is characterized by palpable purpura predominantly in gravity dependent areas such as lower limbs and extensor aspects of upper extremities or on pressure points such as buttocks. Gastrointestinal (GI) manifestations may include abdominal pain, vomiting and diarrhea. Renal involvement may be seen in 30% of cases in the form of hematuria, proteinuria and hypertension.

The disease is usually self-limiting and treatment is mainly supportive. Corticosteroids may be used to treat HSP with significant GI or renal involvement. Prognosis for childhood HSP is excellent with most children experiencing an acute, self-limited course lasting on average 4 weeks.

### CASE REPORT

We report a 6 year old female child presented with complaints of rash over both lower extremities involving the buttocks, joint pain, pain in abdomen and multiple episodes of vomiting for 1 day. Rash started from both lower limbs involving the buttocks and later progressing towards the extensor aspect of both upper extremities and was associated with swelling of face.

Patient also presented with joint pain involving both knees not associated with swelling around the joints. Pain in abdomen was sudden, periumbilical, intermittent and not radiating elsewhere. Child developed 8-10 episodes of vomiting, non-projectile, non-bilious, with 1-2 episodes being blood stained containing streaks of blood.

There was no history of fever or any preceding upper respiratory illness in the child.

On examination, general condition of the patient was fair and vitals were stable. Abdomen was soft and non-tender. There was presence of non-tender, non-blanching purpuric rash over both lower extremities (Figure 1a and b) involving buttocks (Figure 2) and on the extensor aspect of both upper extremities (Figure 3).

Lab findings were suggestive of Anemia with Hb of 9.5 gm/dl, normal WBC count and thrombocytosis with Platelet count of 5.6 lakhs. Serum electrolytes were within normal limits. Urine routine was normal and there was no evidence of any hematuria or proteinuria.

Renal function tests were normal and stool routine was suggestive of occult blood in stool.

USG abdomen was suggestive of non-necrotic sub-centric abdominal lymphadenopathy and minimal ascites.

Diagnosis of HSP was made as per the European League against Rheumatism (EuLaR) or Pediatric Rheumatology European Society (PRES) criteria.<sup>2,3</sup>

Child was treated with oral prednisolone for 5 days, following which child developed subcutaneous edema (Figure 4) and dyselectrolytemia but child finally responded to Inj Methylprednisolone @2 mg/kg/day which was later tapered and omitted successfully. Child also received other supportive care in the form of anti-emetics, antacids, analgesics and fluids.

Our case did not have any renal involvement manifesting in the form of hematuria, hypertension or frank nephritis. Child also did not develop any neurological manifestations.

Patient was advised follow up in the OPD with serial urine analysis and BP monitoring.



**Figure 1: (a) and (b) palpable purpura in lower extremities.**



**Figure 2: Palpable purpura on buttocks.**



**Figure 3: Involvement of extensor aspect of forearm.**



**Figure 4: Upper extremity subcutaneous edema.**

## DISCUSSION

HSP was first described by William Heberden in 1801. Later Schönlein recognized the association between purpura and arthritis while a case reported by Henoch also included gastrointestinal symptoms along with the renal involvement.<sup>4</sup> HSP is the most common vasculitis of the children. 50% cases occur before the age of five and males are affected almost twice as common as females.<sup>5</sup>

The exact etiology and pathogenesis of HSP is yet to be determined. Seasonal variation depicts high prevalence rate in winter and spring and is unusual in summer months. It is also suggested that various triggers like 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.<sup>6</sup> Among all, the preceding infection of  $\beta$ -haemolytic Streptococcus is the most commonly found pattern. Positive throat cultures as well as increased titres of anti-streptolysin O have been often found in many patients.<sup>7</sup> The evidence of prior infection has not been recorded in the present case.

The patient generally presents with the classic tetrad of rashes, poly-arthritis, abdominal pain, and renal disease. Hallmark of HSP is the non-blanching rash which clinically appears as a palpable purpura on the lower legs and arms.<sup>5</sup> Musculoskeletal involvement is generally characterized by the pain and swelling of the joints, with a predilection for large joints such as the knees and ankles. GI manifestations include abdominal pain followed by vomiting and intestinal bleeding. Microscopic hematuria and albuminuria are the prominent renal findings.<sup>8</sup>

Our case had the symptoms of rashes over both lower legs, pain in abdomen and vomiting and pain in both knees indicating GIT and joint involvement. Significant laboratory finding was occult blood test positive in stool. However, there were no signs of hematuria and albuminuria so no renal involvement. The diagnosis of HSP was made by EuLAR or PRES criteria (Table 1).

**Table 1: EuLAR/PRES criteria.<sup>3</sup>**

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

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 and arthralgia 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.<sup>9</sup> The role of the corticosteroids in preventing the long term outcome of

renal complications is controversial. Generally prednisolone is the commonly used steroid for the treatment of HSP. The renal involvement has a high morbidity and mortality, otherwise the disease has an excellent prognosis. A systematic review by Narchi et al stated that even if urinalysis is normal at the presentation, patient should undergo follow up urine testing for at least six months as 97% children will develop abnormal urine findings by that time.<sup>10</sup>

Rarely few cases of complicated HSP as intussusceptions, gastrointestinal bleeding and with cardiac involvement have been reported.<sup>11-13</sup>

**CONCLUSION**

HSP being one of the most common vasculitis 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 has favourable prognosis.

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**REFERENCES**

- Sivaraman V, Fels EC, Stacy P. Ardoin, NELSON Textbook of Pediatrics, 21st Edition. Chapter 192.1. 2019;1.
- Mills JA, Michel BA, Bloch DA, Calabrese LH, Hunder GG, Arend WP, et al. The American College of Rheumatology 1990 criteria for the classification of Henoch-Schonlein purpura. Arthritis Rheum. 1990;33(8):1114-21.
- Ozen S, Ruperto N, Dillon MJ, Bagga A, Barron K, Davin JC, et al. EULAR/PRES endorsed consensus criteria for the classification of childhood vasculitides. Ann Rheum Dis. 2006;65(7):936-41.
- Ballinger S. Henoch-Schonlein purpura. Curr Opin Rheumatol. 2003;15(5):591-4.
- Roberts PF, Waller TA, Brinker TM, Riffe IZ, Sayre JW, Bratton RL. Henoch-Schonlein purpura: a review article. Southern Med J. 2007;100(8):821-4.
- Sohagia AB, Gunturu SG, Tong TR, Hertan HI. Henoch-schonlein purpura-a case report and review of the literature. Gastroenterol Res Pract. 2010;597648.
- Saulsbury FT. Clinical update: Henoch-Schonlein purpura. Lancet. 2007;369(9566):976-8.
- Chen O, Zhu XB, Ren P, Wang YB, Sun RP, Wei DE. Henoch Schonlein Purpura in children: clinical analysis of 120 cases. Afr Health Sci. 2013;13(1):94-9.

9. Tizard EJ, Hamilton-Ayres MJ. Henoch Schonlein purpura. *Arch Dis Childhood Educ Practice Ed.* 2008;93(1):1-8.
10. Narchi H. Risk of long term renal impairment and duration of follow up recommended for Henoch-Schönlein purpura with normal or minimal urinary findings: a systematic review. *Arch Dis Childhood.* 2005;90(9):916-20.
11. Seo MK, Hong J, Yim HE, Pai KS. A Patient with Henoch-Schönlein Purpura with Intussusception and intractable Nephritis. *Childhood Kidney Dis.* 2016;20(2):92-6.
12. Yamazaki T, Akimoto T, Iwazu Y, Sugase T, Takeshima E, Numata A, et al. Henoch-Schonlein purpura complicated with severe gastrointestinal bleeding. *CEN Case Rep.* 2015;4(1):106-11.
13. Lutz HH, Ackermann T, Krombach GA, Grone HJ, Rauen T, Floege J, et al. Henoch-Schonlein purpura complicated by cardiac involvement: case report and review of the literature. *Am J Kidney Dis.* 2009;54(5):9-15.

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