

Case report

Systemic Lupus Erythematosus presenting as Lupus Nephritis

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Abstract

Systemic Lupus Erythematosus (SLE) is an Auto immune disease and multigenic disease in which organs and cells undergo damage mediated by tissue-binding autoantibodies and immune complexes. ^[1] 90% of patients are women of child bearing years. Prevalence of SLE is 4-250/100,000 have been reported with female predominance varying from 4:1 before puberty to 8:1 after puberty. Most common clinical manifestations are systemic (95%): fatigue, fever, malaise, weight loss and musculoskeletal features (95%) and cutaneous manifestations - photosensitivity and rash (70 - 80%) and less common clinical manifestations are renal (30 - 40%), ocular (15%), Thrombosis (15%). The diagnosis of SLE is typically based on the revised (1997) criteria of American College of Rheumatic Association (ACR). ^[2] Anti dsDNA antibodies are highly specific for SLE but they are present only in 70% of cases. ^[3] Manson et al. found high avidity anti dsDNA antibodies in only 13 out of their 16 patients and recommended antinucleosome antibodies to further increase diagnostic specificity. ^[4] Min et al. in a prospective study of 129 patients with SLE found little or no anti dsDNA reactivity in 25 cases during the course of their disease. Among these antinucleosome antibodies were present in 15 patients, 8 patients with anti dsDNA negative SLE had renal disease and all these cases had antinucleosome antibodies. ^[5] We report a rare case of Renal involvement in SLE with absence of rash.

Keywords: Anti dsDNA antibodies, Antinuclear antibodies, Antinucleosome antibodies

Case Report

A 22 year old female presented with fever and joint pains since one month and facial puffiness (Figure1) and swelling of legs since 15 days. There was no history of rash. The joint pains are polyarticular bilaterally symmetrical associated with swelling of wrist and finger joints predominantly. There was no history of hematuria. There was significant family history of similar complaints in her elder sister and she died at age of 23 years due to renal complications.

On general examination pallor present, generalized alopecia and oral aphthous ulcers (Figure 2), bilateral pitting pedal oedema, facial puffiness were present and there was no rash. Systemic examination was normal except for the musculo skeletal examination. Swelling of wrist and proximal interphalangeal joints was present. Local raise in temperature and tenderness was present. No wasting and deformity was observed. Range of movements was decreased and all other joints were normal.

Cardio Gram and imaging techniques like 2D Echocardiography, Chest X-ray and Ultrasonography abdomen shown normal findings.



Fig. 1: Facial puffiness and Alopecia.



Fig. 2: Oral Aphthous ulcers

On laboratory investigations, Haemogram with peripheral smear revealed normocytic normochromic anemia with leucopenia with thrombocytopenia.

Hemoglobin concentration was 7gm%; Erythrocyte Sedimentation Rate (ESR) was 80 mm/hr. Renal Function Tests shown that Serum creatinine was 1.4mg%, Glomerular Function Rate (GFR) was 60 ml/min and Blood urea was 40 mg%. On Complete Urine Examination, there was Presence of protein in the urine and microscopic hematuria. 24 hour urine protein was found to be 2gm per day.

Liver Function Tests, Thyroid Function Tests and Fasting Lipid Profile were normal. Viral markers were found to be non reactive. Specific tests like, Antinuclear Antibodies (ANA) were '3+'; anti dsDNA were '2+'; Complement C3 C4 levels were decreased and C-Reactive Peptide was positive. It was also found that Rheumatoid factor and Anti Streptolysin O titres were negative. Electro

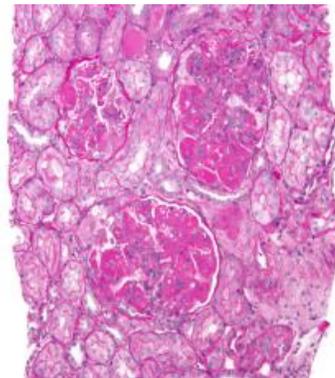


Fig. 3: Light microscopy findings.

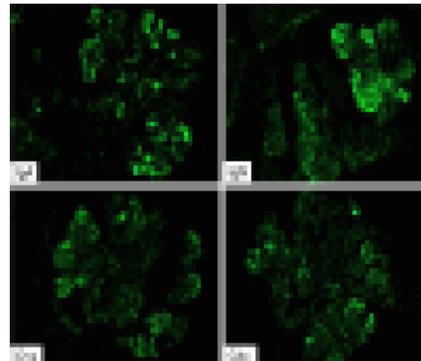


Fig. 4: Immunofluorescence findings.

On Renal biopsy, light microscopy (Figure 3) revealed wire loop lesions, Endocapillary and mesangial proliferation. All these findings were suggestive of diffuse proliferative nephritis. Immunofluorescence (Figure 4) revealed, significant peripheral and mesangial granular deposits of IgG, IgM, IgA, C3c and C1q, suggestive of grade IV Lupus Nephritis.

Based on the clinical, laboratory findings and histopathological examination, the condition was diagnosed as SLE with Grade IV Lupus Nephritis.^[2]

Discussion

Nephritis is usually the most serious manifestation of SLE, particularly since Nephritis and infection are the leading causes of mortality in the first decade of

disease. Since Nephritis is asymptomatic in most Lupus patients, urinalysis should be ordered in any person suspected of having SLE. The classification of Lupus Nephritis International Society of Nephrology (ISN) is primarily histologic. ^[6] Renal Biopsy is useful in planning current and near future therapies. Patients with dangerous proliferative forms of glomerular damage (ISN III & IV) usually have microscopic hematuria and Proteinuria (more than 500 mg/24 hr) and approximately one-half develop Nephrotic Syndrome and most develop hypertension. If Diffuse Proliferative Nephritis (DPGN) is untreated, virtually all patients develop End Stage Renal Disease (ESRD).

Therefore aggressive immunosuppression is indicated (usually systemic glucocorticoids and cytotoxic drugs), unless damage is irreversible. Mycophenolate mofetil is as effective as cyclophosphamide in inducing remission in Lupus Nephritis. This regimen is safe with a lower risk of ovarian failure. ^[7, 8] Overall in USA 20% of individuals with Lupus DPGN die or develop ESRD within 10 years of diagnosis. In children ESRD or deaths has been reported in 20-50% at 10 years follow up due to progressive parenchymal injury, such individuals require aggressive control of SLE and of the complications of renal disease and of therapy. ^[3] Proteinuria is less likely to improve on Lupus Nephritis immunosuppressive therapies. Lupus Nephritis tends to be an ongoing disease, with flares requiring re-treatment over many years.

Considering the atypical presentation and the importance of the early diagnosis and aggressive management for a better prognosis the present case has been reported.

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