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Case Study
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A Rare but Aggressive Renal Manifestation of IgA Vasculitis in an 11-Year-Old Girl

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Abstract
Although rare, crescentic Henoch–Schönlein purpura (HSP) nephritis poses a significant diagnostic and therapeutic challenge due to its variable clinical presentation and unpredictable disease course. Crescentic HSP nephritis is a severe manifestation accounting for less than 5% of HSP nephritis cases and is associated with rapid deterioration of renal function, potentially progressing to end-stage renal disease if not promptly treated. Early diagnosis, close monitoring, and timely recognition of complications are essential for initiating appropriate management and preserving renal function. Lifestyle modifications, including dietary regulation and physical activity, may contribute to symptom control and improvement in overall health. Renal involvement in HSP typically presents with hematuria and mild to moderate proteinuria; however, crescentic involvement represents a more aggressive pathological variant requiring intensive therapy.

Keywords: Henoch–Schönlein purpura, HSP nephritis, crescentic glomerulonephritis, pediatric patient, vasculitis, immunosuppressive therapy, renal biopsy.

INTRODUCTION

Henoch–Schönlein purpura (HSP), also known as IgA vasculitis, is a systemic small-vessel vasculitis predominantly affecting children. It is classically characterized by a tetrad of palpable purpura, arthritis or arthralgia, gastrointestinal manifestations, and renal involvement. The disease most commonly affects children between 3 and 15 years of age. Crescentic HSP nephritis is a rare but severe complication, representing less than 5% of HSP nephritis cases [1].

Renal involvement in HSP commonly manifests as hematuria and proteinuria of mild to moderate severity. In contrast, crescentic HSP nephritis is defined by the presence of crescents in the glomeruli on renal biopsy and is associated with a more aggressive clinical course, particularly in pediatric patients. Despite its low incidence, this condition presents considerable challenges in diagnosis and management due to its unpredictable progression [2].

EPIDEMIOLOGY

Henoch–Schönlein purpura is the most common systemic vasculitis in children, with an estimated annual incidence of 10–20 cases per 100,000 children. It predominantly affects individuals aged 3-15 years, with peak incidence observed between 4 and 6 years of age [3].

The exact etiology of HSP remains unclear; however, immune complex–mediated mechanisms are believed to play a central role. Potential triggering factors include infections, medications, and environmental exposures [4]. Although most cases of HSP are self-limiting, approximately 30–50% of patients develop complications involving the skin, joints, gastrointestinal tract, or kidneys. Renal involvement occurs in nearly 30% of patients, with a small subset progressing to severe disease, including crescentic glomerulonephritis [5].

CASE STUDY

An 11-year-old female child presented with recurrent generalized tonic–clonic seizures occurring every 10 minutes, moderate renal failure, severe anemia, and severe hypertension. She was initially admitted to a hospital in Hyderabad with complaints of diffuse purpuric rash over the extremities, bilateral leg swelling, fever, joint pain, cough, vomiting, and decreased urine output. Based on clinical findings, she was suspected to have HSP nephritis with renal failure and hematuria.

The patient was treated with pulse steroid therapy (five doses of intravenous methylprednisolone) and a single bolus of intravenous cyclophosphamide. A renal biopsy confirmed the diagnosis of HSP nephritis with crescent formation. Although serum creatinine levels initially decreased, hematuria persisted. She was treated with mesna and later discharged.

Subsequently, the patient was readmitted to another hospital with severe hypertension and diarrhea, leading to the discontinuation of immunosuppressive therapy. She underwent three sessions of plasma exchange using fresh frozen plasma and albumin, along with sustained low-efficiency dialysis (SLED). During hospitalization, she developed recurrent seizures, and magnetic resonance imaging revealed findings consistent with posterior reversible encephalopathy syndrome (PRES) with multiple infarcts in the parieto-occipital and thalamic regions, suggestive of central nervous system vasculitis. Despite continued plasma exchange and dialysis, her level of consciousness remained poor, and she was discharged against medical advice.

PATHOPHYSIOLOGY

Crescentic nephritis is mediated by immune mechanisms and may arise from autoimmune conditions such as anti–glomerular basement membrane (anti-GBM) disease, antineutrophil cytoplasmic antibody (ANCA)–associated vasculitis, or immune complex–mediated diseases including lupus nephritis and IgA vasculitis [6]. In immune complex–mediated diseases, deposition of immune complexes in the glomeruli activates the complement system, leading to inflammation and crescent formation [7].

This inflammatory process results in rapid loss of renal function and may progress to end-stage renal disease if not treated promptly. Management involves aggressive immunosuppressive therapy to suppress immune activation and supportive care to preserve renal function [8].

LABORATORY INVESTIGATIONS

Abnormal laboratory findings included normocytic normochromic anemia, elevated serum creatinine indicating impaired renal function, metabolic alkalosis, and reduced complement C3 levels suggestive of immune-mediated disease activity.

TREATMENT

The patient received anticonvulsants for seizure control, corticosteroids for immunosuppression, antihypertensive agents for blood pressure control, and supportive medications including antiemetics, proton pump inhibitors, vitamin supplements, and multivitamins.

DISCUSSION

Crescentic HSP nephritis is a rare but severe complication characterized by crescent formation on renal biopsy. Early diagnosis and prompt initiation of immunosuppressive therapy are essential to prevent irreversible renal damage. Despite aggressive management, patients may develop complications such as severe hypertension, central nervous system vasculitis, and recurrent seizures, highlighting the complexity of managing this condition [9-11].

PREVENTIVE ASPECTS OF MANAGEMENT

Early diagnosis and continuous monitoring are critical for optimal outcomes. Immunosuppressive therapy remains the cornerstone of treatment, with corticosteroids as first-line agents and additional immunosuppressants reserved for refractory cases [10]. Supportive care, including strict blood pressure control and electrolyte management, is essential [11]. Lifestyle modifications such as low-sodium and low-protein diets and regular physical activity may further support renal health [12].

FUTURE PERSPECTIVES

Future research should focus on developing targeted therapies, identifying reliable prognostic biomarkers, and advancing non-invasive diagnostic tools. Precision medicine approaches may help individualize treatment strategies, improve outcomes, and minimize adverse effects. Long-term follow-up and patient education are essential for disease surveillance and prevention of late complications [13-15].

CONCLUSION

Crescentic HSP nephritis is a rare but potentially life-threatening complication of HSP, particularly in pediatric patients. Early recognition, aggressive treatment, and multidisciplinary care are essential to improve outcomes. Further research is needed to refine management strategies and enhance long-term prognosis.

REFERENCES

1. Pohl M. Henoch–Schönlein purpura nephritis in children. *Nat Rev Nephrol*. 2015;11(9):563–573.
2. Narchi H. Henoch–Schönlein purpura nephritis in children: incidence, pathogenesis and management. *World J Pediatr*. 2019;15(2):135–145.
3. Dudley J, Smith G, Llewellyn-Edwards A. Therapy for children with Henoch–Schönlein purpura nephritis: a systematic review. *Int J Nephrol*. 2013;2013:1–10.
4. Coppo R, Andrulli S, Amore A. Clinical outcome in children with Henoch–Schönlein nephritis. *Pediatr Nephrol*. 2006;21(3):354–360.
5. Bogdanović R. Henoch–Schönlein purpura nephritis in children: risk factors, prevention and treatment. *Acta Paediatr*. 2009;98(12):1882–1889.
6. Chang WL, Yang YH, Lin YT. Association between allergic diseases and risks of HSP nephritis. *Pediatr Res*. 2016;79(4):559–565.
7. Sharma A, et al. Crescentic glomerulonephritis in pediatric HSP. *Saudi J Med Allied Sci*. 2021;9:1035–1040.

8. Weiss PF. Drug treatments for nephritis in children with IgA vasculitis. *Clin Rheumatol*. 2018;37(6):1523–1532.
9. Pillebout E, et al. Comparison between adults and children with HSP nephritis. *Pediatr Nephrol*. 2017;32(3):459–466.
10. Wang Y, et al. Urinary MCP-1 levels in children with HSP nephritis. *Clin Nephrol*. 2012;78(5):388–395.
11. Kawasaki Y. Pathogenesis and treatment of pediatric HSP nephritis. *Clin Exp Nephrol*. 2011;15(5):648–657.
12. Sinha R, et al. Henoch–Schönlein purpura nephritis in children: prognosis and treatment experiences. *J Pediatr Nephrol*. 2019;4(1):1–8.
13. Ronkainen J, et al. Long-term results in children with Henoch–Schönlein nephritis. *Kidney Int*. 2002;62(5):1681–1689.
14. Feng D, et al. HSP nephritis with nephrotic proteinuria in children. *Pediatr Rheumatol*. 2018;16(1):1–9.
15. Niaudet P. Henoch–Schönlein purpura nephritis. *Pediatr Nephrol*. 2016;31(7):1031–1043.