European Journal of Case Reports in Internal Medicine

Henoch-Schonlein Purpura in a Newly Diagnosed HIV Patient

Harshil Fichadiya[1], Garuav Mohan[1], Nimit Dalal[2], Hardik Fichadiya[3], Ahmad Al-Awan[1], Raghu Tiperneni[1], Farhan Khalid[1], Ramon Lopez Del Valle[4]

[1]Monmouth Medical Center, Long Branch, NJ, USA

[2]Western Reserve Medical Education, Youngstown, OH, USA

[3]Rutgers Trinitas Regional Medical Center, Elizabeth, NJ, USA

[4]Morsani College of Medicine, University of South Florida, FL, USA

Doi: 10.12890/2022_003363 - European Journal of Case Reports in Internal Medicine - © EFIM 2022

Received: 16/04/2022

Accepted: 19/04/2022

Published: 04/05/2022

How to cite this article: Fichadiya H, Mohan G, Dalal N, Fichadiya H, Al-Awan A, Tiperneni R, Khalid F, Lopez Del Valle R. Henoch-Schonlein purpura in a newly diagnosed HIV patient. *EJCRIM* 2022;**9**:doi:10.12890/2022_003363.

Conflicts of Interests: The Authors declare that there are no competing interest

ABSTRACT

HIV infection is associated with multisystemic manifestations due both to secondary infections caused by a decrease in the CD4+ T-cell count and to the pathogenicity of the HIV virus itself. A common renal manifestation is HIV-associated nephropathy, which is frequently seen in the African population with the APOL1 gene mutation; however, other forms of glomerulopathy such as IgA nephropathy, commonly noted in other ethnicities, are also seen. Vasculitis has rarely been associated with HIV infection and mainly involves small blood vessels, although any size of blood vessel may be involved. The association of Henoch-Schonlein purpura (HSP) with HIV is rare and not well understood. We describe a 53-year-old African American woman with a newly diagnosed HIV infection who presented with a purpuric rash over the bilateral lower extremities with haematuria. Initial work-up revealed renal dysfunction with elevated ESR. Urinalysis was positive for glomerular haematuria and sub-nephrotic range proteinuria. Serum complement level, c-antineutrophil cytoplasmic antibody (ANCA), p-ANCA and anti-nuclear antibody (ANA) were negative. Renal biopsy revealed mesangial IgA deposits with crescent glomerulopathy and fibrinoid necrosis, while skin biopsy revealed leucocytoclastic vasculitis. A diagnosis of HSP was made based on American College of Rheumatology (ACR) criteria. The patient's renal function and purpura improved with a 5-day course of steroid pulse therapy. This case of HSP in a newly diagnosed HIV patient is unusual for the presence of crescentic glomerulopathy.

KEYWORDS

Human immunodeficiency virus (HIV), Henoch-Schonlein purpura (HSP), leucocytoclastic vasculitis, IgA nephropathy

LEARNING POINTS

- Henoch-Schonlein purpura (HSP) associated with HIV infection is uncommon but documented; however, all four features of HSP are rarely seen together.
- Crescent glomerulopathy is rarely seen in HIV-associated HSP.
- HSP associated with HIV is treated with antiretroviral drugs, while the role of steroid and immunosuppressive therapy remains controversial.

INTRODUCTION

The first case of HIV-associated Henoch-Schonlein purpura (HSP) was reported in 1989 by Bauemelon [1]. HIV infection is associated with multiple autoimmune diseases such as immune thrombocytopenic purpura, inflammatory myositis, sarcoidosis, Guillain-Barre syndrome and myasthenia gravis [2]. The pathogenesis of HIV-associated vasculitis is not well understood but some studies suggest it could be due to

the immune dysregulation seen in HIV infection, the formation of immunoglobulins to HIV proteins, and endothelial dysfunction caused by the HIV virus. HSP is an IgA-mediated vasculitis and generally presents as a cluster of abdominal pain, palpable purpura on both lower extremities, arthralgia and IgA glomerulonephritis [2]. HSP associated with HIV is rare and not well understood. Reported cases of HIV-associated HSP demonstrated mesangial deposits of IgA on renal biopsy, skin leucocytoclastic vasculitis and joint manifestations [2]. However, crescentic glomerulonephritis and other manifestations of HSP are rarely seen [2–4].

CASE DESCRIPTION

A 53-year-old African American woman with a medical history of diabetes mellitus and hypertension and a recent history of cocaine use for the past 2 years presented with severe pain and swelling over the bilateral lower extremities. She had recently been diagnosed with HIV infection but had not initiated antiretroviral therapy. She mentioned that the rash had started as multiple maculopapular lesions over her lower extremities which progressively increased in size and worsened to develop into bumpy red lesions (palpable purpura) with ulceration. She complained of cola-coloured urine with peripheral oedema but denied abdominal pain, melena, haematochezia or joint pain.

Physical examination was significant for multiple palpable purpuras with superficial ulcers from the groin to the ankles with tenderness over the joints of the bilateral lower extremities with normal musculoskeletal, abdomen, respiratory, cardiovascular, neurological and HEENT examinations.

Laboratory studies revealed normocytic anaemia with a haemoglobin level of 8.8 g/dl. The WBC and platelet count were within normal limits. The basic metabolic panel revealed elevated blood urea nitrogen of 24 mg/dl, creatinine of 1.3 mg/dl, and hypoalbuminaemia of 3 g/dl. Urinalysis revealed haematuria with RBC casts and sub-nephrotic range proteinuria of 2.85 g/dl; the urine drug screen was positive for cocaine. ESR was elevated. Creatinine phosphokinase, serum complement and cryoglobulin levels, antinuclear antibody, c-ANCA, p-ANCA and rheumatoid factor were within normal limits. HIV-1 antibody was positive with a low CD4+ T-cell count of 299 cells/dl and elevated CD8+ T-cell count of 1476 cells/dl. Hepatitis B and C panels were negative. Ultrasound of the kidneys showed no abnormalities.

A subsequent skin biopsy showed leucocytoclastic vasculitis with a perivascular neutrophilic infiltrate with associated haemorrhage and fibrin deposition/fibrinoid necrosis of vascular walls. Direct immunofluorescence of the skin biopsy stained positive for IgA, IgM and C3. Renal biopsy revealed mesangial hypercellularity (M1), endocapillary hypercellularity (E1), no focal sclerosis (S0), mild tubular atrophy interstitial fibrosis (T0), and cellular crescents (C1) (crescents were seen in 5 out of 21 glomeruli) and focal fibrinoid necrosis with acute tubular necrosis. Immunofluorescence was positive for mesangial IgA (3+), C3 (3+), C1q and Lambda.

A diagnosis of HSP was made using the American College of Rheumatology (ACR) criteria. The presence of two out of the following four constitutes a positive diagnosis: (1) palpable purpura in the absence of thrombocytopenia; (2) granulocytes in the walls of small arterioles and venules; (3) bowel angina; and (4) age ≤20 years at disease onset [2]. The first two criteria were seen in our patient. The patient was treated with 1 g Solu Medrol IV daily for 5 days followed by an oral 60 mg dose of prednisone daily. Improvement in the patient's renal function and skin lesions was noted by day 5 of therapy. She was subsequently discharged with nephrology and infectious disease follow-up for initiation of antiretroviral therapy.

DISCUSSION

There is no clear consensus on the treatment of HIV-associated HSP. Studies show treatment with anti-retroviral drugs helps in inducing remission. The role of steroids and immunosuppression remains controversial. Our patient responded to pulse steroid therapy as in the case reported by Tada et al. [5]. We report this case in order bring attention to the rare association between HIV and HSP and to highlight the presence of crescent glomerulopathy in our patient as an uncommon occurrence. We urge further recognition and reporting of this association to encourage better study of treatment guidelines.

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