Case Report

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A case of Systemic Lupus Erythematosus with class IV nephritis in a 60 year old female

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Abstract

Patients who develop Systemic Lupus Erythematous (SLE) at or after the age of 50 years are considered as late-onset SLE. In this case report, we present a case of class IV Lupus Nephritis in a 60-year-old lady with newly diagnosed SLE. This lady presented with generalized edema and reduced urine output and found to have active urinary sediment, rising serum creatinine, positive ANA and dsDNA and hypocomplementemia. Renal biopsy proved diffuse proliferative glomerulonephritis (Lupus Nephritis – class IV). She was treated with methyl prednisolone and cyclophosphamide as the induction regime. Then treatment was continued according to Euro- Lupus Protocol. She showed clinical and biochemical improvement.

Key words

SLE (Systemic Lupus Erythematosus), glomerulonephritis, Late-onset SLE, Lupus Nephritis (LN)

Introduction

Systemic Lupus Erythematosus predominantly affects females with childbearing age and the incidence declines after menopause. But it might evolve newly in women at or after the age of 50 years and that is called "Late-onset SLE", which constitutes 2-20% of all patients with SLE (1). Here we report a 60-year-old female being diagnosed with SLE complicated with class IV lupus nephritis.

Case presentation

A 60-year-old lady presented to the hospital with erythematous skin lesions preferably involving lower extremities associated with progressive generalized edema. She also had tiredness, fatigability and hair loss. She did not have joint pain or other features of autoimmune connective tissue disorders. After admission, oedema worsened, and she began to produce reduced amount of urine. She had a complex spectrum of disease and suffered from hypothyroidism, sero negative Rheumatoid Arthritis from the age of 57 years, Type2DM and Hypertension. Clinically she was ill looking. She had palpable, non-blanching rash especially over the lower limbs distally. Her blood pressure was poorly controlled with her routine antihypertensive medications.

Laboratory analysis showed urinary active sediment and worsening serum creatinine from 50micromol/l baseline to 120micromol/L. Urine protein creatinine ratio was 207.19mg/mmol (subnephrotic range proteinuria). Full Blood Count showed normocytic normochromic anemia. ESR was 60mm/1st hour and the CRP was normal.

Skin biopsy was not suggestive of true vasculitis and perivascular lymphocyte infiltration. Specialized investigations revealed positive ANA and dsDNA(with titres >1:100 and >150IU/ml, respectively). Blood C3 and C4 levels were reduced (25.5 and 7.9mg/dl respectively), pANCA positive and other autoantibodies including Rheumatoid

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factor and cANCA were negative. Hepatitis B and C screening were negative. Chest Xray showed mild cardiomegaly.

She underwent renal biopsy which showed Diffuse proliferative glomerulonephritis, morphology compatible with Lupus nephritis – class IV and co-existing diabetic nodular glomerulosclerosis. There were no cellular or fibrous crescents. Direct immunofluorescence showed IgG , C3 and C1q diffuse intense granular mesangial and glomerular basement membrane staining along with IgA and IgM light granular mesangial and glomerular membrane staining.

With this diagnosis of class IV Lupus Nephritis, she was treated with pulses of IV Methylprednisolone and IV cyclophosphamide. Then her treatment was continued according to Euro – Lupus Protocol. She improved well clinically and biochemically, oedema settled and Creatinine clearance improved.

Discussion

Systemic Lupus Erythematosus (SLE) is a chronic inflammatory disease with multisystem involvement and approximately 35% of patients will have some evidence of lupus nephritis at the time of diagnosis.

The patient in our case was diagnosed with SLE with Lupus Nephritis according to 2012 SLICC criteria. Even though organ involvement is less and more benign course is noticed in late-onset SLE because of higher chances of having associated co-morbid conditions and more organ damage, it has poorer prognosis (2).

Our patient was a 60-year-old female with SLE with biopsy proven class IV Lupus nephritis, without involvement of other major systems. She has multiple Co-morbidities such as poorly controlled hypothyroidism, Type2DM and Hypertension. So, her prognosis cannot be determined by the severity and the extent of organ involvement in related to SLE per se.

Late-onset SLE patients have lower frequency of cutaneous, neuropsychiatric, and renal manifestations (2). Late-onset SLE patients need lower dose of corticosteroids and cyclophosphamide but develop more complications with cyclophosphamide. Typical presentation with non-specific symptoms results in longer delay of diagnosis (2).

When we consider the serological markers, almost all have ANA positivity. The results of anti-dsDNA in case of Late-onset SLE are contradicting and according to Fu SM et al. anti-dsDNA antibodies might not be most specific marker for SLE (3).

A comprehensive analysis of different age groups including 287 patients by *Mak et al*. concluded that therapy for Lupus nephritis should be individualized rather than not considering the age factor alone. In Late-onset SLE patients with Lupus nephritis, more careful and judicious use of immunosuppressant regimens is needed to minimize the therapy induced complications (4). However, at diagnosis of Lupus nephritis significantly higher proportion of patients in Late-onset SLE have hypertension and higher serum creatinine (4)

Late-onset SLE patients are more likely to die due to treatment related complications, increased background co-morbidities and pre-existing Hypertension. It also concluded that low complement levels during the first year of diagnosis is the only serological marker of poorer survival (5).

Conclusion

This case represents a Late-onset SLE patient with lupus nephritis. Though she has class IV Lupus nephritis, the severity of renal impairment is

relatively lower as her serum creatinine did not rise to more than 120micromol/L (eGFR according to CKD _ EPI formula 42.3ml/min/1.73m^2). The ACR guidelines for the treatment of Lupus nephritis can be followed for Late-onset SLE patients as well. Extra care should be given as they are more prone to treatment related complications such as infections, marrow suppression and osteoporosis.

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