International Journal of Research in Dermatology
Nimra et al. Int J Res Dermatol. 2025 Jul;11(4):340-343
http://www.ijord.com

DOI: https://dx.doi.org/10.18203/issn.2455-4529.IntJResDermatol20252075

Case Report

The clinical spectrum of Henoch-Schönlein purpura: insights from two distinct presentations

Nimra*, G. Anantha Lakshmi, K. B. Harshitha, Tanzeel Fathima

Sri Venkateshwara College of Pharmacy, Osmania University, Hyderabad, Telangana, India

Received: 03 May 2025
Revised: 03 June 2025
Accepted: 13 June 2025

***Correspondence:**
Dr. Nimra,
E-mail: nimrajaf@gmail.com

ABSTRACT

Henoch-Schönlein purpura (HSP), or IgA vasculitis, is an autoimmune small-vessel vasculitis caused by IgA immune complex deposition, primarily affecting children but rarely seen in adults. We present two contrasting cases to highlight its variable presentation and diagnostic challenges. The first case is a 44-year-old woman with recurrent purpuric rash, arthralgia, elevated CRP and ESR, and positive anti-RNP and anti-Sm antibodies, suggesting an autoimmune disorder. However, clinical features and biopsy confirmed IgA vasculitis. She was managed with corticosteroids, topical agents, and supportive care, emphasizing the need to distinguish HSP from other autoimmune diseases. The second case involves a 13-year-old girl with a known history of HSP who presented with hematemesis and melena. Investigations revealed marked leukocytosis, elevated CRP, and gastrointestinal involvement confirmed through endoscopy and biopsy, showing erosive gastritis and pancolitis consistent with GI vasculitis. She was treated with corticosteroid pulse therapy and cyclophosphamide. These cases demonstrate the spectrum of HSP in both adults and children and highlight the importance of correlating clinical symptoms with laboratory and imaging findings for timely and accurate diagnosis. They also reinforce the role of immunosuppressants in managing severe or refractory cases.

Keywords: Henoch-Schönlein purpura, Erythematous purpuric rash, GI vasculitis, Skin biopsy, GI bleed, Pulse steroid, Cyclophosphamide

INTRODUCTION

Henoch-Schönlein purpura (HSP) is an IgA-mediated small-vessel vasculitis that mostly impacts children under ten, with modest male predominance (male-to-female ratio 1.2:1). The global annual incidence varies between 6.2 to 70.3 per 100,000 children under 17 years, peaking between 4-6 years of age. Adult cases are rare but have more severe form of disease, when there is renal involvement with an incidence of 3.4 to 14.3 per million.[1] Diagnosis is clinical and guided by EULAR/PRINTO/PRES (European league against rheumatism/paediatric rheumatology international trials organisation / paediatric rheumatology European society) criteria, requiring palpable purpura plus at least one of the following: arthritis, abdominal pain, renal involvement which is usually considered as classic tetrad, or biopsy-proven vasculitis. Definitive confirmation is made by direct immunofluorescence of biopsy samples, usually skin or kidney.[2-4]

The treatment of IgA vasculitis varies by severity, mild cases are typically managed with supportive care and corticosteroids, while severe or refractory cases may require immunosuppressive agents such as mycophenolate mofetil (MMF) or cyclophosphamide. In cases with severe renal involvement, rituximab, intravenous immunoglobulin (IVIG), or plasma exchange may be considered.[5]

CASE REPORT

We present two distinct cases of HSP that exemplify its diverse clinical spectrum across age groups, from joint and skin involvement an adult to severe gastrointestinal vasculitis in a child, highlighting the critical need for early recognition, individualized management, and vigilance for atypical organ involvement. Our first case demonstrates 44-year-old female patient presented with a history of rash that began over the lower limbs and gradually spread to the abdomen and upper limbs, associated with pain for the past 25 days. She was admitted to a nearby hospital, laboratory tests revealed positive anti-RNP (anti-ribonucleoprotein antibodies) and anti-Sm (anti-Smith antibodies), typically indicative of an autoimmune condition. She was managed with steroids and antibiotics. However, the rashes recurred. On examination, they were erythematous, purpuric, and severe. Her lower limbs were tender and slightly swollen. She was febrile, conscious, but in pain. The patient also complained of severe joint pain for the past 20 days. Her surgical history included cholecystectomy and tubectomy. She had no known comorbidities. Systemic examination did not reveal any significant findings.

Collectively, the findings from Table 1, support systemic inflammation and an autoimmune etiology, which is consistent with HSP. She was managed with intravenous methylprednisolone 125 mg once daily, and oral hydroxyzine hydrochloride 25 mg twice a day, hydroxychloroquine 200 mg once a day and supportive care IV fluids were given. After 5 days of consistent treatment, the patient showed clinical improvement and was stable. Upon discharge, patient was prescribed a tapering dose of oral steroids along with continued hydroxyzine hydrochloride, hydroxychloroquine, and vitamin D supplementation for 8 weeks. Topical therapy included halobetasol propionate/fusidic acid cream and mupirocin ointment. She advised to continue tapering steroids and topical therapy on an outpatient basis. During follow-up, no recurrence of rash was reported.

Table 1: Pathological and laboratory findings of adult patient.

Parameter/test	Result	Interpretation
TLC	13,780 cells/mm³	Leucocytosis; suggests active inflammation
Absolute neutrophil count	10.83×10^9/l	Absolute neutrophilia; supports acute inflammatory process
Platelet count	518,000/mm³	Thrombocytosis; suggests ongoing inflammation
C-reactive protein (CRP)	26.7 mg/l	Markedly elevated, indicates acute inflammation
Erythrocyte sedimentation rate (ESR)	34.76 mm/hr	Elevated, supports acute inflammatory state
Urinalysis (CUE)	No proteinuria, no haematuria	No evidence of renal involvement
Anti-RNP and anti-Sm antibodies	Positive	Suggests autoimmune disease
Skin biopsy (Immunofluorescence)	IgA and C3c deposits in dermal vessels	Consistent with HSP

Table 2: Pathological and laboratory findings of pediatric patient.

Parameters/ tests	Findings	Interpretation
Hemoglobin	Dropped from 14.2 g/dl to 10g/dl	Suggests acute blood loss due to GI bleeding
TLC	34,200 cells/mm^3	Marked leucocytosis suggesting inflammation
Neutrophils	82%	Neutrophilia-suggesting ongoing acute inflammatory response
Platelets	5.26×10^5/µl	Mild thrombocytosis likely due to inflammation or blood loss
CRP	5.83 mg/l	Elevated-indicates systemic inflammation
Sr. creatinine	0.41 mg/dl	Normal renal function
Upper endoscopy	Severe erosive gastritis	GI mucosal involvement likely due to HSP-related vasculitis
Colonoscopy	Pancolitis	Diffuse colonic inflammation, consistent with GI vasculitis
Gastric and colon biopsy	Mucosal congestion, polymorphs, occasional vessels feature suspicious of leukocytoclastic vasculitis	Suggesting GI vasculitis, consistent with HSP flare

The second case demonstrates 13-year-old female, a known case of HSP, presented with 3-4 episodes of hematemesis and melena, accompanied by intermittent abdominal pain. She had experienced similar complaints one month ago. She was on MMF, as per her medication reconciliation. On physical examination, she was dull, lethargic, with purpuric rash seen over the lower limbs. On examination she was hypotensive and tachycardic, abdomen was soft, tender in epigastric region. She was admitted for further evaluation and management.

The findings from Table 2 suggest GI vasculitis due to HSP flare. Initially she was managed with IV fluids, ondansetron, and IV ceftriaxone 1 g for seven days, IV methylprednisolone 125 mg OD was administered for two days but later escalated to 500 mg OD due to suspected GI vasculitis secondary to HSP flare. In view of the severity and extent of GI vasculitis, she received a single dose of IV cyclophosphamide 500 mg, infused over 6 hours in 500 mL of normal saline. The child responded well to treatment and was hemodynamically stable and was discharged with prednisolone 30 mg and planned for monthly cyclophosphamide

DISCUSSION

Our first case highlights the importance of considering HSP as a potential diagnosis, though it is rare in adults. The decision to test for anti-Sm and anti-RNP antibodies was appropriate to rule out other autoimmune diseases such as systemic lupus erythematosus and mixed connective tissue disease. Several differential diagnoses were carefully evaluated and ruled out including idiopathic thrombocytopenic purpura (ITP), disseminated intravascular coagulation (DIC), thrombotic thrombocytopenic purpura (TTP), hypersensitivity vasculitis, and Rocky Mountain spotted fever, as they can present with similar cutaneous findings. However, the presence of arthralgia along with biopsy findings consistent with IgA deposition helped confirm the diagnosis of HSP, in alignment with standard diagnostic guidelines.[6]

HSP is triggered most often by upper respiratory tract infections, certain medications and vaccines[1] which stimulate the immune system to produce abnormal galactose-deficient IgA1 antibodies. These antibodies form immune complexes that circulate and deposit in small blood vessels, particularly in the skin. Once deposited, they activate the complement system, notably C3c, leading to an inflammatory response with neutrophil infiltration and vessel wall damage, a process known as leukocytoclastic vasculitis. This damage increases vascular permeability, causing red blood cells and plasma to leak into surrounding tissue, resulting in the characteristic painful, erythematous, and palpable purpura, especially on gravity-dependent areas like the lower limbs. Elevated inflammatory markers such as CRP, ESR, and thrombocytosis further supported the presence of active systemic inflammation consistent with HSP. The positive IgA anticardiolipin antibody report in this patient is increasingly linked to hypercoagulability especially when combined with thrombocytosis and vascular inflammation. This suggests a potential thrombotic risk, warranting close monitoring and possible thrombophilia workup or prophylaxis.[7,8] The patient was initiated on supportive care with intravenous fluids and a combination of medications aimed at immunosuppression, symptomatic relief, and skin healing. The recurrence of rash following initial steroid therapy highlights the importance of administering an appropriate dose and duration of corticosteroids to effectively control inflammation and prevent relapse in HSP.[5]

In our second case, though patient was on maintenance immunosuppression with MMF, her condition got worse, which required high-dose of IV steroid. In view of severe GI involvement single dose of IV cyclophosphamide was given, which resulted in clinical improvement. While cyclophosphamide is typically reserved for central nervous system or renal involvement in HSP, the literature warrants its use in life-threatening, severe, or refractory GI vasculitis also. Patients who had uncontrolled GI bleeding refractory to steroid, successfully responded to single high dose of cyclophosphamide or treatment can be combined with high dose steroid and immunosuppressants.[9,10] Second-line treatments such as rituximab, azathioprine, or MMF can be used in case of resistance. The prognosis is generally good in patients who received aggressive treatment.[1] Around 18% developed GI bleeding with common presentations were melena or occult blood, with hematemesis being less frequent.[11,12] Given the presence of pancolitis it could mimic ulcerative colitis.[13] Highlights the importance for differential diagnosis. However, patient responded to cyclophosphamide supports that GI involvement was due to HSP flare, underscoring the uniqueness of this case. Severe GI involvement in HSP is generally accompanied by renal disease but in our patient, pancolitis and erosive gastritis with no renal involvement, which is an uncommon but dramatic presentation.[14]

CONCLUSION

This case report demonstrates the clinical spectrum of HSP, which varies from typical adult cutaneous involvement to severe pediatric GI involvement without renal signs. It highlights the importance of early detection, differential diagnosis, and tailored immunosuppressive therapy. This emphasizes that in order to enhance patient care and diagnostic accuracy, general practitioners, rheumatologists, gastroenterologists, nephrologists, and dermatologists must consider HSP in relevant clinical scenarios.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Sohagia AB, Gunturu SG, Tong TR, Hertan HI. Henoch-Schonlein purpura-a case report and review of the literature. Gastroenterol Res Pract. 2010;2010:597648.
2. Roberts PF, Waller TA, Brinker TM, Riffe IZ, Sayre JW, Bratton RL. Henoch-Schönlein Purpura: A review article. South Med J. 2007;100(8):821-4.
3. Gedalia A. Henoch-Schönlein purpura. Curr Rheumatol Rep. 2004;6(3):195-202.
4. 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-Schönlein purpura. Arthritis Rheum. 1990;33(8):1114-21.
5. Castañeda S, Quiroga-Colina P, Floranes P, Uriarte-Ecenarro M, Valero-Martínez C, Vicente-Rabaneda EF, et al. IgA vasculitis (Henoch–Schönlein Purpura): An update on treatment. J Clin Med. 2024;13(21):6621.
6. Roache-Robinson P, Killeen RB, Hotwagner DT. IgA vasculitis (Henoch-Schönlein Purpura). In: StatPearls. Treasure Island (FL): StatPearls Publishing. 2025.
7. Xu L, Li Y, Wu X. IgA vasculitis update: Epidemiology, pathogenesis, and biomarkers. Front Immunol. 2022;13:921864.
8. Hetland LE, Susrud KS, Lindahl KH, Bygum A. Henoch-Schönlein Purpura: A literature review. Acta Derm Venereol. 2017;97(10):1160-6.
9. Gicchino MF, Iafusco D, Marrapodi MM, Melone R, Cuomo G, Zanfardino A, et al. Gastrointestinal Henoch-Schönlein purpura successfully treated with Mycophenolate Mofetil: Description of 2 case reports. Medicine (Baltimore). 2021;100(1):e24093.
10. Uluca Ü, Ece A, Şen V, Yel S, Tan İ, Karabel D. Management of intestinal bleeding with single-dose cyclophosphamide in Henoch-Schönlein purpura: Intestinal bleeding in HSP. Pediatr Int. 2015;57(3):498-500.
11. Guo H, Wang Z-L, Tao Z. Delayed diagnosis of abdominal Henoch-Schonlein purpura in children: A case report. World J Clin Cases. 2023;11(26):6311-7.
12. Wang S, Tang H, Du W, Ding Y. Massive gastrointestinal hemorrhage caused by Henoch-Schoenlein purpura: A case report: A case report. Medicine (Baltimore). 2021;100(50):e28240.
13. Kusumawati NRD, Pratiwi J, Low SL, Paramitha DA, Prawirasatra WA, Pratiwi NR, et al. Ulcerative Colitis with Henoch Schonlein Purpura in Pediatric Patient: A Case-Report. Arch Pediatr Gastr Hepatol Nutr. 2023;2(4):30-5.
14. Li Y, Zhang X, Liu H, Li G, Guan W, Zhang T, et al. Severe gastrointestinal involvement in pediatric IgA vasculitis: a retrospective single-center cohort study in China. Front Pediatr. 2023;11:1194214.

Cite this article as: Nimra, Lakshmi GA, Harshitha KB, Fathima T. The clinical spectrum of Henoch-Schönlein purpura: insights from two distinct presentations. Int J Res Dermatol 2025;11:340-3.