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Young Adolescent Male Patient of Systemic Lupus Erythematosus Presenting With Lupus Nephritis: A Case Report

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Abstract: *Systemic lupus erythematosus is a rare and chronic autoimmune disorder characterized by multisystem inflammation and presence of circulating autoantibodies directed against self antigens. SLE occurs in both children and adults , disproportionately affecting females of reproductive age group . The occurrence of SLE in patient of paediatric is itself very rare and that too in a male child. Here we are describing a case of 15 yrs old boy suffering from SLE who presented with generalised body swelling , malar rash and clinical evidence of renal involvement along with extra-renal sign and symptoms. We are reporting this case for its rarity in male child with clinically documented lupus nephritis.*

Keywords: *Autoimmune, lupus nephritis, systemic lupus nephritis,SLE.*

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I. Introduction

Systemic lupus erythmatosus (SLE) is a rare autoimmune disease with antibodies directed against self antigens which get deposited in various tissues of body. It affects nearly every organ more commonly skin, joints, blood forming cells , blood vessel, central nervous system and the kidneys as the most frequenty affected organ. The disease usually affects female of reproductive age group with reported 2-5 : 1 ratio prior to puberty, 9 : 1 ratio during reproductive years and return to near prepubertal ratios in the postmenopausal period. Children and adolescents though affected rarely, have more widespread organ involvement.Female preponderance suggests hormonal influence, and sex hormone is known to modulate immune system and here the risk of SLE increases with use of estrogen containing contraceptives. Childhood SLE may present acutely, but more commonly it is known to evolve over a period of time [1-2]. SLE, especially the early disease, mimics several other systemic disorders. Therefore diagnosis is usually missed out in the initial stages, and more so if the patient happens to be a young teenager male. Lupus nephritis is one of serious complication of SLE, and is also a major predictor of poor prognosis [3-5].

II. Case report

A 15 yrs old male born of non consanguineous marriage presented with complaints of swelling of face, lower limbs and distension of abdomen along with reduced urine output for 15 days. Swelling first involved face progress to abdomen then lower limbs and was preceded by pain in throat and fever. **Past history:** Patient had similar history one year back. **On general examination**: Revealed mild pallor, left cervical lymphadenopathy, bilateral pedal edema , distended abdomen , periorbital swelling with no clubbing or icterus. A butterfly shaped malar rash , Photosensitivity was present with no oral ulcers . Erythema over palmer aspect of hand. **On gastrointestinal system examination**: Shifting dullness was present indicate ascitis . Examination of other system were normal. Provisional diagnosis of systemic lupus erythmatosus with renal involvement was made. **Diagnosis:** Routine hemogram, serum electrolytes, blood urea , serum creatinine were normal. Total cholesterol was raised , serum albumin and total serum protein were significantly low. HIV1&2 and HBsAg both -ve Routine examination of urine showed nephrotic range proteinuria, RBCs and pus cells. Anti-nuclear antibody and anti-dsDNA tests were strongly positive. ESR was raised and C3 level was low . ECG and echocardiography were normal. Ultrasonography of w/a showed ascites. Renal biopsy revealed Immune complex mediated, diffuse proliferative necrotizing crescentric glomerulonephritis displaying secondary segmental tuft sclerosis involving 50% glomeruli.(class IV lupus nephritis) **Treatment:** patient treated with pulse therapy of methylprednisolone , high doses prednisolone tapered over 4 to 6 months , hydroxychloroquine and mycophenolate mofetil along with albumin infusion and other supportive therapy. Patient responded well with reduction in proteinuria, ESR and C3 returned to normal with resolution of generalised edema.

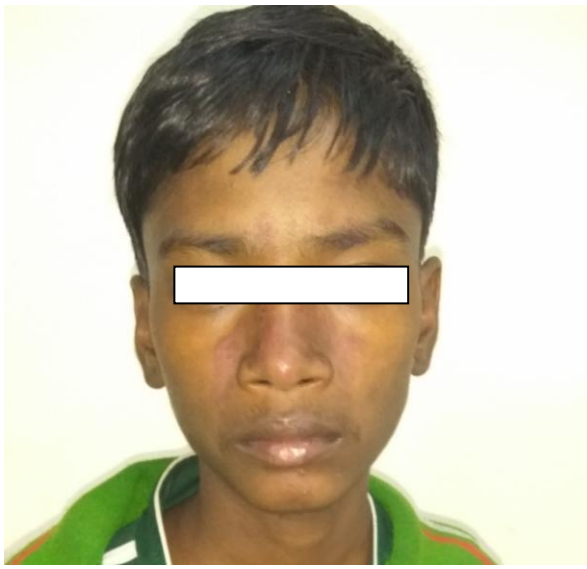

Fig 1: Malar rash

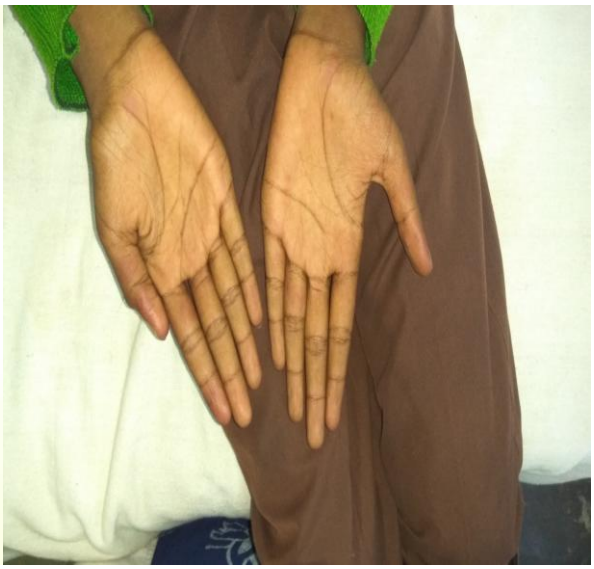

Fig 2: Vasculitic rash over palm

III. Discussion

Systemic lupus erythematosus is a classic autoimmune disease with circulating autoantibodies formation against various self antigens and affect various systems of body. Gender is the strongest risk factor as nearly 90 % of patients are female of reproductive age. The prevalence of SLE in children and adolescents is (1-6/100,000). Only 20% of patients are diagnosed prior to age of 16 years. There is no known etiology of SLE but there is possibility of presence of environmental, hormonal and immunological factors leading to expression of the disease in genetically susceptible host. In SLE both B cells and T cells show functional impairment . B cells show impaired tolerance and their increased autoreactivity enhances ability of B cells to produce autoantobodies on self antigenic exposure following some triggering factor . The abnormal activation of B cells, T cells and antigen presenting cells produce highly pathogenic circulating autoantibodies , the immune complexes so formed get deposited in various body tissues leading to local complement activation, severe local inflammation and ultimately tissue damage.[6]

In SLE, children and adolescents mostly present with fever, fatigue, hematologic abnormalities, arthralgia and arthritis. For the diagnosis of SLE presence of 4 out of 11 criterias by American college of rheumatology 1997 is essential. These criterias include 1.Malar rash, 2. Discoid rash, 3. Photosensitivity, 4.Oral or Nasal ulcers, 5.Arthritis , 6.Serositis , 7.Renal manifestations, 8. Seizures or Psychosis,9..Hematological manifestations, 10.Immunologic abnormalities ,11. Positive anti nuclear antibody test result . In our patient 6 criterias were present which were malar rash, photosensitivity, serositis in form of ascites , renal features like heavy proteinuria was there with positive anti ds-Dna antibody and positive anti nuclear antibody test hence diagnosed as a case of SLE wth renal involvement or lupus nephritis. In 2012, the Systemic Lupus Collaborating Clinics proposed the SLICC criteria for SLE in view of new knowledge of autoantibodies and the importance of low complement.[1,6]

Renal disease in SLE is often asymptomatic, underscoring the need for careful monitoring of blood pressure and urinalyses; in adolescents SLE often presents with nephrotic syndrome and/or renal failure with the predominant symptoms being edema, fatigue, changes in urine color, and nausea/vomiting.[6]

Classification of Lupus Nephritis

	Class	Clinical finding
I.	Minimal mesangial LN	No renal findings
II.	Mesangial proliferative LN	Mild clinical renal disease; minimally active urinary sediment; mild to moderate proteinuria (never nephrotic) but may have active serology.
III.	Focal proliferative LN <50% glomeruli involved A. Active A/C. Active and chronic C. Chronic	More active sediment changes; often active serology; increased proteinuria (approximately 25% nephrotic); hypertension may be present; some evolve into class IV pattern; active lesions require treatment, chronic do not.
IV.	Diffuse proliferative LN (>50% glomeruli involved) all may be with segmental or global involvement (S or G)	Most severe renal involvement with active sediment, hypertension, heavy proteinuria (frequent nephrotic syndrome), often reduced glomerular filtration rate;serology very active. Active lesions require treatment.

	A. Active A/C. Active and chronic C. Chronic	
V.	Membranous LN	Significant proteinuria (often nephrotic) with less active lupus serology.
VI.	Advanced sclerosing LN	More than 90% glomerulosclerosis;no treatment prevents renal failure.

Children with lupus nephritis are treated with immunosupression therapy the goal of which is to produce a clinical remission with normalization of renal function and proteinuria, and a serological remission which is normalization of anti ds-DNA antibody , C3 and C4 levels. All patients are started with prednisolone 1-2mg per kg in divided doses followed by slow tapering over 4 to 6 months beginning 4-6 weeks after achieving serologic remission. The patients with severe forms of lupus nephritis like class III with active lesions and class IV induction therapy with cyclophosphamide 500-1,000 mg/m^2 is done along with pulse intravenous methylprednisolone 1,000mg/m^2 in addition to oral corticosteroids. Maintenance therapy is done by additional cytoxan infusion for three months for 18 months. Mycophenolate mofetil, azathioprine or rituximab may also be used . For extrarenal manifestations of lupus nephritis like malar rash , hydroxychloroquine is used . ACE inhibitors are used to treat associated hypertension. [6]

IV. Conclusion

Childhood SLE is a challenging disease both difficult to diagnose and to treat. It is less often observed in children than adults rare in male child. The clinicians should be aware of the greater risk of systemic complications in children with systemic lupus erythematosus. Lupus nephritis should be suspected, which needs to be confirmed by a renal biopsy . Henceforth, pediatric SLE patients should be continually followed up and appropriate therapy should be initiated depending upon the disease activity to reduce morbidity and mortality.

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