Research Article

A Typical Renal Manifestation of Henoch – Schonlein Purpura in A Child: A Case Report

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Abstract

Henoch–Schönlein purpura (HSP), currently termed IgA vasculitis, is the most common small-vessel vasculitis in children. It is characterized by non-thrombocytopenic palpable purpura, arthritis or arthralgia, gastrointestinal manifestations, and renal involvement. Although the disease is generally self-limiting, renal involvement can lead to significant long-term morbidity and requires early recognition and appropriate management.

We report the case of a 10-year-old female who presented with palpable purpura over both lower limbs, bilateral lower-limb edema, hematuria, and proteinuria following a recent upper respiratory tract infection. Laboratory investigations demonstrated significant urinary abnormalities suggestive of renal involvement. Skin biopsy revealed features of small-vessel vasculitis, while renal biopsy demonstrated IgA-dominant immune complex deposition predominantly within the glomerular mesangium, confirming IgA vasculitis with nephritis. In view of the significant renal involvement, the child was treated with corticosteroids and azathioprine, following which clinical and laboratory improvement was observed.

This case highlights IgA vasculitis presenting with prominent renal involvement requiring histopathological confirmation and immunosuppressive therapy. Early identification and appropriate assessment of renal involvement, timely renal biopsy in selected patients, and multidisciplinary management are essential to reduce the risk of progressive renal disease and improve long-term outcomes in children with IgA vasculitis.

KEYWORDS: IgA vasculitis; Henoch–Schönlein purpura; hematuria; proteinuria; nephritis; skin biopsy; renal biopsy; corticosteroids; azathioprine; vasculitis.

Introduction

A 10-year-old female child, the first born of a non-consanguineous marriage, was brought to paediatric Outpatient department by her mother with a history of reddish- purple rashes over bilateral lowerlimbs which started 1 week followed by cola coloured urine and swelling in her bilateral lower limbs(left>right) associated with generalized tiredness for 3 days

The purpura was insidious in onset and progressive in nature. It initially appeared over the left leg and gradually extended to the thigh. Multiple small, purpurae were noted over both lower limbs(figure 1) and thighs, associated with itching. There was also swelling of the left lower limb(figure 2&3) insidious in onset which started at the level of ankle and progressively increasing up knee , associated with difficulty in walking. The child had a history of lowgrade, intermittent fever and sore throat 1 week ago . There was no history of similar complaints in the past. There was no significant birth history, and developmental milestones were appropriate for age. There was no family history of bleeding disorders.

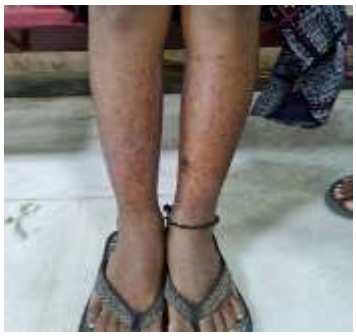

Figure 1. shows purpurae

Figure 2. Shows swelling of left lower limb

Figure.3 Shows swelling of left lower limb

On examination, vitals: pulse rate: 92beats/minute, respiratory rate: 24 cycles/minute blood pressure : 100/70 mmHg, non-pitting pedal edema was noted at the level of bilateral ankle. Local rise in temperature and tenderness were noted. Peripheral pulses were normal. Dermatological examination revealed diffuse, multiple, small maculopapular palpable purpura over both lower limbs and thighs. Systemic examination revealed mild splenomegaly.

Investigation	Result	Reference Range / Interpretation
Complete Blood Count		
Hemoglobin (Hb)	7.9 g/dL	Low (Microcytic hypochromic anemia)
Mean Corpuscular Volume (MCV)	68 fL	Low
Mean Corpuscular Hemoglobin (MCH)	21 pg	Low
White Blood Cell (WBC) Count	13,481/mm³	Mild leukocytosis
Neutrophils	64%	Relative neutrophilia
Lymphocytes	24%	Within normal limits
Eosinophils	6%	Mild eosinophilia
Platelet Count	257,700/mm³	Normal
Urinalysis		
Hemoglobin	2+	Positive
Protein	3+	Significant proteinuria
Specific Gravity	1.015	Normal
pH	7.5	Slightly alkaline
Nitrite	Negative	No nitrituria
Appearance	Cloudy	Abnormal
Bacterial Flora	Discrete bacterial flora	Present

Pyocytes	3/high-power field	Mild pyuria
Red Blood Cells	Numerous well-preserved RBCs	Significant hematuria
Kidney function Test		
Serum creatinine	0.62 mg/dl	Normal
Serum Uricacid	4.2 mg/dl	Normal
Serum Electrolytes		
Sodium	138 mmol/L	Normal
Potassium	4.3 mmol/L	Normal
Chloride	103mmol/L	Normal
Bicarbonate	23mmol/L	Normal
Calcium	9.2mg/dl	Normal
Phosphorous	4.6mg/dl	Normal
Serum Biochemistry		
Total Protein	6.0 g/dL	Lower limit of normal
Albumin	3.1 g/dL	Mild hypoalbuminemia
Autoimmune and Serological Investigations		
Antinuclear Antibody (ANA)	1:80, dense fine speckled pattern	Positive, low titer
Anti-Ro (SSA)	Negative	Negative
Anti-La (SSB)	Negative	Negative
Anti-Sm	Negative	Negative
Anti-dsDNA	Negative	Negative
Rheumatoid Factor	<9.69 IU/mL	Normal (Reference: ≤15 IU/mL)
Antistreptolysin-O (ASO)	238 IU/mL	Mildly elevated (Reference: <200 IU/mL)
Anti-Cardiolipin Antibody	Negative	Negative
Beta-2 Glycoprotein-1 Antibody	Negative	Negative
Lupus Anticoagulant	Negative	Negative
P-ANCA	Positive (1:80)	Positive
C-ANCA	Negative	Negative
Atypical ANCA	Negative	Negative
Anti-Myeloperoxidase (Anti-MPO)	55 U/mL	Markedly elevated (Reference: ≤3.5 U/mL)
Complement C3	107 mg/dL	Normal (90–180 mg/dL)
Complement C4	28.5 mg/dL	Normal (10–40 mg/dL)

A skin biopsy demonstrated features of perivascular neutrophils, lymphocytic infiltrates suggestive of small vessel vasculitis [Figure :1]

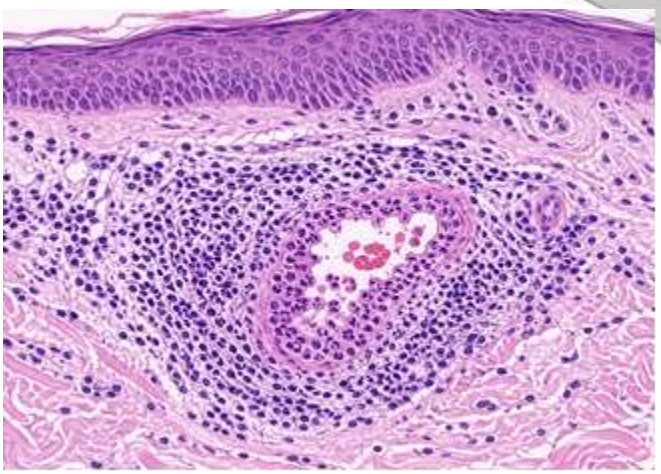

Figure : 4 Skin biopsy shows perivascular neutrophils, lymphocytic infiltrates suggestive of small vessel vasculitis

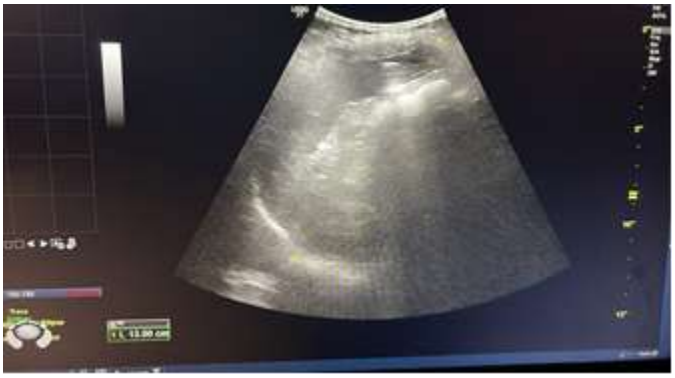

Figure : 5 Ultrasonography of the abdomen showed mild splenomegaly.

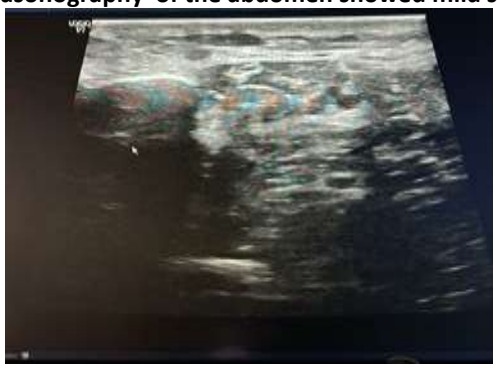

Figure 6: USG of the leg showed diffuse subcutaneous edema around the left lateral malleolus and dorsum of the left foot.

Doppler study was normal. Kidney biopsy shows IgA dominant immune complex deposition in the glomerular mesangium

Based on the history, examination, laboratory findings, skin biopsy and kidney biopsy, the diagnosis is most consistent with IgA Vasculitis (Henoch schonlein purpura) with IgA vasculitis nephritis. Although the patient demonstrated significant MPO-ANCA positivity, the diagnosis of microscopic polyangiitis was excluded based on the renal biopsy, which demonstrated mesangial IgA-dominant immune complex deposition rather than pauci-immune necrotizing glomerulonephritis.

While hypersensitivity vasculitis also shows small vessel vasculitis on skin biopsy, a definitive kidney biopsy showing IgA-dominant immune complex deposition in the mesangium precludes it effectively.

Similarly, post-streptococcal glomerulonephritis was considered because of the antecedent sore throat and mildly

elevated ASO titre; however, normal complement levels and renal histopathology were not supportive. Lupus nephritis was ruled out by the absence of disease-specific autoantibodies (anti-dsDNA, anti-Sm), normal complement levels, and characteristic renal biopsy findings. PAN (Polyarteritis nodosa) was ruled out due to the absence of necrotizing vasculitis of medium-sized arteries in skin biopsy.

As the patient had normal platelet count with palpable purpura, a diagnosis of immune thrombocytopenic purpura was also excluded.

An multidisciplinary team comprising of paediatricians , nephrologists, dermatologists were involved and the patient was initially treated conservatively with intravenous fluid and oral paracetamol when needed along with antibiotics for 7 days. The child was subsequently treated with corticosteroids (oral prednisolone at 1mg/kg/day for 5 days) and immunosuppressive therapy-Azathioprine at 3 mg/kg/day for 2 months.

The patient was reviewed after 2 weeks, 1 month, 2 months, and 6 months in the pediatric nephrology outpatient clinic. At the 2-week follow-up, the palpable purpura and lower limb edema had resolved completely, and the child was able to walk without difficulty. Generalized fatigue improved, and there was no recurrence of fever or skin lesions. At the 1-month follow-up, urinalysis demonstrated a marked reduction in proteinuria and microscopic hematuria. Blood pressure remained within the normal range for age, and serum creatinine and serum electrolytes continued to be normal. Oral prednisolone was gradually tapered, while azathioprine was continued for 2 months under close monitoring.

At the end of 2 months, azathioprine was discontinued after completion of the planned course. The patient remained clinically asymptomatic with no recurrence of purpura, joint symptoms, or edema. Repeat laboratory investigations showed improvement in urinary findings with preserved renal function. At the 6-month follow-up, the child continued to remain well without relapse. Blood pressure, serum creatinine, and estimated glomerular filtration rate were normal, and urinalysis showed no significant proteinuria or active urinary sediment. Long-term follow-up was advised with periodic blood pressure measurement, urinalysis, and renal function assessment to detect any late recurrence or progression of IgA vasculitis nephritis.

DISCUSSION

Henoch–Schönlein purpura (HSP) commonly presents with a well-known tetrad of symptoms: non-thrombocytopenic purpura, polyarthralgia, abdominal pain, and renal involvement. These clinical features are the result of systemic vasculitis. Therefore, the broadly accepted diagnostic criteria define HSP as the presence of palpable purpura along with at least one of the following: widespread abdominal pain, biopsy findings showing dominant IgA deposition, arthritis, or kidney involvement. The purpura, which is the defining feature, appears as raised lesions measuring 2–10 mm in diameter, often occurring in clusters and sometimes accompanied by petechiae and ecchymoses. These lesions are non-blanching due to the leakage of blood into the skin and generally persist for several weeks. They are most frequently observed on the lower extremities, buttocks, and arms. In certain cases, they may be preceded by a maculopapular or urticarial rash that can resemble other dermatological conditions. Localized angioedema, particularly involving the knees, ankles, and hands, may develop before the onset of purpura. Polyarthralgia often leads patients to avoid bearing weight, especially affecting the knees and ankles, although it typically resolves without causing permanent damage. Abdominal pain is usually diffuse and colicky in nature, appearing within a week after the rash and may be associated with nausea, vomiting, and either bloody or mucoid diarrhea. When renal involvement occurs, it generally presents as mild glomerulonephritis characterized by proteinuria, microscopic hematuria, and occasionally red blood cell casts.(1)

HSP is defined by the accumulation of IgA-containing immune complexes within tissues, which activate the complement system via the alternative pathway, resulting in endothelial injury and widespread vasculitis.(2) The condition often follows an unnoticed upper respiratory tract infection and is more prevalent during the autumn and winter months. A range of environmental and infectious triggers have been associated with HSP, including group A β-hemolytic Streptococcus, Mycoplasma pneumoniae, varicella, hepatitis B virus, certain foods, medications, insect bites, and exposure to cold. The disease process involves interactions between leukocytes and endothelial cells, and genetic susceptibility—such as a link with HLA-DRB1*01—also plays a role. (3)

The diagnosis of HSP is primarily clinical, based on the presence of purpura along with other features of the tetrad. There is no single definitive laboratory test for HSP, although investigations are useful for excluding other conditions and assessing organ involvement. Complete blood counts are typically normal or may reveal mild leukocytosis. Serum IgA levels can be elevated but do not correlate with disease severity, while erythrocyte sedimentation rates

may be increased. Throat cultures may be positive for group A β-hemolytic Streptococcus if a recent infection has occurred. Elevated von Willebrand factor levels may reflect endothelial damage. Tests for autoantibodies and complement levels are usually within normal limits. Urinalysis is essential and frequently demonstrates microscopic hematuria and red blood cell casts(4). A definitive diagnosis can be confirmed through a skin biopsy showing leukocytoclastic vasculitis with IgA deposition, and immunofluorescence studies may also reveal deposits of C3 and fibrinogen in the vessel walls.(5) Nüsken and Weber highlighted that renal involvement is the principal determinant of long-term prognosis in IgA vasculitis, making early recognition and careful monitoring of nephritis essential. They emphasized that kidney biopsy remains the gold standard for assessing disease severity and guiding immunosuppressive therapy, while persistent proteinuria, hypertension, and impaired renal function are the strongest predictors of adverse renal outcomes.(9)

Treatment depends on the severity of the disease and the extent of organ involvement. Most cases resolve on their own within 4–6 weeks, with management focused on supportive care and reassurance. In more severe cases, such as those involving significant hemorrhagic complications, hospitalization and close monitoring may be necessary. Joint pain and soft tissue swelling are generally managed with NSAIDs, particularly acetaminophen, while corticosteroids may be required for rapid symptom relief or in cases of severe abdominal pain or scrotal involvement. Patients with significant renal disease may need high-dose corticosteroids or additional immunosuppressive therapy.(6) According to Hastings et al., renal involvement is the major determinant of prognosis in IgA vasculitis. Although most children recover completely, patients with persistent proteinuria, hypertension, or impaired renal function require close follow-up and may benefit from immunosuppressive therapy. Our patient had significant proteinuria and biopsy-proven mesangial IgA deposition, justifying treatment with corticosteroids and immunosuppressive therapy.(8)

The prognosis of HSP is generally favorable, with most patients achieving complete recovery. Recurrences can occur, although they are usually milder. Outcomes are typically better in individuals with minimal renal involvement, whereas persistent kidney disease, such as chronic nephritis, can result in long-term complications including renal insufficiency. Follow-up usually begins within a few weeks after diagnosis to evaluate disease progression and manage symptoms. Early follow-up visits are commonly scheduled every 1–2 months to monitor for complications involving the skin, joints, gastrointestinal tract, and kidneys. Patients with more severe disease, particularly those with renal involvement, may require continued monitoring every 3–6 months. Regular urine tests and blood investigations(kidney function test) are important for assessing kidney function and overall health, and follow-up plans should be tailored according to the patient's clinical course and response to treatment.(7) Jelusic et al. emphasized that IgA vasculitis nephritis is the most significant predictor of long-term morbidity in children and that kidney biopsy should be considered in patients with significant proteinuria or persistent urinary abnormalities to guide treatment. In our patient, biopsy-confirmed mesangial IgA deposition with preserved renal function supported the diagnosis of IgA vasculitis nephritis and justified early corticosteroid and immunosuppressive therapy.(10) Xu et al. reported that the pathogenesis of IgA vasculitis involves dysregulated IgA1 glycosylation, formation of pathogenic immune complexes, and activation of the alternative and lectin complement pathways, ultimately resulting in small-vessel injury. In our patient, biopsy-proven mesangial IgA deposition with significant proteinuria is consistent with these pathogenic mechanisms and supports the diagnosis of IgA vasculitis nephritis.(11) Peruzzi and Coppo reported that although IgA vasculitis nephritis and IgA nephropathy have similar histopathological features, the presence of systemic manifestations such as palpable purpura, arthritis, and gastrointestinal symptoms distinguishes IgA vasculitis. In our patient, the combination of palpable purpura, biopsy-proven mesangial IgA deposition, and significant proteinuria confirmed the diagnosis of IgA vasculitis nephritis.(12) Williams et al. reported that although emerging urinary biomarkers may improve the early detection of IgA vasculitis nephritis, their clinical utility remains limited because of insufficient validation and variability among studies. Therefore, conventional investigations such as urinalysis, assessment of proteinuria, and kidney biopsy continue to be the cornerstone for diagnosing and monitoring renal involvement. (13)

CONCLUSION

HSP is a sudden-onset, self-limiting multisystem vasculitis that mainly occurs in children. It affects multiple organ systems, with a palpable purpuric rash being a characteristic feature present in every case. Treatment is usually centered on relieving symptoms, although some patients may need immunosuppressive or immunomodulatory therapy. Although the use of corticosteroids is still controversial, studies suggest they may be useful in patients with

severe renal involvement, marked abdominal pain, and soft tissue edema. Even though most cases resolve spontaneously, close observation is necessary to identify rare but serious complications. Early diagnosis, proper management, and Multidisciplinary collaboration, including nephrologists, rheumatologists, and dermatologists, is crucial along with detailed patient education are essential for controlling symptoms and preventing severe outcomes.

LEARNING POINTS

- IgA vasculitis should be considered in children presenting with palpable, non-thrombocytopenic purpura, particularly following a recent upper respiratory tract infection.
- Renal involvement may be clinically significant and can present with hematuria and substantial proteinuria even when serum creatinine and renal function remain normal.
- Renal biopsy can be crucial in selected patients with significant or persistent urinary abnormalities, as demonstration of IgA-dominant mesangial immune-complex deposition can confirm IgA vasculitis nephritis and help assess severity.
- Positive MPO-ANCA does not always indicate microscopic polyangiitis. In this case, the renal biopsy showing mesangial IgA-dominant deposits rather than pauci-immune necrotizing glomerulonephritis supported IgA vasculitis nephritis.
- Long-term renal surveillance is essential, even when renal function is initially preserved, with periodic assessment of blood pressure, urinalysis, proteinuria, and renal function.
- Multidisciplinary management involving paediatricians, nephrologists, and dermatologists can facilitate timely diagnosis, treatment, and follow-up in children with significant multisystem involvement.

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