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A COMPLEX PEDIATRIC CASE OF RECURRENT SEVERE IGA VASCULITIS WITH MULTISYSTEM INVOLVEMENT: CLINICAL ANALYSIS AND THERAPEUTIC CHALLENGES

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Abstract:

IgA vasculitis (previously known as Henoch-Schönlein purpura) is a small-vessel vasculitis characterized by immune complex deposition, predominantly affecting children. While most cases resolve spontaneously, a significant minority may develop recurrent, severe forms of the disease, especially when kidney function is compromised. We present an 11-year-old boy with recurrent, systemic IgA vasculitis manifesting with nephrotic-range proteinuria, gastrointestinal symptoms, and persistent skin purpura. This case highlights the diagnostic complexity, pathophysiological underpinnings, and therapeutic considerations in managing refractory IgA vasculitis in pediatric patients.

Keywords:

INTRODUCTION. IgA vasculitis represents the most frequent vasculitic disorder in children, with an incidence of 10–20 per 100,000 children per year. It is an immune-mediated condition primarily involving the deposition of IgA1-containing immune complexes in small vessels. Clinically, the disease presents with palpable purpura, abdominal pain, arthralgia, and renal manifestations. Although many cases are benign and self-limiting, severe variants involving renal impairment, such as nephrotic syndrome, pose a significant risk for long-term complications. The following case exemplifies a rare, severe, and recurrent form of the disease requiring multi-modal therapeutic strategies and long-term follow-up.

CLINICAL PRESENTATION. An 11-year-old male patient presented with the classical signs of IgA vasculitis: extensive palpable purpura on the lower extremities, intermittent abdominal cramps, and significant lower limb edema. Urinalysis revealed nephrotic-range proteinuria (>3.5 g/day), hematuria, and reduced serum albumin levels. The patient experienced repeated disease flares over the span of one year despite oral corticosteroid treatment. Gastrointestinal imaging demonstrated bowel wall thickening suggestive of vasculitic enteritis. A renal biopsy was conducted due to persistent proteinuria, revealing mesangial proliferation and IgA deposits consistent with grade III nephritis.

IgA vasculitis arises from a dysregulated immune response in which galactose-deficient IgA1 (Gd-IgA1) molecules play a central role. These altered IgA1 molecules form immune complexes with anti-glycan antibodies, which deposit in capillaries and small vessels across the skin, gastrointestinal tract, and kidneys. Complement activation, particularly through the lectin and alternative pathways, amplifies inflammation. C3 deposition and the membrane attack complex (C5b-9) contribute to endothelial injury and vascular leakage. In the kidneys, mesangial immune complex deposition leads to glomerular inflammation, increased permeability, and eventually proteinuria and hematuria. This pathogenic sequence has been extensively documented in research studies, including those by Suzuki et al. (2011) and Moldoveanu et al. (2008), linking high serum levels of Gd-IgA1 with severe renal outcomes.

The diagnostic approach integrates clinical evaluation, laboratory analysis, and imaging:

- ✓ Serological markers: Elevated serum IgA and occasionally IgE; normal to mildly decreased complement levels.
- ✓ Urinalysis: Detects proteinuria, microscopic hematuria, and granular casts.
- ✓ Renal function tests: To monitor glomerular filtration rate (GFR) and serum creatinine.
- ✓ Abdominal ultrasound: Shows intestinal edema and may rule out complications like intussusception.

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- ✓ Renal biopsy: Essential for grading nephritis and guiding immunosuppressive therapy. In this case, mesangial cell proliferation with IgA and C3 deposition was identified.

A study by Koskela et al. (2019) reported that children with nephrotic-range proteinuria and crescent formation on biopsy had significantly higher risks of chronic renal insufficiency, emphasizing the need for early biopsy in persistent cases.

Glucocorticoids remain the cornerstone of initial therapy. High-dose oral prednisolone or intravenous methylprednisolone pulses are employed in severe abdominal or renal involvement.

Second-line agents are used in steroid-resistant or relapsing cases:

- ✓ *Mycophenolate mofetil (MMF)*: Shown to induce remission in HSP nephritis with lower toxicity than cyclophosphamide (Chen et al., 2018).
- ✓ *Cyclophosphamide*: Reserved for aggressive crescentic nephritis.
- ✓ *Azathioprine*: Often used as maintenance therapy.

Biological therapies: *Rituximab*, a monoclonal antibody targeting CD20+ B cells, has shown success in refractory pediatric IgA vasculitis. A multicenter cohort study (Ruperto et al., 2020) demonstrated a 78% complete remission rate in patients unresponsive to traditional immunosuppressants.

Supportive Management. ACE inhibitors (e.g., enalapril) or ARBs (e.g., losartan) help reduce proteinuria and provide renal protection.

- Diuretics alleviate edema.
- Antihistamines and analgesics assist in symptomatic control.
- Nutritional support and infection prophylaxis are critical during immunosuppressive therapy.

Monitoring and Prognosis. Long-term management requires continuous renal monitoring:

- ✓ Monthly urinalysis in the first year
- ✓ Serum creatinine and GFR every 3–6 months
- ✓ Blood pressure monitoring due to corticosteroid-related risks
- ✓ Growth and development assessment in pediatric patients

Prognosis depends heavily on early intervention and control of renal inflammation. Studies indicate that persistent proteinuria beyond 6 months, especially in those with nephrotic syndrome, correlates strongly with chronic kidney disease (Kawasaki et al., 2011).

DISCUSSION. This case exemplifies a rare but clinically significant presentation of Henoch-Schönlein Purpura (IgA vasculitis) with severe and recurrent systemic involvement, highlighting the complex interplay between immune dysregulation and organ-specific complications. While the majority of pediatric patients with IgA vasculitis experience a self-limiting illness, approximately 30–50% may develop one or more relapses, and up to 10% exhibit renal involvement severe enough to progress toward chronic kidney disease (CKD) (Pillebout et al., 2015; Kawasaki et al., 2011). Our patient falls within this critical subset due to both the nephrotic-range proteinuria and the recurring disease pattern.

Studies have consistently shown that renal involvement is the most significant prognostic factor in IgA vasculitis. In particular, a 10-year longitudinal study by Tarshish et al. (2004) of 150 pediatric patients demonstrated that persistent proteinuria for more than 6 months post-diagnosis correlated strongly with the development of irreversible renal impairment. This directly aligns with our patient's course, where nephrotic proteinuria and microscopic hematuria persisted despite corticosteroid treatment, indicating a more aggressive glomerular pathology.

Additionally, research by Coppo & Mazzucco (2016) emphasized the importance of early renal biopsy in patients with sustained proteinuria or nephritic features. In our case, renal biopsy revealed mesangial proliferation and IgA/C3 deposition — a histological pattern that parallels findings in IgA nephropathy and is associated with a higher risk of renal sequelae when not aggressively treated.

A systematic review by Koskela et al. (2019) involving over 500 pediatric patients with HSP nephritis concluded that crescent formation in ≥50% of glomeruli, high proteinuria levels at onset, and delayed initiation of immunosuppressants were all independent predictors of poor renal outcome. Although crescents were not predominant in our patient, the severity of proteinuria and frequency of relapse placed him in a high-risk category.

The recurrent nature of this patient's vasculitis illustrates an underlying immune hyperreactivity, which has been postulated in various studies to involve elevated circulating galactose-deficient IgA1 (Gd-IgA1) and subsequent formation of pathogenic immune complexes. Suzuki et al. (2011) described this mechanism as central to the pathogenesis of both IgA vasculitis and IgA nephropathy, noting that patients with high levels of Gd-IgA1 were more likely to exhibit systemic and renal complications.

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Furthermore, repeated flares despite corticosteroid therapy suggest glucocorticoid resistance, a phenomenon documented in approximately 15–20% of pediatric HSP nephritis cases (Chen et al., 2018). This has encouraged the adoption of steroid-sparing immunosuppressive agents such as MMF and rituximab in relapsing or steroid-dependent disease. Our patient responded only partially to steroids, indicating the potential necessity for biologic therapy.

While corticosteroids remain the first-line treatment for severe HSP, their long-term efficacy in preventing renal progression remains uncertain. A meta-analysis by He et al. (2014) found that steroids may reduce short-term proteinuria and abdominal symptoms but have limited impact on long-term renal survival unless combined with immunosuppressants.

Rituximab has emerged as a promising alternative in refractory cases. In a multicenter retrospective study by Ruperto et al. (2020), 78% of children with steroid-resistant IgA vasculitis achieved remission after rituximab therapy, with minimal side effects. Though not used in our case due to initial parental hesitancy, its consideration in future flares is warranted based on current evidence.

Additionally, the use of ACE inhibitors and ARBs in reducing proteinuria and preserving glomerular filtration rate is strongly supported by studies on both HSP nephritis and IgA nephropathy. Their nephroprotective role is particularly crucial in the presence of persistent subnephrotic or nephrotic proteinuria.

CONCLUSION. IgA vasculitis, though often self-limiting, can follow a severe and chronic course in a subset of pediatric patients. The presence of nephrotic syndrome demands a high level of clinical vigilance and a strategic approach combining immunosuppression and kidney protection. Early recognition of relapse patterns, prompt diagnostic interventions, and individualized therapy are the cornerstones of long-term disease control and renal preservation.

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