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Adult-Onset IgA Vasculitis Complicated by Kidney Failure at Disease Onset in a Nepalese Patient: A Case Report

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Abstract:-

➢ ***Introduction:***

Immunoglobulin A (IgA) vasculitis, previously known as Henoch-Schönlein purpura, is an immune complex-mediated small vessel vasculitis primarily affecting children. While rare in adults, it can present with more severe manifestations, particularly involving the kidneys. This case report details the presentation and management of adult-onset IgA vasculitis with significant renal involvement.

➢ ***Case Report:***

A 43-year-old male with a history of bipolar disorder presented with facial swelling, shortness of breath, and decreased urine output following an upper respiratory infection. Initial investigations revealed elevated blood pressure and renal impairment. Despite supportive treatment, his condition worsened, leading to a referral to a tertiary care center. He exhibited symptoms consistent with IgA vasculitis, including joint pain, rash, and nephrotic-range proteinuria. The diagnosis was confirmed through a skin biopsy and 24-hour urine collection. The patient was treated with intravenous methylprednisolone, oral prednisone, and an ACE inhibitor. His renal function improved with this regimen.

➢ ***Discussion:***

Adult-onset IgA vasculitis can present with severe kidney involvement, including nephrotic-range proteinuria and elevated serum creatinine, which are associated with poorer outcomes compared to pediatric cases. The patient's management, involving glucocorticoids and an ACE inhibitor, aligns with current treatment recommendations for significant renal involvement. Long-term prognosis in adults remains challenging, with a higher risk of end-stage kidney disease compared to children. Vigilant monitoring and tailored treatment strategies are crucial for improving outcomes.

➢ ***Conclusion:***

This case underscores the potential severity of adult-onset IgA vasculitis and highlights the importance of early diagnosis and aggressive management to mitigate long-term renal complications. Ongoing research is necessary to refine treatment approaches and enhance outcomes for adults with this condition.

Keywords:- *IgA Vasculitis, Henoch-Schönlein Purpura, Proteinuria, Prednisolone, ACE- Inhibitor, Vasculitis.*

I. INTRODUCTION

Immunoglobulin A (IgA) vasculitis, which used to be known as Henoch-Schönlein purpura, is a type of immune complex vasculitis that mainly targets small blood vessels. Heberden first reported the condition in 1802, and Schönlein later identified the link between purpura and joint pain in 1837. Henoch expanded the description by including gastrointestinal symptoms and kidney involvement in the syndrome [1]. The exact cause of its development is not well understood. Still, it is thought that an unidentified antigenic trigger might lead to an increase in circulating IgA and complement activation, resulting in widespread inflammation of small blood vessels [2].

Immunoglobulin A (IgA) vasculitis is the most prevalent type of systemic vasculitis in children, typically affecting those aged 3 to 15 years, with an incidence of 3 to 27 per 100,000 [3]. In adults, IgA vasculitis is rarer [4], with an annual incidence estimated at 0.1 to 1.8 per 100,000 people [1]. Retrospective studies indicate that 20 to 30 percent of IgA vasculitis cases occur in adults, who tend to experience more severe kidney complications than children [5,6]. Most research indicates a slight predominance in males, with incidence rates of 6.1 per 100,000 for men and 3.7 per 100,000 for women [7].

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The characteristics of IgA vasculitis in adults were well demonstrated in a retrospective review of 250 French patients with a median age of 50. At the time of diagnosis, 96 percent of patients exhibited palpable purpura, 61 percent experienced arthritis, and 48 percent reported gastrointestinal symptoms. Nearly one-third of the patients developed kidney insufficiency, defined as a creatinine clearance of less than 50 mL/minute per m^2, within four months of their initial presentation [8]. Adults with IgA vasculitis (IgAV) display clinical symptoms similar to those seen in children [8]. However, there are notable differences: intussusception is rarely observed in adults, and they face a greater likelihood of developing severe kidney issues, such as end-stage kidney disease (ESKD) [4,6]. IgA vasculitis is usually diagnosed by observing clinical symptoms, including palpable purpura without thrombocytopenia or coagulopathy, and two or three additional features: arthritis or arthralgia, abdominal pain, and kidney disease [9].

We present the case of a 43-year-old man who was diagnosed with IgA vasculitis following an upper respiratory tract infection that was complicated by renal involvement.

II. CASE REPORT

A 43-year-old male with a known history of Bipolar Disorder, managed with Lithium 300 mg daily for the past three years, presented with a 15-day history of facial swelling and shortness of breath. The patient reported that his symptoms began with swelling in the periorbital region, gradually progressing to involve his entire face. Additionally, he experienced acute shortness of breath that initially occurred with exertion, such as walking uphill or climbing stairs, but eventually became present even at rest. Over the past three days, he also noted a decrease in urine output and the presence of hematuria. Notably, a week before the onset of these symptoms, the patient had experienced a sore throat.

On physical examination, the patient appeared moderately built with normal vital signs except for a blood pressure of 140/100 mmHg in the sitting position. The rest of the physical examination was unremarkable. Initial laboratory investigations, including Complete blood count(CBC), Renal function test(RFT), Serum Lithium, Complement levels, Acute phase reactants, and Spot Protein Creatinine ratio(PCR), are summarized in Table 1.

Table 1 Laboratory Investigations Showing Decreased Hemoglobin level, Increased Acute Phase Reactants, Deranged Renal Function Test, and Increased Amount of Protein in 24-Hour Urine Collection.

Test	Observed value	Reference range
Complete Blood Count		
Hemoglobin	9.2 g/dL	13-18 f/dL
Red Blood cells	3.28 millions/ µL	4.7-6.0 millions/ µL
Total leucocyte count	11100 cells/ µL	4000-11000 cells/ µL
Neutrophil	80 %	40-70 %
Lymphocyte	18 %	20-25 %
Monocyte	1 %	2-8 %
Eosinophil	1 %	1-6 %
Mean corpuscular volume(MCV)	86.2 fL	76-98 fL
Packed cell volume	28.1 %	40-52 %
Total Platelet count	137000 cells/ µL	150000-450000cells/ µL
Erythrocyte Sedimentation Rate(ESR)	58 mm/hour	0-20 mm/hour
C-reactive Protein(CRP)	29 mg/L	<5 mg/L
Renal Function Test		
Serum Urea	119 mg/dL	17-43 mg/dL
Serum Creatinine	3.33 mg/dL	0.72-1.18 mg/dL
Sodium (Na+)	139 mmol/L	135-145 mmol/L
Potassium (K+)	5 mmol/L	3.5-5.5 mmol/L
Phosphorus	6.7 mg/dL	2.5-4.5 mg/dL
Calcium (Total)	7.2 mg/dL	8.4-10.2 mg/dL
Spot Protein Creatinine Ratio(PCR)		
Spot Urinary Protein	782.6 mg/dL	
Spot Urinary Creatinine	190.1 mg/dL	
Spot Urinary PCR	4.11	<0.2
Urinary Protein/24 hour	4674 mg/24 hour	20-140 mg/24 hour
Complement level		
Complement 3 (C3)	0.88 g/L	0.80-1.85 g/L
Complement 4 (C4)	0.22 g/L	0.10-0.40 g/L
Antinuclear antibodies	Positive	NA
Lithium, Serum	0.77 mmol/L	0.40-1.20 mmol/L

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The initial management plan was initiated based on the assumption of primary IgA nephropathy. The patient received supportive care, intravenous fluids, and blood pressure control targeting optimal levels using an angiotensin-converting enzyme inhibitor (ACE-I) and moderate protein restriction in his diet. This approach gradually decreased the patient's serum creatinine, as depicted in Figure 1. However, on the sixth day of hospitalization, the patient's serum creatinine unexpectedly rose to 5.03 mg/dL and continued to increase over the next two days (Figure 1). Due to the worsening renal function, the patient was referred to a tertiary care center for further management. Despite the recommendation, the patient delayed visiting the tertiary care center for a week, feeling subjectively better and choosing to avoid hospital admission.

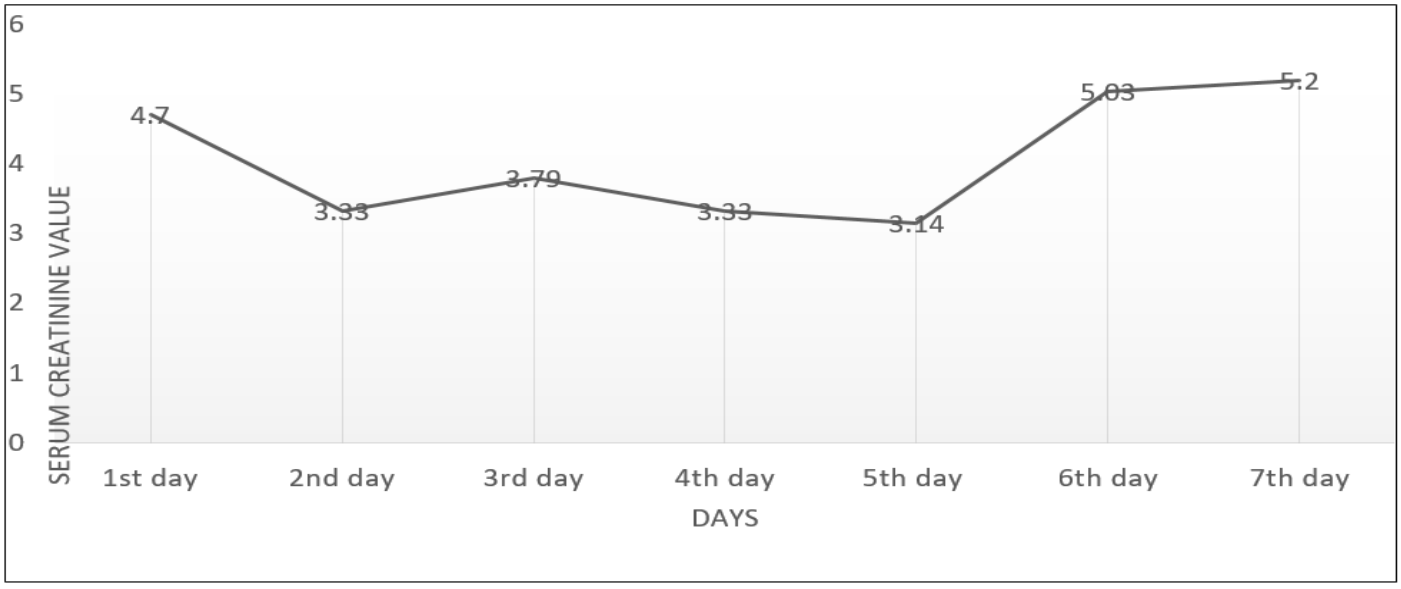

Fig 1 Serum Creatinine Trend—Change in Serum Creatinine levels from the day of Admission through the 7th day of Hospitalization.

Upon admission to the tertiary care center, the patient reported new symptoms, including joint pain and a rash on his bilateral lower limbs and back. He also complained of persistent hematuria and generalized body swelling. Physical examination revealed an erythematous macular rash that was itchy but non-painful, appearing in crops and symmetrically distributed on his back and lower limbs (Figure 2). The patient described migratory arthralgia primarily involving the hips and knees, accompanied by periarticular swelling. His vitals were stable, but abdominal distension was noted, with other examination findings remaining unremarkable.

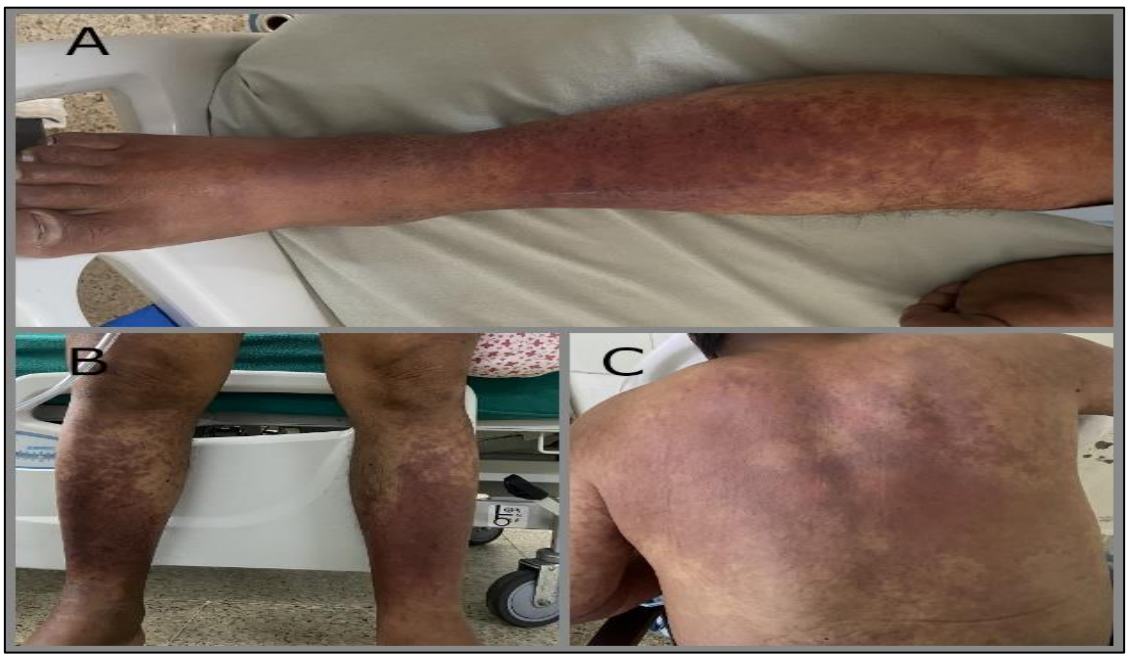

Fig 2 Clinical Manifestations of IgA Vasculitis Rash. A: Erythematous Rash in Crops on the Right Lower Limb. B: Symmetrical Distribution of the Rash on Bilateral Lower Limbs. C: Erythematous Rash on the Back.

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Based on the clinical criteria, the patient was diagnosed with IgA vasculitis (Henoch-Schönlein purpura). A 24-hour urine collection revealed protein excretion of 4641 grams. The patient was started on intravenous methylprednisolone 500 mg, ACE inhibitors, and supportive therapy. A skin biopsy of the lower legs was performed, which showed that the epidermis was lined by keratinized stratified squamous epithelium with basal layer vacuolar alteration. The underlying fibro collagenous dermis showed moderate lymphocytic perivascular infiltrates with extravasated red blood cells and hemosiderin pigments suggestive of lymphocytic vasculopathy. This confirmed our diagnosis of IgA vasculitis. The patient's medication regimen was maintained, and kidney function was closely monitored. The trend of his kidney function is depicted in Figure 3. His creatinine levels stabilized in the low 3 mg/dL range over a few days, and he was subsequently discharged with a prescription for Prednisolone 40 mg daily and an ACE-I, with a follow-up plan to monitor urine protein excretion and serum creatinine every two weeks for the first month, then monthly for the next six months.

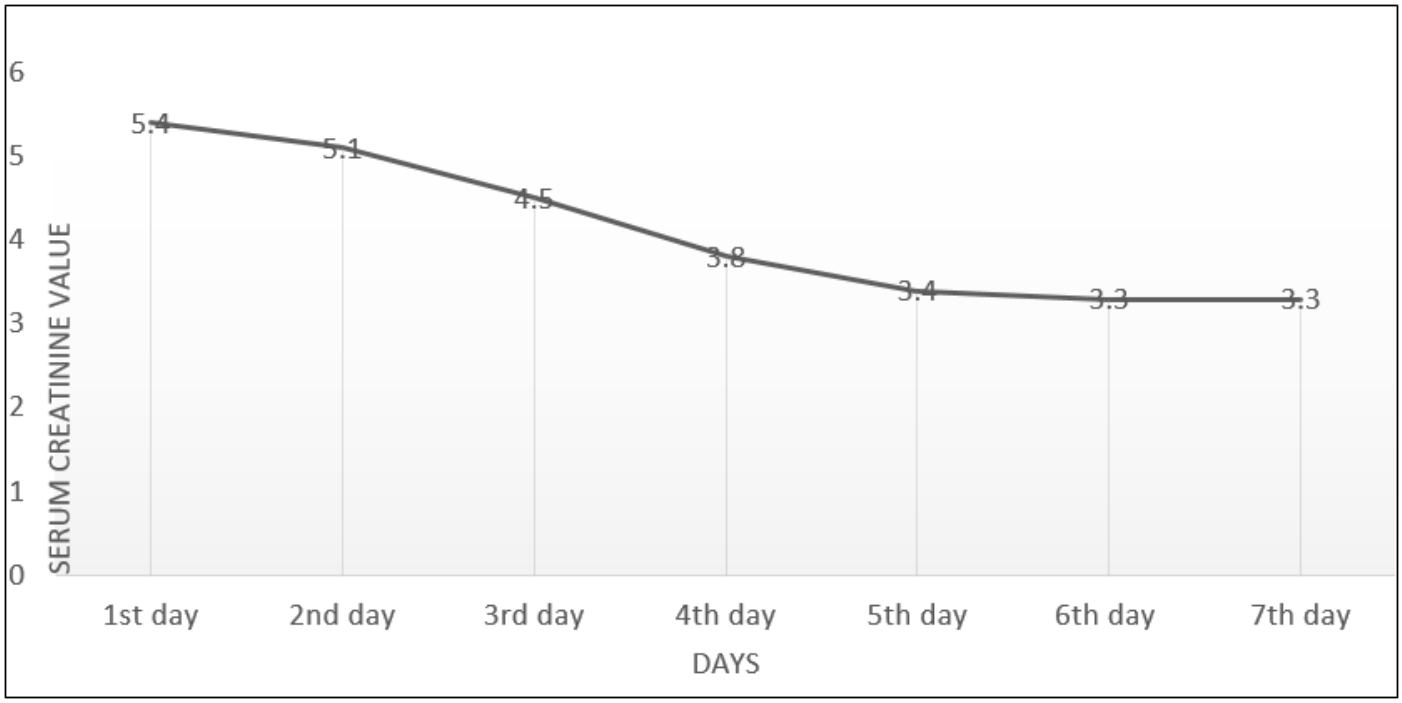

Fig 3 Serum Creatinine Trend—Change in Serum Creatinine levels after Starting Treatment with Glucocorticoids and ACE-I

III. DISCUSSION

IgA vasculitis (IgAV), or Henoch-Schönlein purpura, is a small vessel vasculitis characterized by IgA deposition primarily affecting the skin, joints, gastrointestinal tract, and kidneys [8]. Although more common in children, adult-onset IgAV presents distinct challenges, particularly regarding kidney involvement, which is often more severe and carries a worse prognosis compared to pediatric cases [6]. The kidney manifestations resemble those seen in IgA nephropathy, suggesting a shared pathogenesis. Elevated galactose-deficient IgA1 (Gd-IgA1) levels in both conditions indicate a common underlying mechanism, potentially triggered by environmental factors such as infections [10].

In this case, the patient presented with classical features of IgA vasculitis, including palpable purpura, arthralgia, abdominal pain, and hematuria, along with significant renal involvement characterized by elevated serum creatinine, nephrotic-range proteinuria, and hypertension. This constellation of findings underscores the severity of kidney involvement, a common and concerning complication in adult-onset IgAV. While kidney involvement (IgAV nephritis) typically presents as microscopic or macroscopic glomerular hematuria with or without red blood cell casts and mild to moderate proteinuria, a minority of patients—particularly adults—develop more severe manifestations, including nephrotic-range proteinuria, elevated serum creatinine, and hypertension, which are associated with a worse kidney prognosis [11,12]. Unlike in children, where IgAV nephritis is generally mild, adults are more prone to moderate to severe disease, emphasizing the need for vigilant monitoring and early intervention to prevent long-term complications.

The diagnosis of IgAV nephritis is primarily based on clinical presentation. In children, the development of hematuria, proteinuria, and elevated serum creatinine associated with palpable purpura, abdominal pain, and arthritis/arthralgia is often so characteristic that no further investigation is needed. However, in adults, especially when the presentation is atypical or severe, further confirmation through skin biopsy may be warranted to solidify the diagnosis and guide management [5].

Our patient was treated with a combination of glucocorticoids and an ACE inhibitor, aligning with current recommendations for managing significant proteinuria and

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impaired renal function in IgAV nephritis [13,14]. In adults with IgAV nephritis, the treatment approach is largely influenced by the severity of kidney disease at presentation and the potential for progression to kidney failure [14]. For patients with more severe kidney involvement—such as our patient, who had nephrotic-range proteinuria and elevated serum creatinine—immunosuppressive therapy with glucocorticoids is generally recommended [14]. The patient was administered pulse IV methylprednisolone followed by oral prednisone to reduce inflammation and prevent further renal deterioration. Adding an ACE inhibitor was critical for reducing proteinuria and protecting kidney function. Regularly monitoring urine protein excretion and serum creatinine is essential in these cases to assess treatment efficacy and detect any signs of disease progression early [14]. However, it is important to acknowledge that not all patients respond to glucocorticoids, and alternative immunosuppressive therapies may be necessary. For patients who are resistant to glucocorticoids or have contraindications, agents like mycophenolate mofetil, rituximab, or cyclosporine have shown promise in reducing proteinuria and stabilizing renal function. However, their use is supported more by observational studies and case series than by large, randomized trials [15,16].

The long-term kidney prognosis in IgAV nephritis is generally worse in adults than in children, possibly due to concurrent chronic kidney disease (CKD) or a longer duration between disease onset and clinical presentation [4]. A meta-analysis of studies evaluating risk factors for CKD in IgAV nephritis found that older age at disease onset, low glomerular filtration rate (GFR), and an initial presentation with nephrotic syndrome or nephritic-nephrotic syndrome are associated with unfavorable kidney outcomes [17]. In adults, reported rates of end-stage kidney disease (ESKD) are significantly higher than in children, ranging from 10 to 30 percent at 15 years [8,18,19].

Given the severity of IgAV nephritis in adults, as evidenced by our patient's presentation, it is imperative to tailor the treatment approach based on individual risk factors and disease severity. Ongoing research is needed to optimize treatment strategies, particularly for those with severe or refractory disease, to improve long-term outcomes in this population.

IV. CONCLUSION

This case report highlights the clinical presentation and management of adult-onset IgA vasculitis, a condition often characterized by severe renal involvement. While treatment with glucocorticoids and an ACE inhibitor can be effective, the long-term kidney prognosis remains challenging in adults. The risk of developing end-stage kidney disease is significantly higher compared to children, emphasizing the need for early diagnosis, tailored management, and ongoing research to improve outcomes.

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