Case Report

Rapidly progressive glomerulonephritis as presenting feature of systemic lupus erythematosus in a Bangladeshi male patient: a rare case report

Haque S[a], Jahan I[b], Chowdhury TA[c], Rahim MA[d], Ananna MA[d], Iqbal S[e]

Abstract

Rapidly progressive glomerulonephritis is one of the most dramatic and tragic presentations of lupus nephritis (LN) or renal manifestation of systemic lupus erythematosus (SLE). A 35-year-old Bangladeshi gentleman presented with worsening oedema, scanty, high colored, frothy urine and deteriorating renal function. He had puffy face, anaemia, oedema, normal jugular venous pressure (JVP), high blood pressure (150/90 mm Hg), ascites and bilateral pleural effusions. Diagnostic work-up confirmed SLE with class IV LN. His initial response to specific therapy showed improvement.

Key words: *male patient, presentation, rapidly progressive glomerulonephritis, systemic lupus erythematosus.*

(BIRDEM Med J 2020; 10(2): 137-138)

Introduction

Systemic lupus erythematosus (SLE) is an uncommon diagnosis, further uncommon in a male patient. Renal involvement is an important determinant of outcome in SLE, with 38% prevalence rate and 40-60% of those having the disease during initial presentation.[1] Rapidly progressive glomerulonephritis (RPGN) is one of the most dramatic and tragic presentations of lupus nephritis (LN) or renal manifestation of SLE.[2] Here, we present case history of a young Bangladeshi male patient, who had RPGN as initial manifestation of SLE.

Case report

A 35-year-old hypertensive male got admitted with a 20-day history of worsening oedema, scanty, high colored, frothy urine, without any preceding history of fever, sore throat, skin rash, joint pain, chest pain or shortness of breath. However, he had one episode of epistaxis.

He had puffy face and there was anaemia, oedema, normal jugular venous pressure (JVP) and high blood pressure (150/90 mm Hg). He had proteinuria (++). Systemic examination revealed ascites and right sided pleural effusion. Other examination finding revealed no abnormality.

He had anaemia (haemoglobin 7.9 gm/dL, normocytic, normochromic), normal total and differential white cell counts and platelet counts, high erythrocyte sedimentation rate (104 mm in 1st hour), slightly raised C-reactive protein (11.4 mg/L), normal reticulocyte counts (2%), negative direct and indirect Coomb's test and raised lactate dehydrogenase (1090 U/L) levels. There was evidence of rapid deterioration of renal function (serum creatinine became 3.8 mg/dL from 1.1 mg/dL through 1.5 mg/dL within 1 month) with proteinuria (+++), hematuria [25-40 red cells/high power field (HPF)], pyuria (plenty of pus cells/HPF) and granular cast (4-6/HPF). Chest x-ray revealed bilateral pleural effusion, more marked in right side (Figure 1), abdominal ultrasonography showed normal sized kidneys with slightly increased cortical echogenicity and minimal ascites and bilateral mild pleural effusions.

Author information

a. Samiha Haque, Resident Medical officer, Nephrology, BIRDEM General Hospital, Dhaka, Bangladesh.

b. Ishrat Jahan, Assistant Registrar, Nephrology, BIRDEM General Hospital, Dhaka, Bangladesh.

c. Tufayel Ahmed Chowdhury, Registrar, Nephrology, BIRDEM General Hospital, Dhaka, Bangladesh.

d. Muhammad Abdur Rahim, Mehruba Alam Ananna, Associate Professor, Nephrology, BIRDEM General Hospital, Dhaka, Bangladesh.

e. Sarwar Iqbal,Professor, Nephrology, BIRDEM General Hospital, Dhaka, Bangladesh.

Address of correspondence: Samiha Haque, Resident Medical officer, Nephrology, BIRDEM General Hospital, Dhaka, Bangladesh. Email: trinasamiha@gmail.com muradrahim23@yahoo.com

Received: January 31, 2020 **Accepted:** February 29, 2020

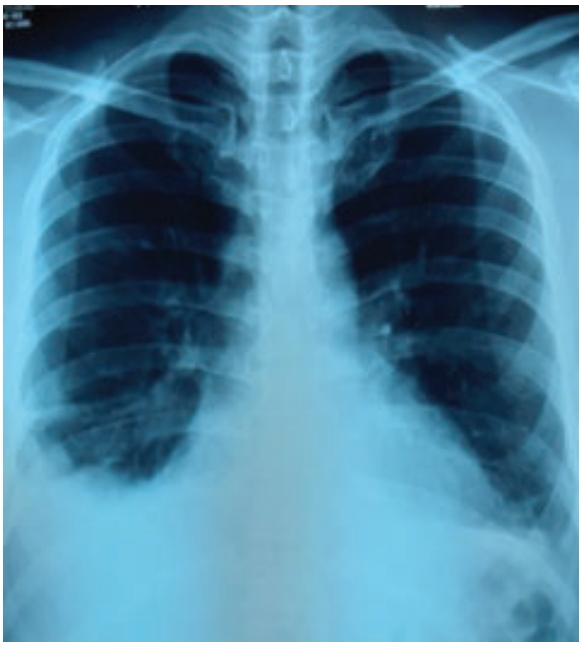

Figure 1 Chest x-ray postero-anterior view showing bilateral pleural effusions

Antinuclear antibody (ANA) (10: 600, cut off 1: 500) and anti-double stranded deoxyribineucleic acid (anti-ds DNA) (>240 IU/mL, ref <20) were positive, while other vasculitis markers, components of extractable nuclear antigens (ENA) and viral markers remained negative. C3 level was low (0.59 g/L, ref 0.9-1.8). 24-h urinary total protein was 4.22 g and serum albumin was 16.5 g/L. A renal biopsy confirmed class IV lupus nephritis (LN) with active lesions and crescent. He was started with intravenous (IV) methyl prednisolone, mycophenolate mofetil, hydroxy-chloroquine, diltiazem and prazocin. His initial response to therapy showed improvement.

Discussion

Patients with LN may have heterogeneous presentations; they may have asymptomatic urine abnormality through acute nephritic syndrome, nephrotic syndrome to chronic kidney disease and end-stage renal disease (ESRD) or in various combinations of these features. Rarely, patient may present with RPGN, may require renal replacement therapy for a temporary basis and progress to ESRD.[1-3]

RPGN forms around 2-10% of all renal biopsies.[4] It has been estimated that up to 30% of cases of active LN result in RPGN.[5] RPGN is a clinical syndrome with rapid loss of renal function and have potential to lead to ESRD within weeks to months. RPGN has poor outcome, specially, if not recognized and treated early.[3] Timely recognition and appropriate therapy often fail to guarantee a full recovery.

Whenever indicated[1], a renal biopsy should be undertaken in patients with SLE, having features of renal involvement, as, often, urine albumin-creatinine ratio and other markers fail to correlate with biopsy findings and treatment is directed towards histological classification on renal biopsy findings.

Class IV active disease merits active intervention with IV methyl prednisolone along with other immunosuppressive agents.[1] Every patient with a diagnosis of SLE should have hydroxyl-chloroquine as indicated by guidelines[1] because of its proven benefits in preventing life- and organ-threatening flares.

Outcome of treatment in LN is variable. Patients presenting with RPGN may have a poor outcome, as many patient may progress to ESRD.[2] However, delay in making a diagnosis and initiation of appropriate intervention remain amongst the other outcome determinants. Nevertheless, no intervention can guarantee a successful renal recovery. In refractory LN, many agents are on way of research.[1]

In conclusion, any patient, who presents with rapid decline in renal function without an identifiable aetiology, should raise suspicion of RPGN, merit urgent investigation and treatment, as delay in establishing diagnosis and treatment initiation may compromise outcome.

Conflict of interest: Nothing to declare.

References

1. Kidney Disease: Improving Global Outcomes (KDIGO) Glomerulonephritis Work Group. KDIGO Clinical Practice Guideline for Glomerulonephritis. Kidney Inter 2012; 2(suppl.): 139-74.
2. Komolafe OO. Rapidly Progressive Glomerulonephritis: A Wild Card Manifestation of Lupus Nephritis. Saudi J Kidney Dis Transpl 2018;29(2):443-51.
3. Jastrzêbska K, Gozdowska J, Perkowska-Ptasiñska A, Durlik M. Rapidly Progressive Glomerulonephritis Associated with Systemic Lupus Erythematosus. Arch Clin Nephrol 2016;2(1):28-31.
4. Nagaraju SP, Laxminarayana SLK, Kosuru S, Parthasarathy R, Attur RP, Rangaswamy D, et al. Clinicopathological characteristics and outcomes of diffuse crescentic glomerulonephritis - A single center experience from Southern India. J Clin Diagn Res 2017;11:OC21-4.
5. Appel GB, Cameron JS. Lupus nephritis. In: Feehally J, Floege J, Johnson RJ, editors. Comprehensive Clinical Nephrology. 3rd ed. Philadelphia, PA: Mosby Elsevier; 2007. p. 291-03.