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A RARE CLINICAL PRESENTATION OF JUVENILE SYSTEMIC LUPUS ERYTHEMATOSUS WITH LUPUS NEPHRITIS, HEPATITIS AND CELIAC DISEASE IN SICKLE TRAIT

Paediatrics

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ABSTRACT

Background: Systemic lupus erythematosus (SLE) is chronic autoimmune disease, characterized by multisystem inflammation and presence of circulating autoantibodies directed against self-antigens. Renal involvement (lupus nephritis) in juvenile SLE is more common and more severe than that seen in adults. **Clinical description:** We report a case of juvenile SLE in a 9year old undernourished female presenting with periorbital puffiness, bipedal edema and abdominal distension, associated with low grade fever, skin rashes, polyserositis, hepatitis and generalized lymphadenopathy and hypertension. Laboratory investigations suggested hypoalbuminemia, anemia, thrombocytopenia, positive urinary protein/creatinine ratio, Raised ESR, deranged LFT, ANA by IF showed speckled pattern with +3 intensity and positive TTG IgA antibody. **Management:** Patient was treated with corticosteroids, IV cyclophosphamide and other immunosuppressive agents, diuretics, antihypertensive drugs, IV albumin, diet modification (gluten free diet) and macro and micro nutrients. **Conclusion:** Due to complex and chronic nature of SLE, patients with SLE to be treated by pediatric rheumatologist in a multidisciplinary clinic with access to a full complement of pediatric subspecialist.

KEYWORDS

systemic lupus erythematosus, antinuclear antibody, immunofluorescence, tissue transglutaminase IgA antibody, Liver function test

INTRODUCTION:

Systemic lupus erythematosus (SLE) is chronic autoimmune disease, characterized by multisystem inflammation and presence of circulating autoantibodies directed against self-antigens. SLE predominantly affects females, with reported 2-5:1 ratio before puberty, 9:1 ratio during reproductive years, and return to near-prepubertal ratio in the postmenopausal period. Childhood SLE is rare before 5 year of age and is usually diagnosed in adolescence, with a median age at diagnosis of 11-12 years[1]. Up to 20% of all individuals with SLE are diagnosed before age 16 year. The pathogenesis of SLE remains largely unknown, but several factors likely influence risk and severity of disease, including genetics, hormonal milieu, and environmental exposures. In children, renal involvement (lupus nephritis) is more common and is more severe than that in adults. Lupus nephritis is the most important cause of morbidity and mortality in SLE. Gastrointestinal manifestations are not uncommon in patient with SLE. There is an increased frequency of positive serologic celiac disease markers in patients with systemic lupus erythematosus [2]. The determination of celiac disease (CD) in patient with SLE is clinically important because the a patient with SLE and CD share a variety of autoantibodies, common HLA type and may frequently have overlapping symptoms and findings.[3]

CASE DESRIPTION:

A 9year old female child, presented with hyperpigmented skin rashes and history of hair fall, recurrent mouth ulcers and recurrent loose stools since 2 years; low grade fever, weight loss, abdominal pain, bipedal pitting edema and abdominal distension since 3 months and periorbital puffiness since 15 days. On examination, cachexic look, patches of alopecia, multiple hyperpigmented rashes over entire body, periorbital edema, pallor, generalized lymphadenopathy, hepatosplenomegaly with moderate ascites and bipedal pitting edema. In physical examination, patient was febrile, radial pulse 85/min, respiratory rate 22/min and BP-130/90 (hypertensive).

Table 1: Laboratory Investigations

Laboratory parameter	Value	Interpretation
Haematological		
Haemoglobin	7.8 g/dL	Moderate anemia
Total leukocyte count	7800/mm3	WNL
Platelet counts	80000/mm3	Thrombocytopenia
ESR	101 sec	Raised
Renal function test		
Creatinine	0.4mg/dl	Normal
Calcium	7.4mg/dL	Hypocalcemia
Liver function test		
Prothrombin time-INR	3.1	Deranged
S. Albumin	1.0g/dL	Hypoalbuminemia
Total protein	4.9g/dL	Hypoproteinaemia
SGPT	225	Elevated
CRP	126	Raised
Anemia profile		
Sickling test	Positive	
HPLC	Sickle cell trait	
DCT/ ICT	Negative	
S. Iron	54	Normal
LDH	200	Normal
Urine examination		
Routine microscopy	Albumin +3	Albuminuria
Urine albumin by heat	+3	
Urinary protein/creatinine ratio	5.5	Significant
24hour urinary protein	94.31(0-15)	Proteinuria
Thyroid profile (TSH)		Euthyroid
Lipid profile		
Total cholesterol	161	Normal
Triglyceride	266	
Complement levels		
S. C3 level	<0.3 (0.9-1.8)	Hypocomplementemia
S. C4 level	<0.06 (0.1-0.4)	
ANA profile		
ANA	Speckled pattern, +3 intensity	Positive
Anti-Smith antibody	+3	Positive
Anti SSA antibody	+3	Positive
TTG-IgA	182.56 U/mL	Positive
2D echocardiography	Pericardial effusion	
CT-scan(Thorax, abdomen)	Hepatosplenomegaly, Moderate ascites, pericardial effusion, abdominal LAP, Bilateral pleural effusion	

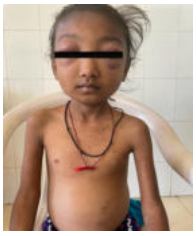

Figure:1

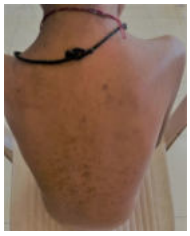

Figure:2

Figure 1, 2: Clinical Presentation of SLE Patient

Hemogram reports suggested moderate anemia and thrombocytopenia with raised ESR. Anemia workup done to rule out the cause of anemia, patient was found to be sickle cell trait. Chronic infections (Tuberculosis, seropositivity) were ruled out for generalized lymphadenopathy. Clinically patient presented with generalised anasarca with polyserositis (ascites, pleural and pericardial effusion); cardiac function abnormalities were ruled out and patient was euthyroid. As patient had abdominal pain with tenderness in right hypochondrial region with ascites, Liver function tests were found deranged; coagulation abnormality and elevated aminotransferase enzymes were noted along with hypoproteinaemia and hypoalbuminemia. Due to persistence of severe hypoalbuminemia despite albumin transfusion, to rule out the route of protein loss, nephrotic workup done-urinary protein/creatinine ratio and urine albumin positive, hypocomplementemia, hypertension suggestive of nephrotic syndrome and oral steroids (prednisolone) was started along with antihypertensive measures in form of salt restriction, high protein diet, oral furosemide, spironolactone and enalapril.

In a view of haematological profile (Anemia, thrombocytopenia), positive inflammatory markers (ESR, CPR), Lymphadenopathy, hyperpigmented rashes with periorbital puffiness with nephrotic syndrome in a female child of pre-pubertal age points high index of suspicion towards underlying rheumatological disorder; ANA profile was carried out revealed strongly positive ANA titre (+3) with anti-Smith antibody. Keeping a provisional diagnosis of SLE with Lupus nephritis, Rheumatologist and Nephrologist opinion was taken, Review of ANA result led us to a diagnosis of Systemic Lupus Erythematosus when combined with 4 other criteria that she had fulfilled viz; serositis, rashes, nephritis and haematological manifestations. She was commenced on high dose prednisolone while awaiting renal biopsy and ophthalmologic examination prior to commencing hydroxychloroquine. Renal biopsy was planned for staging of lupus nephritis and treatment, but the parents refused for invasive procedures, thus DMARDs (disease modifying antirheumatic drugs)-hydroxychloroquine, Immunosuppressive agents – Inj Methylprednisolone for 3 days and Inj Cyclophosphamide single dose was given.

As patient had history of recurrent loose stool with weight loss since last 2-3 years, presence of wasting and pallor on examination with hypoalbuminemia, to differentiate between protein losing enteropathy and malabsorption syndromes, TTG IgA was done showed positive result and provisional diagnosis of celiac disease was made and upper GI endoscopy was planned for later and gluten free diet was advised to parents.

Progressive disease control was achieved with prednisolone, antihypertensive medications, monthly cyclophosphamide with dietary medications was discharged successfully with plan for regular follow up.

DISCUSSION:

SLE in children is more active and is associated with more rapid accrual of damage than is SLE in adults. SLE may presents with pancytopenia or isolated cell line depletion, unusually elevated ESR depend upon disease activity and CRP is usually normal except in presence of infection[4]. Our patient had anemia with thrombo cytopenia with raised ESR and CRP. A higher frequency of renal disease and a concomitantly lower frequency of cardiopulmonary involvement have been reported in Juvenile SLE. This increased frequency and severity of renal disease was associated with increased disease activity, which was primarily observed in the renal domain rather than in other organ systems. children with childhood-onset SLE had more active disease at diagnosis and developed damage over time more rapidly. Reasons for greater disease activity and more damage in childhood-onset SLE remain to be elucidated. Hormonal changes observed during puberty might add to the imbalance of the immune system in children with SLE.[5]

The American College of Rheumatology (ACR) criteria[6] for the diagnosis of SLE revised in 1992 require that at least 4 out of the 11 criteria should be present either serially or simultaneously[7]. Our patient fulfilled the following five of the ACR criteria; serositis, nephritis, skin rash, positive ANA and Haematological criteria.

CONCLUSION:

This case report shows the diverse clinical manifestation of SLE in a paediatric patient that constitutes a diagnostic challenge due to its varied and alternating clinical spectrum. The clinician should be aware of the various manifestation and complications in children with SLE, so that appropriate therapy can be initiated early to reduce morbidity and mortality.

REFERENCES:

1. Kliegman, St Geme, Blum, Shah, Tasker, Wilson. Systemic lupus erythematosus. In Nelson textbook of pediatrics Volume 1, 21st edition. International edition. Elsevier; 2020.p 1274-1280.
2. F. Alenzi et al; The value of screening for celiac disease in systemic lupus erythematosus: A single experience of a tertiary medical center; Rheumatol Ther (2020) 7:649-656
3. Y. Sahin et al; Serological screening for celiac disease in children with systemic lupus erythematosus; Eur J Rheumatol 2019; 6(3): 142-5
4. AdisaAK, Adamu H, Asani MO, Aliyu I; Systemic lupus erythematosus in a 7 year old girl:A first case report from Northern Nigeria; Niger J Paed(2015);42(2):158-161
5. Hermine I. Brunner,Dafna D. Gladman,Dominique Ibanez, Murray D. Urowitz,and Earl D. Silverman; Difference in Disease Features Between Childhood-Onset and Adult-Onset Systemic Lupus Erythematosus; ARTHRITIS & RHEUMATISM 2008; 58(2) p 556–562
6. Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum1997;40:p1725
7. Narayanan N. Systemic Lupus Erythematosus. In: Parthasarathy A, Menon PS, Gupta P, Nair MK, Agrawal R, Sukumaran TU editors. IAP Textbook of Pediatrics. 5th ed. New Delhi (India): Jaypee Brothers Publishers Ltd, 2013:871 -3.