ISSN No:-2456-2165

Systemic Lupus Erythematosus and Henoch-Schönlein Purpura

Jaouad Yousfi*, Amal Elouakhoumi, Mouna Zahlane, Lamiaa Essaadouni
Department of Internal Medicine, University Hospital of Mohammed VI, Marrakesh, Morocco
Auteur correspondant*: Jaouad Yousfi, Department of Internal Medicine, University Hospital of Mohammed VI, Marrakesh, Morocco

Abstract:- Systemic lupus erythematosus (SLE) is an autoimmune disease that can cause kidney damage. However, lupus-unrelated renal injury has rarely been described in patients with SLE. We report the observation of a 28-year-old woman presenting with vascular purpura, inflammatory joint pain and abdominal pain. The skin biopsy showed vasculitis while direct immunofluorescence demonstrated IgA deposits. Two months later, the patient presented with malar rash and lower limb edema. She tested positive for anti-nuclear antibodies (1/1280 with a homogeneous pattern) and for anti-dsDNA (300 IU). Proteinuria was at 4.18g/24h. Renal biopsy revealed proliferative glomerulonephritis with IgA deposits. The diagnosis of SLE and IgA vasculitis with renal involvement was established. The patient was treated with corticosteroids and cyclophosphamide.

Keywords:- *Systemic Lupus Erythematosus -IgA Vasculitis – Nephropathy.*

I. INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune disease characterized by cutaneous and visceral involvement. Lupus nephropathy is the most common visceral manifestation of systemic lupus. However, the presence of renal involvement of other origin has been reported in patients followed for SLE. Thus, IgA vasculitis with renal involvement has been rarely associated with systemic lupus. We report a new observation of IgA vasculitis nephropathy associated with systemic lupus erythematosus in a young woman.

II. CASE REPORT

A twenty-eight-year-old patient presented with vascular purpura occurring four days before the admission, abdominal pain, bilateral and symmetrical inflammatory joints pain and diarrhea.

The clinical examination revealed an apyretic patient, purpura (Figure1) and ecchymosis of the lower limbs, slightly infiltrated, not confluent and without necrotic lesions. Ankle, knee and wrist joints were painful without any osteoarticular deformities. Abdominal examination showed diffuse abdominal tenderness without organomegaly. Cardiac and pulmonary auscultation were normal.

The biological assessment showed a biological inflammatory syndrome, negative viral serologies (HIV, Syphilis, CMV, hepatitis B and C). Elevated serum immunoglobulin A**at 30 g/l (**threshold of 10 U/ml)**.** The skin biopsy showed vasculitis with IgA deposits. The diagnosis of IgA vasculitis (Henoch-Schönlein purpura) was established. The patient was treated with oral corticosteroids(1mg/kg/day of prednisone) with clinical improvement.

Two months later, the patient presented with a malar rash (Figure 2), a photosensitivity and an inflammatory polyarthralgia. Clinical examination showededema of the lower limbs, without purpura. The urine test strip was positiveforred blood cells (2x) and proteins (3x). The biological assessment revealed lymphopenia at 810/mm^3, C-reactive protein at 6 mg/l and erythrocyte sedimentation rate of 23 mm. Antinuclear antibodies were positive at 1/1280 with a homogeneous fluorescence. Anti-dsDNA werepositive at 300 (threshold of 24). Proteinuria was 4.18g /day, albuminemia was at 35g/L and the renal function was normal (urea at 0.36 g/l andcreatinine at 8.2 mg/l). The renal biopsy showed a proliferative glomerular nephropathy with IgA deposition without IgM or IgG or C1q. The diagnosis of IgA vasculitis nephritis associated with SLE was made.

In addition to the corticosteroid therapy, the patient was treated with monthly intravenous bolus of cyclophosphamide. The 6 months follow-up showed clinical and biological improvement.

ISSN No:-2456-2165

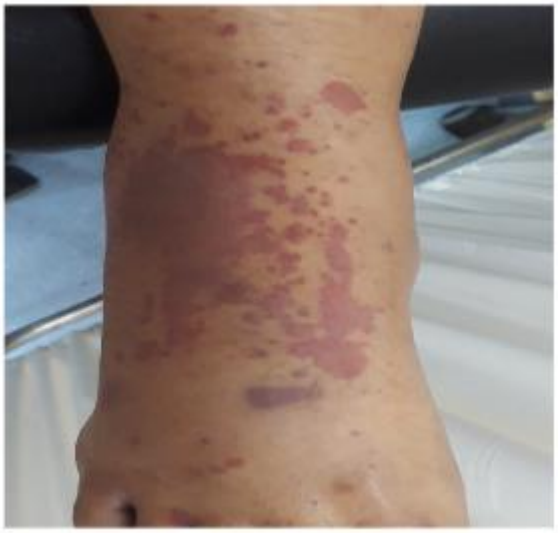

Figure1 : vascular purpura of the lower limbs

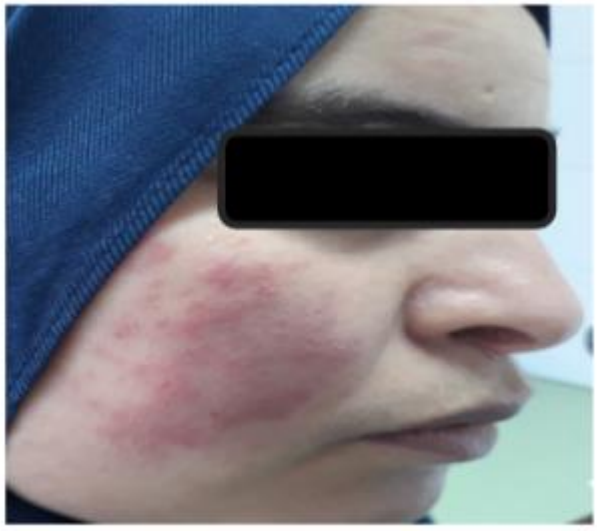

Figure2 : malar rash

III. DISCUSSION

SLE is a chronic autoimmune disease affecting multiple organs. IgA vasculitis is a systemic small-vessel vasculitis which can affect different organs including the kidneys. It is aleukocytoclastic vasculitis characterized by IgA1-immune deposits, complement factors and neutrophil infiltration. This inflammation affects mainly dermal, gastrointestinal and glomerular capillaries.[1]

IgA vasculitis is the most frequently encountered childhood vasculitis with an incidence of 10 cases per 100,000 child population. In adults, it is rare with an incidence of 3.4 to 14.3 cases per million. The clinical presentation of IgA vasculitisis generally more severe in the elderly andis characterized by nonthrombocytopenic palpable purpura, arthritis or arthralgias, gastrointestinal, renal involvement and, rarely, other systemic manifestations.[2]

Renal involvement in IgA vasculitis occurs more frequently in adults. The risk of progression to renal insufficiencyranges from 5% to 30%. Patients withIgA nephropathypresent with hematuria and proteinuria (50%), acute nephritic syndrome (8%), nephrotic syndrome (13%), and both in 29% of cases. Haematuria and proteinuria may follow relapses of purpura or may recur long after the extrarenal manifestations have resolved, with variable intensity[1].

Because of the rarity of IgA vasculitis with renal involvement in adults, its association with systemic lupus in the same patient is exceptional.[3]

The pathological study of the renal biopsy showed mesangial proliferation with isolated IgA deposits without other immune deposits. In contrast, lupus nephropathy is characterised by glomerular, tubular and interstitial lesions, with deposits of immunoglobulins (IgM, IgG +/- IgA) and complement (C1q, C3 and C4) [4]

The distinction between IgA nephrotpathy and lupus nephritis in SLE patients has important prognostic and therapeutic implications[5].

IV. CONCLUSION

Our observation highlights the importance of looking for other etiologies of renal involvement in lupus patients. Although rare, IgA vasculitis with its renal involvement can be associated with systemic lupus.

REFERENCES

[1]. Davin JC. Henoch-Schönlein purpura nephritis: pathophysiology, treatment, and future -strategy. Clin J Am Soc Nephrol. 2011;6:679 –89.

[2]. Saulsbury, Frank T. "Epidemiology of Henoch-Schönlein purpura." Cleveland Clinic journal of medicine vol. 69 Suppl 2 (2002): SII87-9

[3]. Bernardino V, Mendes-Bastos P, Rodrigues A, Riso N. IgA vasculitis (formerly Henoch-Schönlein purpura) in an adult with systemic lupus erythematosus. BMJ Case Rep. 2015;2015:bcr2015210121. Published 2015 Sep 9. doi:10.1136/bcr-2015-210121

[4]. Murata, C., Rodríguez-Lozano, A.L., Hernández-Huirache, H.G. et al. IgA vasculitis (Henoch – Schönlein Purpura) as the first manifestation of juvenile Systemic Lupus Erythematosus: Case-control study and systematic review. BMC Pediatr 19, 461 (2019)

[5]. Al-Attrach I, Al-Shibli A, Al-Riyami L, et al. Systemic lupus erythematosus with severe nephritis that mimicked Henoch-Schoenlein purpura. Arab J Nephrol Transplant 2011;4:159–61.