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Case Report
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Focal Segmental Glomerulosclerosis in Systemic Lupus Erythematosus

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Abstract

Nephrotic syndrome is a common presentation of lupus nephritis. In patient with Systemic Lupus Erythematosus (SLE) and the nephrotic syndrome, the two most common renal biopsy findings are diffuse proliferative lupus nephritis [World Health Organization (WHO) class IV] and membranous lupus nephritis (WHO class V). Lupus nephritis is divided into 6 distinct morphologic classes according to WHO. On the other hand non-lupus nephritis in patient with confirmed SLE is rarely reported. The relevant glomeruler lesions most frequently defined are amyloidois and Focal Segmental Glomerulo Sclerosis (FSGS). Thirty-six year-old female patients admitted with acute renal damage and nephrotic syndrome clinical findings. She fulfilled the following criteria of the American College of Rheumatology for SLE diagnosis: positive anti-dsDNA and ANA, leukopenia and cutaneous maculea. FSGS was detected in kidney biopsy. We believe that the patient had SLE and developed FSGS and we believe that lupus nephritis needs new classification.

Keywords: Acute kidney damage; Focal segmental glomerulosclerosis; Systemic lupus erythematosus

Introduction

Nephrotic Syndrome (NS) is a common presentation of Lupus Nephritis (LN). LN is divided into 6 distinct morphologic classes according to World Health Organization (WHO). LN is disunited 6 distinct morphologic classes according to the WHO classification: I-normal, II-mesangial proliferative, III-focal proliferative, IV-diffuse proliferative, V-membranous VI-advanced sclerosing glomerulonephritis [1, 2]. Beyond these lesions, renal biopsies from patients with Systemic Lupus Erythematosus (SLE) may show pathogenic and morphologically non-SLE-related changes [3]. Some of these cases have been described including several different renal lesions in which amyloidosis and focal segmental glomerulosclerosis predominate. It is important that other nephritic diagnosis in SLE patients may have different prognosis and treatment. Morphology, behavior and the best treatment approach for non-LN cases in SLE patients are not well known.

Case Presentation

We report on the case of a 36 year-old, Arabic woman. She fulfilled the following criteria of the American College of Rheumatology for SLE diagnosis: positive anti-dsDNA and ANA, leukopenia and cutaneous maculea. She reported three weeks history of edema of lower extremities that progressed anasarca, dyspnea and cough, night urination and decrease of urinary volume. The patient had no chronic illness, no drug or substance use. At the initial physical examination, blood pressure was 100/80 mmHg, body temperature 36.8°C, and heart rate 92 beats/min and rhythmic. It was observed edema of eyelid and 3+ edema of pretibial. Laboratory tests revealed serum creatinine: 1.6 (0.5-1.1) mg/dL, urea: 110 mg/dL, sodium:137 (134-148) mmol/L, potassium: 5.0 (3.5-5.5) mmol/L, hemoglobin: 9.38 (11-16) g/L, ESR: 66 mL/h, C-RP: 10.5 (0-5) mg/L, WBC: 3.71 (4-10.5)10^9, platelets: 259 (150-450) 10^9 /L, serum albumin: 1.8 (3.5-5.1)g/dL. Anti-dsDNA: >300.00 U /mL, pozitif ANA: 1.5 (0-1), Comp leman C3:42.9 (90-180) mg/dL, Comp leman C4: 6.1 (10-40) mg/dL, IgA: 181 (82-453) mg/dL, IgG: 1390 (751-1560) mg/dL, IgM: 158 (46-304) mg/dL. Sediment microscopy of urine showed 10-15 RBCs/HPF (high power field) and 5-10 WBCs/hpf; proteinuria was detected at 3.8 g/day. Ultrasonographic evaluation revealed normal kidney size, increased to grade 1 echogenicity. In the following days, urine output fell below 400 cc. Serum creatinine varied from 1.6 mg/dl to 5.7 mg/dl and patient was hipervolemic. Immediately started treatment with

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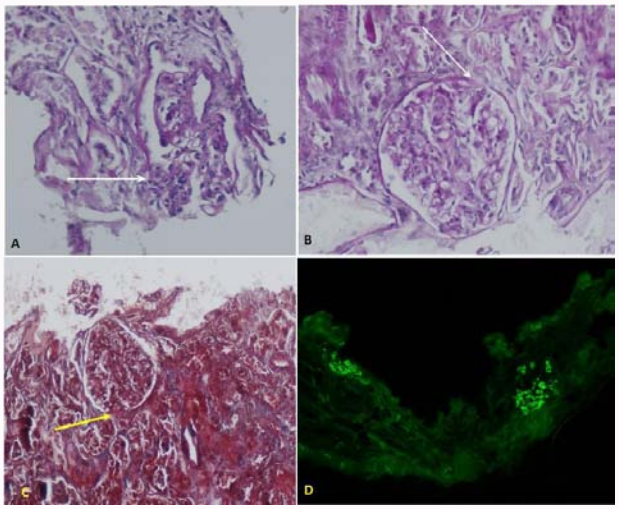

Figure 1: (A) Segmental sclerosis in the glomerulus, H&E × 200 (Arrow); (B) Adhesion of glomerular capillaries to bowman capsule, PAS × 200 (Arrow); (C) Adhesion of glomerular capillaries to bowman capsule, Tricrom × 100 (Arrow); (D) Coarse granular 3+ staining with C3 in mesengial areas, immunofluorescense ×100.

hemodialysis and kidney biopsy was performed. We thought she was diffuse proliferative lupus nephritis of SLE. Immediately we treated the patient with intravenous prednisolone 1000 mg three days. After she was treated with 48 mg oral methylprednisolone. As she was followed in our service during 25 days, her renal function was normally and proteinuria within 25 days went down to 1.8 g/day. She had no edema anywhere, and did not need hemodialized. She was treated with methylprednisolone (1 mg/kg/day) and miklofenat mofetil (2 g/day). FSGS was detected in kidney biopsy. Endothelial tubuloreticular inclusions were also seen. Pathology was consistent with FSGS. Renal biopsy did not reveal glom ruler lesions by the immunofluorescense was negative for C1q, IgG, IgM and IgA, but 3 positive for C3 (Figure 1). Two months after discharge from hospital laboratory test revealed: serum creatinine: 0.5 mg/dl, ürea: 33 mg/dL, albumin: 3.9 g/dL, urinary microscopy is normal and protein excretion is 200 mg/day.

Discussion and Conclusion

Our patient presented at least four American College of Rheumatology criteria for diagnosis of SLE (leukopenia, cutaneous maculae, positive ANA, positive anti-dsDNA) and other clinical findings consistent with this diagnosis (hypocomplementemia and arthralgia). It is generally believed that renal abnormalities are caused by LN in well-documented SLE patients. Thus, renal biopsy in SLE is usually recommended not for diagnostic purposes, but rather to determine the type and extent of renal involvement [4], as well as to evaluate the activity and chronicity indices. Interestingly, in rare cases, clinically significant renal diseases not associated with LN have been described in patients with SLE. There are only a few reports that this association is often isolated.

In SLE patients, a wide variety of renal lesions not associated with LN may be seen. Renal biopsy plays an important role in determining glomerular disease associated with SLE. SLE patients clinically presenting with nephrotic syndrome indicating Minimal Change Disease (MCD), Mesangial Proliferation (MsP) or FSGS, while on electronic microscopy, diffuse podocyte foot process removal in absence of sub-epithelial or sub-endothelial deposition is the only morphological feature and now diagnosed as lupus podocytopathy. There are 17 previous reports of FSGS in SLE [3-10]. All reported patients presented with NS, normal or more frequently decreased renal function and hypertension. FSGS was preceded 3 to 18 years by renal abnormalities, which were related mostly to a previous LN. How the non-LN develops in patients with SLE is still poorly understood. The association between SLE and some diseases, such as thin-membrane disease, hypertensive nephrosclerosis, IgA nephropathy and infection-related GN is probably coincident. On the other hand, the more frequent association with FSGS and amyloidosis may be pathogenetically concerned to SLE [3,4]. Our patient had the histological diagnosis of FSGS.

Lupus podocytopia is susceptible to glucocorticoid therapy. However, the relapse rate can reach up to 90% in treatment with glucocorticoid alone. Glucocorticoid plus other immunosuppressive agents can significantly reduce the relapse rate. Lupus podocytopathy with FSGS offers higher sensitivity to acute renal damage and less sensitivity to glucocorticoid treatment [3,11,12]. We treated for FSGS after the biopsy; the patient experienced a full remission during the course of the disease. When non-LN was diagnosed, it was treated with myclofenate mofetil in addition to corticosteroids for FSGS.

Finally, non-LN may be seen in SLE patients regardless of clinical or serological disease activity. Renal biopsy is an important tool in the identification of these lesions. These kidney lesions have a broad morphological spectrum; one of the most common is FSGS. The combination of corticosteroids and other immunosuppressive agents in SLE patients with acute renal damage and FSGS has increased the success of treatment.

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