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A Young Male Patient of Systemic Lupus Erythematosus (SLE) Presenting With Early Lupus Nephritis:-A Case Report

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Abstract: *SLE is one of several diseases known as "the great imitators" because it often mimics or is mistaken for other illnesses. SLE symptoms vary widely and appear and disappear unpredictably. Diagnosis can thus be elusive, with some patients having unexplained symptoms of untreated SLE for years. Autoimmunity plays a major role in the pathogenesis of lupus nephritis. Lupus nephritis(LN) is one of the most serious complications of systemic lupus erythematosus(SLE) since it is the major predictor of poor prognosis. The immunologic mechanisms include production of autoantibodies directed against nuclear elements. Here is a case report of a male patient who presented with Anasarca, Respiratory Distress, Proteinuria and was diagnosed with lupus nephritis.*

Keywords: *SLE, Lupus Nephritis, Auto-Immunity.*

I. Introduction

SLE is a chronic, occasionally life-threatening, multisystem disorder. Patients suffer from a wide array of symptoms and have variable prognosis that depends upon the severity and type of organ(s) involved. Females are more commonly affected in SLE; In children female to male ratio is 3:1.In contrast, the ratio in adults ranges from 7–15:1. In older individuals, the ratio is approximately 8:1.[1]The overall prevalence and incidence of SLE ranges from 1.4 to 21.9% and from 7.4 to 159.4 cases per 100,000 people, respectively [2].SLE can affect several organs and systems, including the joints, skin, brain, heart, lungs, blood vessels and kidneys.

Lupus nephritis (LN) is one of the most serious SLE complications since it is the major predictor of poor prognosis. Up to 25% of these patients still develop end-stage renal disease (ESRD) 10 years after onset of renal compromise [3].In addition, LN may develop early in the course of SLE thus becoming a major predictor of poor prognosis. However, in about 5% of the cases, LN may appear several years after the onset of SLE (i.e., delayed LN) [4]. The group with delayed LN is positively associated with Sjögren syndrome(SS), lung involvement, and anti -phospholipid syndrome as compared with early LN (i.e., those SLE patients who develop LN during the first 5 years of the disease) [5].

II. Case Report

15 years old male presented with complaints of progressive breathlessness since 15 days, cough, intermittent fever without chills and rigors and decreased urine output since 10 days. On general examination Patient was having Pallor +1, Anasarca, Tachycardia, Tachypnoea with RR of 30/min, BP was 150/90 mm of Hg. On systemic examination, on auscultaton B/L basal crepts were present. Rest of the systemic examinations were normal. Patient had history of previous hospitalization in private sector since last 4 days and referred for Intensive care as patient had complaints of increased breathlessness and expectoration of pink frothy sputum. He was then intubated and initiated on diuretics. Investigations revealed Hb-7 g/dl, WBC 4800 cells/mcL, Platelets 1,45,000 cells/mcL, Creatinine-3.4 mg/dL, Urea-90 mg/dL, S.Electrolyte- WNL, Albumin-1.8 gm/dl, Total Protein- 4.2 gm/24 hour, Urine dipstick test-severe proteinuria with 4+ Albumin and 24 Hrs Urinary Protein-4 gm/l

Patient was treated with furosemide and FFP to relieve the congestive changes. On further investigation and evaluation of proper history, patient was found to be diagnosed with SLE 2 months back and was on Hydroxychloroquine and steroids since 1 month, Anti ds-DNase was Positive and C3 was low. CXR was found to be S/O Congestive changes and B/L blunting of CP angles. USG- S/O Moderate collection on Right side and Mild to moderate collection on Left side. Diagnostic and therapeutic tapping was done to relieve the breathlessness and reports were S/O Transudative collection. ECG had Non-specific ST-T changes, Echo- S/O Pericarditis. After stabilizing the patient and extubating on third day, suspecting lupus nephritis, patient was advised for renal biopsy.

Renal biopsy revealed partial to complete encircling fibro-cellular crescents in 50 % of glomeruli while rest have mild increase in mesangial matrix and cells with mild neutrophilic exudation present, Basement membrane showed mild thickening, Patchy tubular atrophy, interstitium shows mild oedema and Patchy Inflammatory cell infiltrate, Immunofluorescence study showed IgA:+- coarse granular along capillary wall; IgG,IgM,C1q & Kappa: Positive coarse granular along capillary wall +1; C3 and Lambda: Positive Coarse Granular along capillary wall +2.Patient was diagnosed with Lupus Nephritis Class IV-S(A/C) (ISN/RPS 2004).

Patient was treated with anti-hypertensives, pulse therapy of steroids, cyclophosphamide 3 doses and Hydroxychloroquine. 8 units of FFP and 1 unit RCC was given. He was discharged on Anti-hypertensives, Hydroxychloroquine and steroids.

III. Discussion

Patient was a known case of SLE and presented with multiorgan involvement mainly kidney.The patients had laboratory abnormalities such as elevated serum creatinine levels, low albumin levels, or urinary protein or sediment suggesting active lupus nephritis. Symptoms related to active nephritis may include peripheral edema secondary to hypertension or hypoalbuminemia. Extreme peripheral edema is more common in persons with diffuse or membranous lupus nephritis, as these renal lesions are commonly associated with heavy proteinuria.[6] Patients with active lupus nephritis often have other symptoms of active systemic lupus erythematosus (SLE), including fatigue, fever, rash, arthritis, serositis, or central nervous system (CNS) disease. These are more common with focal proliferative and diffuse proliferative lupus nephritis.[7]

Evaluating renal function in patients with systemic lupus erythematosus (SLE) to detect any renal involvement early is important because early detection and treatment can significantly improve renal outcome.[6] Renal biopsy should be considered in any patient with SLE who has clinical or laboratory evidence of active nephritis, especially upon the first episode of nephritis.[6,8] So Patient Renal Biopsy was done and Patient was diagnosed as a case of lupus nephritis stage IV according to the classification revised by the International Society of Nephrology (ISN) and the Renal Pathology Society (RPS) in 2004. This classification is based on light microscopy, immunofluorescence, and electron microscopy findings from renal biopsy specimens.

The principal goal of therapy in lupus nephritis is to normalize renal function or, at least, to prevent the progressive loss of renal function. Therapy differs depending on the pathologic lesion.[6, 9] .Corticosteroid therapy should be instituted if the patient has clinically significant renal disease. Use immunosuppressive agents, particularly cyclophosphamide, azathioprine, or mycophenolate mofetil, if the patient has aggressive proliferative renal lesions, as they improve the renal outcome. They can also be used if the patient has an inadequate response or excessive sensitivity to corticosteroids.[9,10,11]

The first guidelines for managing lupus nephritis have been issued by the American College of Rheumatology.[12] Patients with clinical evidence of active, previously untreated lupus nephritis should have a renal biopsy to classify the disease according to International Society of Nephrology/Renal Pathology Society criteria. All patients with lupus nephritis should receive background therapy with hydroxychloroquine, unless contraindicated. This recommendation was based on a prospective controlled trial showing lower flare rates in those who continued hydroxychloroquine, compared with those who switched to placebo.[13] Glucocorticoids plus either cyclophosphamide intravenously (IV) or mycophenolate mofetil orally for induction in patients with ISN class III/IV disease. Patients with ISN/RPS class I and II nephritis do not require immunosuppressive therapy. Administer ACE inhibitors or angiotensin-receptor blockers if proteinuria is 0.5 g/24 h or more. Maintain blood pressure at or below 130/80 mm Hg. Patients with end-stage renal disease (ESRD), sclerosis, and a high chronicity index based on renal biopsy findings are unlikely to respond to aggressive therapy. In these cases, focus therapy on extrarenal manifestations of systemic lupus erythematosus (SLE) and on possible kidney transplantation.

IV. Conclusion

All the patients of SLE presenting with deranged Renal function test should be promptly investigated and suspected for lupus nephritis and treatment should be started at the earliest after diagnosis is established. Though its expected that Lupus Nephritis usually manifests around 4-5 years after diagnosis, early presentation can occur in patients with relatively shorter duration of illness.

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