VIA MEDICA

CASE REPORT

Forum Dermatologicum
DOI: 10.5603/fd.102135

ISSN 2451–1501, e-ISSN 2451–151X

IgA vasculitis in an adult patient

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ABSTRACT

Henoch–Schönlein purpura (HSP) is a form of small-vessel vasculitis characterized by the deposition of immunoglobulin A (IgA) in vascular walls. It predominantly affects children, often following a respiratory tract infection, and is marked by a classic tetrad of symptoms: palpable purpura, arthritis, abdominal pain, and haematuria. Though typically a paediatric condition, HSP can also occur in adults, where it may present with similar but sometimes more severe manifestations.

This article presents a case of IgA vasculitis with renal involvement in an adult patient treated with dapsone. Henoch–Schönlein purpura in adults presents unique challenges and requires a comprehensive, interdisciplinary approach to management. The absence of standardized treatment guidelines for adults highlights the need for further research and development of clear protocols. Collaboration among specialists in dermatology, nephrology, and other relevant fields is essential for optimizing patient care and outcomes in managing this complex condition.

Forum Derm.

Keywords: vasculitis, purpura, IgA vasculitis, dapsone, IgA nephropathy

INTRODUCTION

Henoch–Schönlein purpura (HSP) was classified by the International Chapel Hill Consensus Conference 2012 as a small-vessel vasculitis associated with immune complexes [1]. In this condition, immune complexes, primarily IgA, deposit in the walls of blood vessels in the skin and internal organs. While 90% of cases occur in children under the age of 10, the prevalence in adults is much lower, ranging from 8 to 18 cases per million annually [2]. The most characteristic feature is skin involvement, presenting mainly as palpable purpura. Additionally, systemic symptoms often include abdominal pain, gastrointestinal bleeding, arthritis, and nephritis [3].

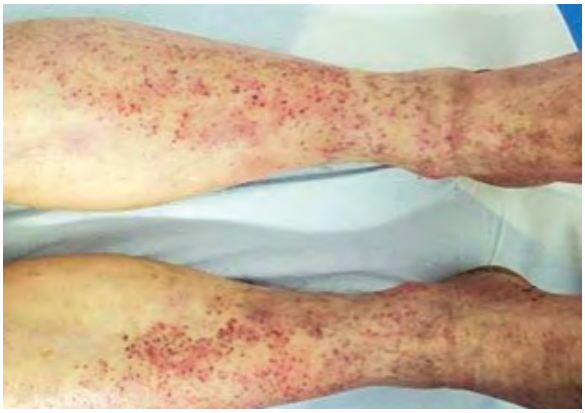

Figure 1. Palpable purpura on the patient's lower legs

CASE REPORT

A 50-year-old female patient was admitted to the department of dermatology due to palpable purpura that had been developing for several days and involved upper legs, lower legs and skin of the abdomen (Fig. 1). The diascopy sign was positive. The eruption of lesions was preceded by a respiratory tract infection. The patient denied symptoms of other systems, including abdominal pain and arthralgia. Laboratory tests revealed elevated marker of inflammation [C-reactive protein (CRP) was 2.8 mg/dL, white blood cells (WBC) was 14.4 × 10^3/μL, neutrophils (NEU) was 12.7 × 10^3/μL]. A general urine test showed *haematuria*, the presence of proteins as well as the presence of leached erythrocytes. The 24-hour proteinuria was 2.18 g abdominal ultrasound did not show abnormalities. Antibodies perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA), cytoplasmic anti-neutrophil cytoplasmic antibodies (c-ANCA) and antinuclear antibodies (ANA) were absent in the serum. Due to the significant progression of the disease, a skin

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Received: 26.08.2024 Accepted: 7.11.2024 Early publication date: 22.11.2024

Table 1. Classification criteria for IgA-related vasculitis [10, 11]

ACR classification criteria (1990)	EULAR/PRINTO/PRES classification criteria (2010)
Two of the following criteria: • age ≤ 20 at disease onset • palpable purpura • acute abdominal pain • biopsy showing granulocytes in the walls of the small arterioles or venules	Purpura or petechia AND One of the following four criteria: • abdominal pain • arthritis or arthralgia • renal involvement • leukocytoclastic vasculitis with predominant IgA deposits or proliferative glomerulonephritis with predominant IgA deposits
Sensitivity 87.1%; specificity 87.7%	Sensitivity 100%; specificity 87%

ACR — American College of Rheumatology; EULAR/PRINTO/PRES — European League Against Rheumatism/Paediatric Rheumatology International Trials Organisation/Paediatric Rheumatology European Society; IgA — immunoglobulin A

biopsy was not performed. The suspicion of IgA vasculitis was established based on the clinical presentation. The patient has been referred to the nephrology department. A kidney biopsy was performed, with histopathological examination revealing an increase in mesangial cells and matrix, non-thickened capillary walls, and only minimal interstitial changes in the form of tubular atrophy and fibrosis. Testing for amyloidosis returned a negative result. Direct immunofluorescence (DIF) examination showed staining with anti-IgA sera (+++), C3 (+), and lambda light chains (++), with kappa light chains (+). No reactivity was observed with anti-IgG, IgM, C1q, or fibrinogen sera. Based on the clinical presentation, laboratory results and direct immunofluorescence findings a diagnosis of Henoch–Schönlein purpura was established. Treatment with dapsone, at 50 mg/day was initiated with good clinical response. Topical therapy was performed with mometasone furoate. After 6 months of therapy, no skin changes, abnormalities in laboratory tests, haematuria, or proteinuria have been observed in the patient.

DISCUSSION

IgA vasculitis (IgAV) is a systemic, non-infectious inflammatory disease affecting small vessels. It occurs much less frequently in adults compared to children and is associated with a higher risk of severe outcomes [4]. The main symptoms include purpura, joint pain and/or arthritis, gastrointestinal vasculitis, and glomerulonephritis. Diagnosis is primarily clinical, with possible consideration for a skin biopsy. Histopathological examination typically reveals leukocytoclastic vasculitis and direct immunofluorescence (DIF) shows IgA and complement C3 deposits [5, 6]. The exact causes of IgA vasculitis remain unknown, with infections and medications being well-documented triggers. A systematic review by Rasmussen et al. [7] identified antibiotics, particularly beta-lactams, vaccines (mainly for influenza and measles), TNF-alpha blockers, antihypertensives, anticoagulants, NSAIDs, and other analgesics as common triggers. Recent reports also suggest COVID-19 as a potential cause of Henoch–Schönlein purpura, with more severe outcomes in adults [8, 9]. The American College of Rheumatology (ACR) proposed classification criteria in 1990, updated in 2010 by EULAR, PRINTO, and PRES (Tab. 1) [10, 11].

Treatment of IgA vasculitis in adults remains debated due to the lack of correlation between initial renal function assessment and long-term outcomes, as well as the possibility of spontaneous remission in severe cases or progression to chronic kidney disease in mild cases [4]. Treatment options depend on the clinical presentation and may include colchicine, dapsone, corticosteroids, azathioprine, mycophenolate mofetil, cyclosporine A, cyclophosphamide, rituximab, and other immunosuppressives (intravenous immunoglobulins, plasmapheresis) [12]. Immunosuppressive therapy is often used for severe glomerulonephritis. Randomized trials have shown success with cyclosporine and mycophenolate mofetil in steroid-resistant kidney disease. Dapsone and rituximab also show good therapeutic effects in patients with skin and renal involvement [13]. The prognosis for chronic kidney disease development post-Henoch–Schönlein purpura is uncertain, with clinical studies showing varying rates of kidney failure and proteinuria [14, 15].

CONCLUSIONS

IgA vasculitis (formerly Henoch–Schönlein purpura) is a leukocytoclastic vasculitis with an immunological basis, characterised by IgA deposits in small vessel walls of the skin and internal organs. It necessitates an interdisciplinary approach and collaboration among various specialists. However, clear treatment guidelines for adults are still lacking.

Article information and declarations

Acknowledgements

None.

Author contributions

Each author made an equal contribution to the creation of this case report.

Conflict of interest

The authors declare no conflicts of interest.

Funding

There was no funding.

Ethics statement

Case report, consent of the bioethics committee is not required.

Supplementary material

None.

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