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IgA Vasculitis – An In-Depth Literature Review

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Abstract

Introduction and Objective: Immunoglobulin A vasculitis (IgAV), previously Henoch-Schönlein purpura, is a systemic leukocytoclastic vasculitis of small vessels, characterised by the deposition of immunoglobulin A complexes in their walls. It represents the most prevalent form of vasculitis in children and may progress to IgA nephropathy, the leading cause of primary glomerulonephritis. This review provides a comprehensive overview of IgAV, covering its epidemiology, pathogenesis, clinical features, diagnosis, treatment, complications, and prognosis.

Materials and Methods: A narrative review was conducted, using PubMed articles up to 2024, focusing on recent data. Key sources included clinical guidelines (EULAR/PRINTO/PRES 2010 for paediatrics, ACR 1990 for adults) and contemporary studies on genetic, immunological, and therapeutic aspects. Data were synthesised to compare disease presentation and management in children and adults.

Results and Conclusions: IgAV predominantly affects children aged 3–15 years (incidence 3.0–27.0 per 100,000), often triggered by upper respiratory tract infections. In adults, the condition is rarer but more severe. Diagnosis relies on clinical criteria, with biopsy reserved for atypical cases. Treatment is primarily symptomatic, employing glucocorticoids for acute

symptoms and immunosuppressive agents for severe cases. Complications, such as gastrointestinal and renal disorders, are more frequent in adults. Prognosis is favourable, with 94% of paediatric cases resolving within two years, although adults are at greater risk of chronic kidney disease and relapses. Early intervention and monitoring are critical for improving outcomes.

Keywords: Immunoglobulin A vasculitis, Henoch-Schönlein purpura, leukocytoclastic vasculitis, IgA nephropathy, paediatric vasculitis, adult vasculitis, glomerulonephritis, glucocorticoids, immunosuppression

Introduction

Immunoglobulin A vasculitis (IgAV), previously known as Henoch-Schönlein purpura (HSP), is defined as a systemic, leukocytoclastic vasculitis affecting small blood vessels. It is characterised by the deposition of immunoglobulin A (IgA) immune complexes within the vessel walls [2,4]. IgAV represents the most common form of vasculitis in the paediatric population, while IgA nephropathy is the leading cause of primary glomerulonephritis [2,4,9,16,31,32,33]. In children, the condition frequently manifests following an upper respiratory tract infection [2]. The hallmark clinical features include palpable purpura, which is not associated with thrombocytopenia and predominantly affects the posterior surfaces of the body [1]. This is often accompanied by abdominal pain, arthralgia, or arthritis (most commonly involving the knees, ankles, wrists, and fingers, with spinal or shoulder joint involvement being less frequent) [1]. In some cases, gastrointestinal bleeding or glomerulonephritis may occur. Rarely, the disease can involve other internal organs, such as the lungs or the central nervous system [16,33]. The clinical presentation of IgAV most frequently constitutes a combination of symptoms pertaining to multiple organ systems; however, the disease may be restricted exclusively to the skin or to the kidneys (IgA nephropathy) [4, 33]. Although IgAV typically follows a self-limiting course, recurrences are observed in certain instances [1,2,31,33].

History of the Disease

The history of immunoglobulin A vasculitis (IgAV) spans over two centuries [29]. The first documented case of HSP was described in 1802 by William Heberden, who reported a 5-year-old boy presenting with a characteristic constellation of symptoms, including joint swelling and arthritis, macroscopic haematuria, abdominal pain, tarry stools, and a red rash [29,30]. The disease is named after two researchers who conducted detailed analyses of its clinical features: Johann Schönlein and his student Eduard Henoch. In 1837, Schönlein discovered the connection between inflammation and joint pain with cutaneous purpura ("purpura rubra"). His student, Henoch, in 1838 reported cases of children with purpura, abdominal pain, bloody diarrhea, and joint pain [33]. He described the observed cases in 1886 [1,29,30,33]. Henoch emphasized the systemic effects of the condition, which was initially named Henoch-Schönlein purpura [33].

The direct link between HSP and vascular involvement was established by Gaidner in 1948, followed by subsequent descriptions of the critical role of immunoglobulin A in the pathogenesis of the disease [30].

Epidemiology

The majority of cases of IgA vasculitis (IgAV) occur in children aged 3 to 15 years, statistically more frequently in boys, with a ratio of 1.5:1 [2,3]. Between 75-90% of reported cases in children occur below the age of 10, with the highest recorded incidence of 70.3/100,000 individuals in the age range between 4 and 7 years. This appears to be related to the fact that this age group is the most susceptible to infections [33]. In the pediatric population, the annual incidence is estimated at 3.0 - 27.0 cases of HSP/100,000 children. This is approximately 30 times more frequent than in the adult population [33]. In children, this disease entity exhibits seasonality—it is more frequently observed in autumn and winter [1,2]. The observed phenomenon is likely related to greater exposure to infections during the mentioned seasons [33]. No such correlation has been observed in adults to date. The number of new cases statistically remains at a similar level over the years, with a slight upward trend. However, the actual incidence appears to be underestimated because, in the case of children with only cutaneous purpura, hospitalization is not required, resulting in a lower number of

reported cases [33]. Nearly 50% of IgA vasculitis cases are preceded by an upper respiratory tract infection of streptococcal etiology (Streptococcus spp.). Other frequently reported pathogens include the hepatitis A virus, Helicobacter pylori, parvovirus B19, and Mycoplasma pneumoniae [2,9,33]. A similar correlation has also been noted with gastrointestinal infections, connective tissue inflammations, and urinary tract infections [1,33]. A significant decrease in the number of cases was observed in connection with the COVID-19 pandemic. Most likely, due to restrictions introduced by the governments of many countries, such as mandatory wearing of protective masks or the necessity of quarantine, the occurrence of viruses and other pathogens affecting the respiratory system decreased [33,34]. Among other risk factors for developing HSP, vaccines, medications, insect venoms, and infections are also mentioned [1,33]. A 3-4 times higher tendency to develop HSP has been observed in children of Caucasian and Asian races compared to those of Black race [33].

Pathogenesis

The etiology of IgAV remains incompletely elucidated, involving complex interplay of immunological, environmental, and genetic factors. Although numerous infectious and chemical triggers have been identified, the mechanisms by which they elicit an immune response leading to IgA immune complex deposition in small vessel walls are not fully understood. It has been observed that approximately 50% of cases of IgA vasculitis are associated with a prior upper respiratory tract infection (URTI) [33]. Common pathogens associated with URTI include beta-haemolytic Streptococcus group A, parainfluenza virus, parvovirus B19, adenovirus, and herpesvirus [2,4,17].

Helicobacter pylori infection is also implicated as a risk factor, with eradication therapy significantly improving disease outcomes [4,33]. Despite the demonstrated association in studies between vaccinations and the subsequent development of IgA vasculitis, the risk has been determined to be low and outweighed by the risk of not vaccinating children against childhood diseases [33]. Biological drugs and targeted therapies are also mentioned as possible risk factors for the development of IgAV [33]. A significant disease associated with IgA vasculitis is familial Mediterranean fever, in which IgAV symptoms most commonly affect the gastrointestinal tract. Environmental factors, such as vaccines (notably MMR, hepatitis B, and SARS-CoV-2 vaccines) and SARS-CoV-2 infection itself, are recognised as potential triggers [4].

Genetic predisposition may increase susceptibility to IgAV, with associations noted for human leukocyte antigens (HLA), particularly HLA-DRB1*01 and HLA-B35. The presence of HLA-DRB1*03 and HLA-DRB1*07 antigens has a protective effect against the development of IgAV, whereas HLA-DRB1*01 and HLA-DRB1*11 increase the predisposition to the disease [33]. HLA-DR1*0103 is strongly linked to IgAV risk, while HLA-B35 is associated with IgA nephropathy [2,4,5,6,7].

In affected small vessels, elevated levels of IgA and complement component C3 are observed, with a predominance of galactose-deficient IgA1 (Gd-IgA1) [17]. The reason for the exclusive involvement of IgA1, but not IgA2, in pathogenesis remains unclear [1]. Normally, intact IgA is cleared by hepatocytes in the liver, but Gd-IgA1 complexes evade this mechanism, depositing in small vessel walls and renal mesangium.

Additionally, patients with IgAV exhibit elevated serum levels of galactose-deficient IgA1, IgA-class anticardiolipin antibodies, transforming growth factor-beta (TGF-β), and circulating IgA immune complexes. These complexes form in response to antigenic exposure, such as infection, and deposit in the walls of small vessels, particularly capillaries in the skin, joints, kidneys, or gastrointestinal tract. This may activate the complement system, amplifying the inflammatory response. Immune complex deposition in cutaneous capillaries results in palpable purpura and petechiae, while gastrointestinal deposits may lead to bleeding [16].

Clinical Manifestations in Children

In the paediatric population, the majority of IgAV cases are characterised by a mild, self-limiting course, distinguishing HSP from other systemic vasculitides [8,9,10]. The disease typically presents acutely, with initial symptoms often preceded by a viral infection of the upper respiratory tract or gastrointestinal system occurring 1–2 weeks prior. The initial symptoms of IgAV appear in children within days or weeks [33]. The predominant feature is palpable purpura, primarily localised to the lower extremities and less commonly affecting the trunk or upper limbs [9,10]. This purpura is not associated with thrombocytopenia or a bleeding diathesis. In addition to purpura, the three most common manifestations include arthritis or arthralgia, renal involvement with proteinuria exceeding 0.3 g/day, and abdominal pain, which is often diffuse, colicky, and detectable on physical examination. Abdominal pain may be accompanied by gastrointestinal bleeding [3,8,9,10]. Up to 50% of cases in the pediatric population have mild gastrointestinal symptoms (abdominal pain, nausea, vomiting)

[33]. IgA nephropathy occurs with a frequency of 20-54% among cases in children with HSP [33]. In children under 2 years of age, subcutaneous oedema is frequently observed in distal parts of the body, such as the hands, scalp, or ears [3,8,9]. Kidney damage caused by the disease is the most significant prognostic factor, worsening the prognosis [9,31]. 20-80% of children with IgAV exhibit symptoms of kidney damage—hematuria and/or proteinuria—and 1%–7% may develop kidney failure or even its end-stage [31]. Renal involvement is the most critical prognostic factor, significantly worsening the outlook [9]. Approximately 94% of paediatric patients achieve complete recovery within 2 years, with most symptoms resolving within 30 days [8,9,10].

Clinical Manifestations in Adults

IgAV is considerably less common in adults than in children, with a peak incidence around the age of 50 [8,9,10,11]. The clinical presentation is similar to that in the paediatric population [12]. However, abdominal pain is less frequent in adults [12]. Notably, IgA nephropathy tends to be more severe and manifests earlier in the disease course in adults. Renal involvement in adults is most commonly indicated by haematuria and proteinuria. Unlike in children, approximately 75% of adult cases present with systemic symptoms and organ damage [8,9]. These may result from oedema and/or ischaemia, leading to gastrointestinal manifestations such as bloody diarrhoea, nausea, vomiting, and severe pain. Similar mechanisms may affect other organs, frequently resulting in inflammation of the lungs, scrotum, testes, or bladder [8]. In rare cases, complications such as intracranial haemorrhage, focal neurological deficits, or ischaemic stroke may occur [9,14,15]. Adults are also at higher risk of end-stage renal disease, disease recurrence, and complications [8,9,10,12]. In individuals with glomerular involvement observed in a kidney biopsy ≥50%, the likelihood of progression to chronic kidney disease ranges between 5% and 20% [31].

Diagnosis

Diagnosis of IgAV is primarily based on characteristic clinical features. In cases of atypical cutaneous lesions or ambiguous symptoms, a skin biopsy may be performed [1]. No diagnostic tests are required to confirm the diagnosis [2]. Current diagnostic criteria for IgAV in children are based on the EULAR/PRINTO/PRES 2010 guidelines, while the American

College of Rheumatology (ACR) 1990 criteria are used for adults [2]. The paediatric criteria demonstrate 100% sensitivity and 87% specificity, while the adult criteria offer 99% sensitivity and 86% specificity [2,8,13]. According to EULAR/PRINTO/PRES, a mandatory criterion is palpable purpura or petechiae, unrelated to thrombocytopenia, typically on the lower limbs, plus at least one of the following: acute-onset abdominal pain, arthritis or arthralgia (commonly of the knees or ankles), renal involvement, or histopathological evidence of leukocytoclastic vasculitis with predominant IgA deposits in skin or glomerular IgA deposits in renal biopsy [17]. The ACR criteria for adults include age under 20 years, palpable purpura, abdominal pain, and biopsy evidence of granulocytic infiltration in small vessel walls, with 87.1% sensitivity and 87.7% specificity [13].

Laboratory tests serve a supportive role in diagnosis and monitoring. Elevated serum IgA levels are observed in 50–70% of patients [1,33]. Blood counts may show normal platelet levels, leukocytosis, and, in cases of gastrointestinal bleeding, normochromic anaemia. Inflammatory markers (CRP, ESR) may be elevated, particularly if infection triggered the disease, or normal. Renal involvement may manifest as elevated serum creatinine, haematuria, or proteinuria in urinalysis. Imaging studies, such as abdominal ultrasound or CT, are performed to evaluate suspected intussusception, obstruction, or perforation in patients with abdominal symptoms. Renal biopsy is indicated for nephritic or nephrotic syndrome, massive proteinuria, or acute renal failure. The differential diagnosis of immunoglobulin A vasculitis is notably broad due to the disease's propensity to cause multisystem complications, which can mimic a variety of other conditions. It should encompass disorders presenting with cutaneous petechiae, such as meningococcal sepsis (a life-threatening emergency), drug reactions, thrombocytopenia, and haemolytic uraemic syndrome [29]. Additionally, HSP must be differentiated from septic arthritis, bacterial endocarditis, polyarteritis nodosa, cryoglobulinaemia, granulomatosis with polyangiitis, Goodpasture's syndrome, and autoimmune diseases (e.g., systemic lupus erythematosus) [1,17,29]. In addition to these systemic conditions, severe abdominal symptoms in IgAV, such as colicky pain or gastrointestinal bleeding, may closely resemble those seen in inflammatory bowel diseases, notably Crohn's disease [29].

Treatment

The selection of appropriate treatment is challenging due to the complexity of symptoms and the diverse organ manifestations of the disease [8,9]. In the majority of cases, symptomatic treatment is sufficient, given the typically mild disease course [18,21].

8.1. Glucocorticosteroids

Glucocorticosteroids (GCS) are the cornerstone of treatment, with oral prednisolone at a dose of 1 mg/kg/day demonstrating a good response in acute purpura [8,13,19,21,22]. Early initiation of GCS may benefit renal function [21,22]. However, their administration during the purpuric phase does not prevent the development of IgA nephropathy [22]. GCS are also effective in managing gastrointestinal and joint inflammation [21]. Nevertheless, their use as a standardised treatment remains controversial due to the limited reliability of studies evaluating their efficacy [20,21,22].

8.2. Non-Steroidal Anti-Inflammatory Drugs

Non-steroidal anti-inflammatory drugs (NSAIDs) have been beneficial in managing joint pain and arthritis but have no effect on cutaneous symptoms [21,22]. Their use is contraindicated in cases of renal or gastrointestinal involvement, which are among the most common organ manifestations [8,9,18,19,21].

8.3. Colchicine and Dapsone

Colchicine, primarily used in acute gout attacks, exerts anti-inflammatory effects by inhibiting neutrophil activity [21,22]. Although clinical trials have not demonstrated its efficacy in HSP, case reports describe successful treatment of recurrent cutaneous purpura [21,22]. Similarly, dapsone, which inhibits inflammatory mediators such as IL-8, reactive oxygen species, and TNF, may be effective in treating refractory purpura [8,21,22].

8.4. Immunosuppressive Therapy and Plasmapheresis

In severe disease forms, immunosuppressive therapy may be employed. Additionally, plasmapheresis and intravenous immunoglobulins have shown benefits in advanced disease stages [19,20,21,22]. Plasmapheresis is used in both adults and children with HSP who

experience significant, rapid deterioration of renal function or crescentic glomerulonephritis on renal biopsy. Some studies report the use of plasmapheresis in paediatric patients with severe gastrointestinal involvement or disease recurrence in transplanted kidneys [1,28].

8.5. ACE Inhibitors and ARBs

Angiotensin-converting enzyme inhibitors (ACEIs) or angiotensin receptor blockers (ARBs), typically used in the treatment of arterial hypertension, are also employed in cases where the kidneys are affected by a pathological process [21,22]. Their beneficial effects in reducing proteinuria and improving glomerular filtration parameters have been demonstrated, thereby contributing to better long-term prognosis [21,24,31]. The rationale for dose escalation to achieve a therapeutic effect has also been confirmed. Standard doses of losartan used in the treatment of hypertension range between 50 mg and 100 mg per day. In cases of severe IgA nephropathy, doses as high as 200 mg per day may be considered. However, this requires an individualized approach in each case and close monitoring [21,22,23,24]. Nevertheless, drugs from these groups do not exhibit anti-inflammatory activity [22].

8.6. Rituximab

Rituximab, a monoclonal antibody targeting the CD20 antigen on B lymphocytes, may inhibit the production of pathological IgA antibodies [18,20]. It is used in recurrent or treatment-refractory disease [18,19]. Its mechanism of action is not fully understood [19,20], but studies suggest it halts disease progression and promotes remission [18,19,20,22].

Complications

Among adult patients, the most prevalent category of complications associated with Henoch-Schönlein purpura, pertains to gastrointestinal manifestations. These complications are reported to affect up to 44% of individuals diagnosed with this condition [26]. The spectrum of gastrointestinal sequelae includes, but is not limited to, intestinal perforation, intussusception, and intestinal necrosis. In contrast, severe abdominal complications occur in only 5% of children [17,26]. Paediatric patients are more likely to experience genitourinary complications, such as orchitis in 10% of boys [26]. Avascular necrosis of the femoral head,

presenting as hip pain, is another reported complication, potentially linked to prolonged glucocorticoid therapy or direct immune complex-mediated injury [26,27].

Prognosis

IgAV is typically self-limiting or responsive to treatment, with mild to moderate cases carrying a favourable prognosis. Persistent purpura, age over 10 years, severe gastrointestinal symptoms, and relapses are risk factors for glomerulonephritis in IgAV [17]. In adults, onset after age 50, particularly in men, is associated with poorer outcomes, with gastrointestinal symptoms serving as the strongest predictor of relapse [2,26]. In children, risk factors for relapse include arthritis or arthralgia, abdominal symptoms, and glucocorticoid use during the initial episode [17,25]. Relapses occur in 2–30% of paediatric cases, most commonly as cutaneous manifestations, and may occur up to 10 years after the initial episode [2]. IgA vasculitis (IgAV) in elderly patients is characterized by a significantly worse prognosis, with the risk of developing end-stage renal disease reaching approximately 32% [31]. In contrast, among children with IgA nephropathy, the risk is considerably lower, ranging from 1% to 7% [31]. In recurrent IgAV and in adult patients without defined disease etiology malignancy should be excluded [8,17,26]. Patients with nephropathy require ongoing outpatient monitoring, including regular urinalysis and blood pressure measurements.

Summary

Immunoglobulin A vasculitis (IgAV, Henoch-Schönlein purpura) is characterised by the deposition of IgA immune complexes in the small vessels of the skin and various internal organs. It primarily affects children, with rarer but more severe presentations in adults, associated with a higher risk of complications. The multifactorial pathogenesis of IgAV remains incompletely understood, necessitating a multifaceted approach to diagnosis and treatment. Early intervention and vigilant monitoring are essential to optimise outcomes in this complex vasculitic disorder.

Disclosure

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