

## Case Report

### Symptomatic Lupus Nephritis in a Male Child: An Unusual Presentation

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**Abstract:** Systemic lupus erythematosus (SLE) is a classical autoimmune disorder with a predilection to occur in females of reproductive age group. Paediatric onset SLE has a low incidence and is more common among female children. We describe a case of a 8 year old boy suffering from this disease who presented with fever, polyarthritis and malar rash along with clinical and pathological evidence of renal involvement. This case is being reported for its rarity in presentation in a male child with clinically symptomatic and pathologically documented lupus nephritis.

**Keywords:** SLE, nephritis, children

#### INTRODUCTION

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disorder in which there is widespread circulation of autoantibodies and deposition of immune complexes in various tissues leading to a wide spectrum of clinico-pathological manifestations. Although the disease predominantly involves women of child bearing age, patients in any age group can be affected. Kidneys are one of the most frequently involved organs in SLE.

#### CASE REPORT

A male child, aged 8 years presented with complaints of low grade fever, facial rash and pain in the wrists and knee joints for the last 9 months. He also complained of swelling around both his ankles which was gradually increasing during the last 3 months. Past medical history and drug history were insignificant.

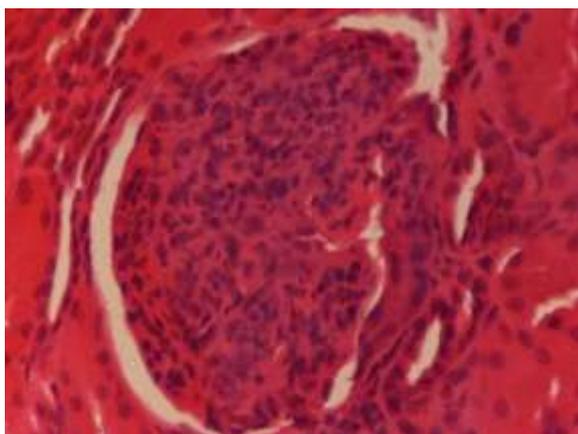
General examination revealed average built individual with mild pallor and bilateral painless pitting pedal edema. The wrists and knee joints were bilaterally tender but no deformity was present. In addition, he also had a butterfly shaped malar rash and his blood pressure was 150/90 mm of Hg. There was no lymphadenopathy or hepatosplenomegaly. Examination of respiratory, cardiovascular and nervous systems were within normal limits. A provisional diagnosis of SLE was made and the patient was admitted for thorough investigations.



**Fig. 1: SLE: Typical malar rash seen on the cheeks**

Subsequently, his haemogram revealed haemoglobin of 10.5 gm/dl, total leucocyte count of 9,500/mm<sup>3</sup> with platelet count being 2,96,000/mm<sup>3</sup>. Peripheral smear examination showed normocytic to microcytic RBCs with mild hypochromia with no immature cells. A raised erythrocyte sedimentation rate (ESR) of 80 mm/hour was noted. On serum biochemistry, total protein was 5.0 gm/dl, albumin 2.1 gm/dl, urea 56 mg/dl and creatinine 1.0 mg/dl. Antinuclear antibody (ANA) and anti ds-DNA were positive in the titres of 1:640 each. Urine examination showed proteinuria 4+, RBCs 10/HPF and hyaline casts 2/HPF.

Subsequently a kidney biopsy was performed in the patient to assess the extent of renal involvement. Two creamish white biopsy fragments were obtained, measuring 1.4x0.1 cm and 0.5x0.1 cm, respectively. Haematoxylin and eosin stained sections of these fragments showed 5 glomeruli with mild diffuse mesangial hypercellularity and increase mesangial matrix. Vessels showed mild endothelial proliferation with insignificant change in the tubules. No evidence of segmental necrotic lesions, haematoxylin bodies or wire loop lesions were detected. Our diagnosis went in favour of diffuse proliferative glomerulonephritis (WHO Class-IV lesion).



**Fig. 2: Lupus Nephritis: Tissue sections of the kidney showed glomeruli with mild diffuse mesangial hypercellularity and increase mesangial matrix. Haematoxylin and Eosin x 40 X**

Our patient was given an aggressive regimen of intravenous methylprednisolone with cyclophosphamide. He is doing well after 12 months of follow up period.

## DISCUSSION

Systemic lupus erythematosus is a classic example of multisystem autoimmune disease that is caused by failure of recognition of self proteins by the body's own defence mechanism and subsequent production of autoantibodies against a wide range of body proteins [1]. Although the disease can involve people of any age or sex, 85-90% patients are females in the reproductive age group. A higher incidence of SLE has been reported in African-Americans as compared to Whites [2]. SLE diagnosed in paediatric age group (0-12 years) has a low incidence of 0.36-2.5 per 1,00,000 population per year with a female preponderance in the ratio 4.5:1 [3, 4].

The exact etiology of SLE is not known. Kyttaris *et al.* [1] proposed that a variety of environmental, hormonal and immunological factors lead to the expression of the disease in a genetically susceptible host. Abnormal activation of B-cells, T-cells and antigen-presenting cells cause production of highly specific pathogenic autoantibodies. These circulating

antibodies and immune complexes bind to target organs, activate complement and incite an inflammatory damage resulting in the plethora of clinical and pathological manifestations associated with the disease. The organs damaged in SLE chiefly include synovial joints, kidneys, skin, lung, cardiovascular and central nervous systems [1].

In children, SLE commonly presents with intermittent or persistent symptoms of fever, rash, arthritis and nephritis. Haematolymphoid abnormalities in the form of anaemia, thrombocytopenia, leucopenia, lymphadenopathy and hepatosplenomegaly are more frequently seen than adults [4]. For the diagnosis of SLE, presence of at least 4 out of the 11 criteria as described by American College of Rheumatology (1997) is essential [5]. Our patient satisfied five of these criteria viz., arthritis, malar rash, nephritis, positive ANA and anti ds-DNA.

Font *et al.* [6] prospectively analyzed 34 patients of SLE with initial presentation in childhood and compared their disease pattern with that of adults. They observed that in paediatric onset SLE, fever, nephropathy and lymphadenopathy were the more common presenting features at the time of diagnosis while malar rash, chorea and raised anti-cardiolipin antibody were more frequent during evolution of the disease.

Around 30-75% cases of childhood onset SLE have associated nephropathy and these cases usually manifest after 10 years of age [4, 7]. Currently, lupus nephritis is defined as the presence of more than 3+ or 0.5 gram/24 hour proteinuria or presence of urinary cellular casts of any type [5]. The spectrum of presentation is diverse, ranging from asymptomatic proteinuria to overt nephrotic syndrome or even renal failure. In the presence of active disease, serum C3 and C4 levels tend to be reduced [1]. A distinct feature of lupus nephritis is that it has a tendency to progress over time and often the pathological findings fail to correlate with the clinical features. Hence, renal biopsy is essential to assess the extent of kidney damage and categorize the severity of lesions, so that appropriate management plan can be formulated [4, 7].

World Health Organization (WHO) [8] has classified renal lesions of lupus nephritis into 6 histological categories as follows: Class-I. Minimal mesangial glomerulonephritis (GN); Class-II. Mesangial proliferative GN; Class-III. Focal GN; Class-IV. Diffuse GN; Class-V. Membranous GN and Class-VI. Advanced sclerotic GN. This classification has prognostic significance and helps in choosing the therapeutic modality [4, 7].

In children, Class-IV lesion is most common accounting for up to 50% of the cases and is more frequent among boys than girls. Moreover, patients with

this lesion are often clinically symptomatic with proteinuria, haematuria and hypertension. However, this lesion carries the worst prognosis due to its high risk of progressing to end-stage renal disease [4, 7]. Therefore, it is of utmost importance to identify patients with Class-IV lupus nephritis early in the course of the disease, so that deterioration of renal function can be retarded with aggressive immunosuppressive and/or cytotoxic drug therapy [4, 9].

The main aim of treatment in lupus nephritis is to achieve clinical and serological remission. Favourable response to therapy is indicated by reduction of proteinuria, serum creatinine and ESR, and return of Anti ds-DNA, C3, C4 levels to normal range [10]. WHO Class-I and II lesions do not require any specific therapy but need regular follow up for monitoring any progression to a more severe renal disease. Class-III lesions can be managed by low dose corticosteroids. Class-IV lesions need aggressive treatment with glucocorticoids (oral Prednisolone or i.v. Methylprednisolone) alone or in combination with cytotoxic agents like, Cyclophosphamide, Azathioprine and Rituximab (in refractory cases). Duration of treatment is variable depending up on the severity of renal dysfunction. The treatment of Class-V lesions remains controversial [9, 10]. With appropriate management, progression to end-stage renal disease and mortality can be significantly reduced. The long-term outcome of childhood lupus nephritis is similar to that seen in adults [9-11].

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