Annals of Case Reports
Djafer S, et al. Ann Case Rep: 10: 102257
www.doi.org/10.29011/2574-7754.102257
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Case Report

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Adult IgA Vasculitis (Henoch-Schönlein Purpura) with Cardiac Involvement: A Case Report and Review of the Literature

Salim Djafer*, Tristan Born, Armin Zgraggen, Samuel Rubeli, Sabine Adler

Department of Rheumatology and Immunology, Medical University Clinic Basel, Cantonal Hospital Aarau, Switzerland

***Corresponding author**: Salim Djafer, Department of Rheumatology and Immunology, Medical University Clinic Basel, Cantonal Hospital Aarau, Switzerland

Citation: Djafer S, Born T, Zgraggen A, Rubeli S, Adler S (2025) Adult IgA Vasculitis (Henoch-Schönlein Purpura) with Cardiac Involvement: A Case Report and Review of the Literature. Ann Case Report. 10: 2257. DOI:10.29011/2574-7754.102257

Received: 10 April 2025, **Accepted:** 15 April 2025, **Published:** 17 April 2025

Introduction

IgA Vasculitis, formerly known as Henoch-Schoenlein Purpura (HSP) represents a systemic immune complex-mediated vasculitis mainly targeting small blood vessels throughout various bodily regions. Predominantly observed in pediatric populations, this condition also manifests in adults, albeit less frequently. The disease is characterized by leukocytoclastic vasculitis foremost impacting the skin, kidneys, joints, and gastrointestinal tract by a pericapillary localized inflammation characterized by the accumulation of Immunoglobulin A (IgA) and complement component 3 (C3). In children, IgA vasculitis is usually responsive to steroid treatment only and generally follows a benign clinical course. The etiology of HSP remains unclear and is in general believed to be a combination of genetic susceptibility and exaggerated immune system response, triggered by various factors such as viral infections, medications, and environmental triggers [1].

The hallmark manifestation of HSP is palpable purpura, characterized by small, elevated, reddish-purple lesions on the skin. These lesions predominantly appear on the lower extremities, buttocks, and, to a lesser extent, on the upper extremities. Often, the purpura is preceded by symptoms including joint and abdominal pain, gastrointestinal hemorrhage, and renal involvement, as initially described during the 19th century by the German physician Johann Lukas Schönlein and one of his students, Eduard Henoch. Cardiac manifestations are exceedingly rare in pediatric and adult patients and typically arise from vasculitis affecting the coronary arteries and their smaller branches.

We detail a case of HSP with concurrent cardiac, renal, and gastrointestinal involvement. This work was complemented by a review of the literature on adult cases of HSP purpura with cardiac involvement.

Case Description

We report the case of a 54-year-old male with no prior history of cardiac or renal disease, whose prior pharmacotherapy was limited to Olmesartan 20 mg daily for the management of primary arterial hypertension.

In early May 2023, without any previous symptoms, the patient exhibited petechiae, predominantly on the lower extremities, alongside isolated necrotic foci. A skin biopsy conducted at the family physician's office demonstrated leukocytoclastic vasculitis and the physician prescribed topical betamethasone, which led to an initial amelioration of the dermatological symptoms.

However, three weeks after this initial presentation, the patient reported significant hematochezia, without accompanying abdominal pain, prompting him to seek treatment at our emergency department. Upon presentation, the patient was afebrile, with normal heart rate and blood pressure. Physical examination identified purpura on both legs, partially merging into ulcerative/necrotic areas on the distal aspect of the right lower leg,

accompanied by mild edema. Cardiac and pulmonary assessments were unremarkable. Laboratory analyses were within normal limits, including hemoglobin, leukocyte count, platelet count, creatinine, and electrolytes. Urinalysis revealed proteinuria, microhematuria, and nephritic sediment, indicating renal involvement. For the initial treatment, the patient received oral prednisolone starting at a dosage of 75 mg daily, alongside azathioprine 200 mg daily. A significant amelioration of the skin lesion was observed within the subsequent week and glucocorticoids were tapered.

Nevertheless, the patient sought care again at our emergency department ten days later, still taking 30mg of prednisone, presenting with a fever peaking at 39.4°C, chills, and hypotension. Clinical examination revealed pronounced edema in the lower legs, novel lesions on both forearms, and macroscopic hematuria. Consequently, the patient was admitted to the internal medicine department for comprehensive diagnostic evaluation and treatment. During admission, the patient experienced dyspnea and chest tightness. Radiographic examination revealed cardiac decompensation, affirmed by laboratory findings indicating pro-BNP level of 8738 ng/L (reference interval < 400 ng/L) and an elevated troponin concentration of 85 ng/L (reference interval < 0.04 ng/mL). Electrocardiography did not demonstrate acute myocardial infarction; however, echocardiography identified severe hypokinesia of multiple coronary flow areas spanning from inferoseptal to apical regions and inferior basal segments. Continuous 24-hour monitoring failed to detect significant cardiac arrhythmias, and the patient's dyspnea promptly improved following the administration of high-dose intravenous glucocorticoids and loop diuretics. Follow-up echocardiographic and coronary CT imaging revealed no remarkable pathology, demonstrating minimal non-stenotic coronary plaques with intact coronary anatomy. Also, a CT body scan did not reveal any underlying malignant disease. A renal biopsy was conducted during the patient's hospital stay, revealing IgA nephropathy characterized by cellular crescents in 5 out of 38 glomeruli, accompanied by a single instance of glomerulosclerosis and no arteriosclerosis.

In addition to the intravenous glucocorticoids 125mg/d, immunosuppressive therapy with cyclophosphamide was initiated and doses of 1.2g thrice at three-week intervals were administered. Additionally, methotrexate was started as renal function showed normal eGFR and continued up to date with 20mg subcutaneously once a week. In the weeks following discharge, the patient then went into remission and remained asymptomatic up to date. Glucocorticoids could be tapered to 5mg a day over 6 months and completely reduced over 12 months. During follow-up, cardiac magnetic resonance revealed no abnormalities (Figure 1,2 and Table 1).

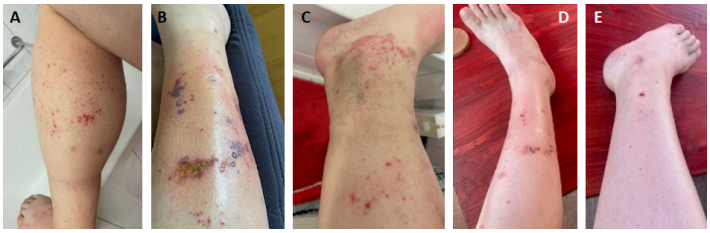

Figure 1: Skin lesions before (A-C) and after (D-E) treatment initiation.

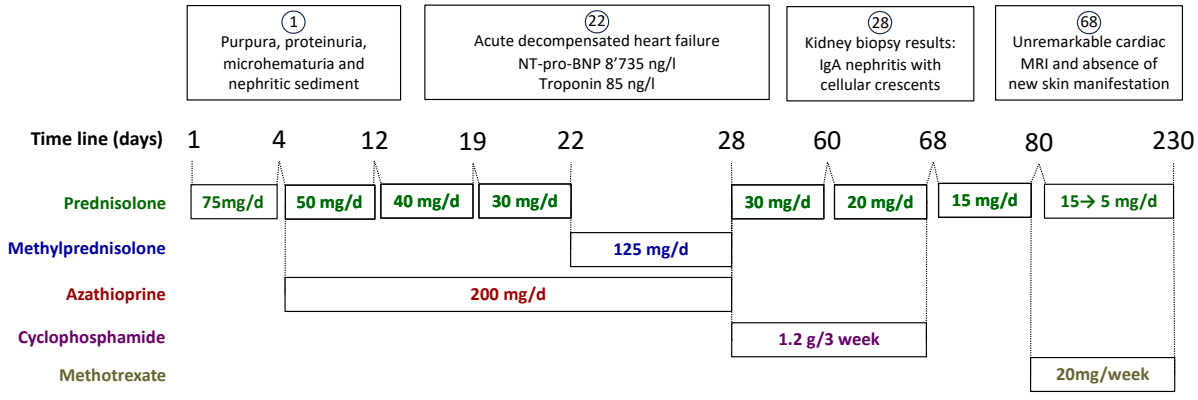

Figure 2: Summary of patient's clinical course together with treatment timing.

First author, year (reference)	Sex, Age	Cardiac involvement	Immunosuppression regimen	Cardiac outcome
Abdel-Hadi O, 1981[1]	Male, 29	Myocardial infarction	No reported immunosuppression	Persistent mild apical dyskinesia
Kereiakes DJ, 1984[2]	Male, 24	Myocarditis and heart failure	Prednisone, cyclophosphamide	Persistent congestive heart failure despite resolution of inflammation on the second myocardial biopsy
Satoh M, 1991[3]	Male, 64	Heart failure	No specified treatment	Not specified
Agraharkar M, 2000[4]	Male, 63	Myocardial necrosis and heart failure	Methylprednisolone	Death
Carmichael P, 2002[5]	Male, 69	Arrythmia	Methylprednisolone	Death
Hayakawa K, 2003	Male, 28	Transient myocardial ischemia	No reported immunosuppression	Complete remission
Hayakawa K, 2003[6]	Male, 20	Transient myocardial ischemia	Prednisolone	Complete remission
Polizzotto MN, 2006[7]	Male, 71	Arrythmia	Methylprednisolone	Complete remission
Lutz HH, 2009[8]	Male, 19	Arrythmia	Prednisolone, Cyclophosphamide	Complete remission
Sundriyal D, 2013[9]	Male, 21	Pericarditis	Prednisolone	Complete remission
Michas G, 2017[10]	Male, 19	Myocarditis and heart failure	No reported immunosuppression	Complete remission
Bando K, 2018[11]	Male, 60	Heart failure	Prednisolone	Improvement of the heart failure without complete remission
Torosoff, 2018[12]	Male, 61	Arrythmia and heart failure	Rituximab, Corticosteroid	Complete remission
Mank V, 2021[13]	Male, 61	Hemopericardium	Corticosteroid	Complete remission
Our patient	Male, 59	Heart failure	Prednisolone, Methylprednisolone, Cyclophosphamide, Azathioprine, Methotrexate	Complete remission

Discussion

This case highlights a rare HSP presentation with significant cardiac, renal, and gastrointestinal manifestations in an adult. While HSP is primarily known in pediatrics, where it often takes a benign course, cases in adults are more often severe [14-24].

In adults, its incidence ranges from 3 to 15 cases per million annually, with hallmark features including purpura, arthralgia, gastrointestinal symptoms, and renal impairment.

Notably, our literature review identified only 14 other cases of HSP with cardiac involvement. The cardiac manifestations in these cases included heart failure, arrhythmias, and myocardial ischemia, indicating a potential underappreciation of cardiac risks in HSP.

This patient presented with characteristic skin lesions and progressed to gastrointestinal bleeding and cardiac symptoms. The timely initiation of high-dose glucocorticoids and immunosuppressive therapy proved crucial in stabilizing his condition.

The management of HSP remains challenging, with treatment strategies generally involving immunosuppressive therapy ranging from glucocorticoid courses only to more aggressive B-cell targeted therapies tailored to individual clinical presentations. Our approach, combining glucocorticoids with cyclophosphamide and methotrexate, was associated with a favourable outcome in this severe presentation.

In summary, this case reinforces the need for clinicians to remain alert to the potential for cardiac involvement in adult HSP. Prompt recognition and aggressive management are vital, as these patients may experience severe life-threatening complications.

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