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CASE REPORT ON SYSTEMIC LUPUS ERYTHEMATOSUS IN A MALE CHILD WITH ALL ELEVEN CHARECTERISTICS OF AMERICAN COLLEGE OF RHEUMATOLOGY CRITERIA.

Pediatrics

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ABSTRACT

Systemic lupus erythematosus(SLE) is a chronic autoimmune disease characterised by multisystem inflammation and presence of circulating autoantibodies directed against self antigen. Male patients with SLE present mainly with renal involvement and seizures rather than photophobia and skin manifestations. The outcome also seems to be more serious in males. Here we have reported a case who had malar rash, discoid rash, oral ulcers, serositis- Peritonitis, arthritis, renal involvement in the form of Proteinuria, Microscopic haematuria,neurological involvement like seizure, headache, postive MRI brain findings, haematological involvement like anemia,thrombocytopenia , immunological involvement (Anti ds DNA positive) , ANA positive. Thus it had all eleven characteristic according to AMERICAN COLLEGE OF RHEUMATOLOGY REVISED CLASSIFICATION CRITERIA for diagnosis of SLE. Thus this case is unique and rare.

KEYWORDS

SLE, male child, rare presentation

INTRODUCTION

Systemic lupus erythematosus(SLE) is a chronic autoimmune disease characterised by multisystem inflammation and presence of circulating autoantibodies directed against self antigen. Of individuals with SLE, 90% are females making gender the strongest risk factor of SLE. The increase frequency of SLE among women may be attributed to the effect of estrogen and its hydroxylation, decreased androgen levels, hyperprolactinemia and differences in gonadotropin-releasing hormone (GnRH) signalling[1]. Reported cases on male SLE presented mainly with renal involvement and seizures and have a more rapid progression [2].

Table 1. American College of Rheumatology criteria for SLE

CRITERION	FEATURE
1-malar rush	erythema on malar eminences
2-discoid rush	erythematous lesions with keratotic scalling
3-oral ulcer	oral and nasopharyngeal ulcers
4-photosensitivity	dermal sensitivity to sun light
5-arthritis	non-erosive oligo arthritis
6-serositis	pleuritis, pericarditis
7-renal	glomerulonephritis, proteinuria
8-hematologic	leucopenia, anemia, thrombocytopenia
9-neurologic	seizure, stroke, phsycosis
10-immunologic	anti ds DNA, anti cardiolipin antibodies
11-antinuclear ab.	existence of antinuclear antibody

CASE

An 11 years old male child presented with puffiness of face followed by swelling of legs and distension of abdomen, oliguria, photosensitivity, mucosal ulcers ,on and off fever and headache for 4 months. He had pain and swelling of right elbow joint for 5 days and 2 episodes of generalised seizure, mucosal bleed and high grade fever for 1 day prior to admission in the hospital. He had past history of fever with rash 8 months back, pain and swelling of both knee and ankle joint 4 months back which subsided in two months and yellowish discolouration of eyes and urine 4 months back for which herbal medication was taken and symptoms got relieved. His mother died when he was 3 years old suffering from SLE and had similar rash and swelling of the body for 1 ½years and died due to renal complications. He was delivered by normal vaginal delivery in their district civil hospital, his birth weight was 2.8kg, cried immediately after birth and his perinatal and postnatal history was uneventful. His development history was normal and he attained his milestones at an age similar to children in his neighbourhood.According to kuppuswamy scale he belonged to upper lower socioeconomic class.

Examination on the day of examnation showed that the patient was drowsy , his pulse rate was 140/min ,regular in rhythm, nomal in volume and charecter, no radioradial or radiofemoral delay, all peripheral pulses are bilaterally and symmetrically palpable, respiratory rate was 56/min regular thoraco abdominal,temperature was 102 F, axillary,Blood Pressure was 160/100 mm Hg. He was pale. Icterus, cyanosis and clubbing was absent. He had bipedal oedema .Lymph nodes were not significantly enlarged and were not palpable. He had non scarring alopecia . There was a haemorrhage in nasal side of sclera in right eye. Rashes were present all over the body including malar rash, discoid rash, vasculitic rash.his nails were discoloured but clean and cut.

On Systemic examination the abdomen was distended, flanks were full ,Umbilicus was in midline, slit like, there was no organomegaly, fluid thrill was absent, shifting dullness present, normal bowel sounds were heard. Neurological examination on the second day of admission showed no focal neurological deficit. No cognitive impairment was found on psychiatric evaluation. Opthalmological examination was normal. Musculoskeletal examination showed that the right elbow joint was swollen, tender, warm, range of motion was limited in right elbow joint. Other joints were normal with range of movement within normal limit. Coarse crepitations in the chest was found . Cardiovascular examination was within normal limit Routine blood examination showed Haemoglobin 7.6 g/dl , Total Leukocyte Count 2900 /cu mm, Differential Leukocyte Count Neutrophil 76% Leukocyte 20% (580/cumm) Eosinophil 2% Monocyte 2% Basophil 0 Platelet 15000/cumm, Erythrocyte Sedimentation Rate 80(↑) ,Reticulocyte count- 2.5%, Peripheral Blood Smear- RBC normocytic normochromic, few hypochromic, no nRBC seen, WBC normal morphology, Platelets markedly reduced. PT 12.6s(N) APTT 29s(N) INR 0.97 , Liver Function Test :-Total Serum Bilirubin 0.3mg/dl(N), Direct Bilirubin 0.10mg/dl(N) Total Protein 5.8g/dl(↓), Albumin 1.6g/dl(↓), Globulin 4.2g/dl (↓), Aspartate transaminase 164u/l (↑), Alanine transaminase 86 u/l(↑), Alkaline Phosphatase 144u/l (N) GGT 141u/l(↑), Lactate dehydrogenase 484u/l (↑) , Blood Urea 36mg/dl(N), serum Creatinine 0.73mg/dl , serum sodium 127.6mg/dl, serum potassium 4.33mg/dl, ANA POSITIVE, ANTI ds DNA POSITIVE(>1000), pANCA positive , C3 21.10 mg/dl(↓),C4 <8mg/dl (↓).Direct & Indirect Coomb's Test was negative. Thyroid profile: T3 0.6 ng/ml(↓), T4 30 ng/ml(↓), TSH 1.52 µIU/ml(N). Lipid profile showed serum Cholesterol 106 mg/dl(↓), TGL 425 mg/dl(↑), HDL 12 mg/dl(↓) . Serum Ferritin >1000 ng/ml, CRP <6 mg/dl (N), Bone marrow study was normocellular . Urine routine showed pH 6.5, Specific Gravity 1.010, Glucose, Ketones and Bilirubin not detected, protein 4+, RBC plenty/HPF, Bacteria- not detected. Hyaline cast present, Urine Culture and Sensitivity was sterile, 24 hours urinary protein- 0.5 gm/24 hrs/L. CSF analysis was normal, culture and sensitivity was sterile, JE negative, Dengue- negative. Chest X ray was normal,X ray knee, elbow & ankle joint was normal (No erosive changes), USG abdomen showed bilateral renal parenchymal disease with ascites. MRI BRAIN was suggestive of advanced stage of CNS SLE, generalized atrophic changes in cerebral and cerebellar hemispheres, subtle FLAIR hyperintense lesion in right corona radiata without DWI restriction, suggestive of chronic ischaemic/ vasculitic lesions.

According to AMERICAN COLLEGE OF RHEUMATOLOGY REVISED CLASSIFICATION CRITERIA for SLE he had 1)Malar rash, 2)Discoid rash, 3)Oral ulcers, 4)Serositis- Peritonitis, 5)Arthritis.7/Renal involvement-Proteinuria, Microscopic

haematuria,8)NEUROLOGICAL- Seizure, Headache, postive MRI BRAIN findings, 9)HEMATOLOGICAL- Anemia, Thrombocytopenia , 10)IMMUNOLOGICAL ABNORMALITIES- Anti ds DNA Positive , 11)ANA positive

Patient was treated conservatively, PCV and Platelet transfusion was given. After rulling out infection methypredisone pulse therapy was given, Sunscreen are applied locally. However the patient expired due to advance couse of the disease.

DISCUSSION:

Systemic lupus erythematosus (SLE) is a chronic multisystem disease, occasionally life-threatening. [3] , The prevalence of SLE is found to be lower in males than in females, especially after puberty. [4] Additionally, gender may produce different characteristics in the manifestation of SLE. [5]. Male patients with SLE present mainly with renal involvement and seizures rather than photophobia and skin manifestations. The outcome also seems to be more serious in males[6].

In studies it has been found that men with lupus tend to have a similar frequency of occurrence of renal disease, skin manifestations, arthritis and central nervous system manifestations as women,but lesser prevalence of photosensitivity and higher prevalence of serositis. [7] Kasitanon N et al in their study that included 1378 patients with SLE have found that, with a median follow-up of 6.1 years, 118 patients died (8.6%). The overall cumulative probability of survival after disease diagnosis at 5, 10, 15 and 20 years was 95%, 91%, 85% and 78%, respectively. Based on a multivariate model, age at SLE diagnosis >50 years (hazard ratio = 5.9; P <0.001) and male gender (hazard ratio = 2.4; P = 0.004) were associated with poorer survival. [8] The major cause of death in the first few years of illness was active disease (e.g., central nervous system, renal or cardiovascular disease) or infection due to immunosuppression. [9] Our case is unique because it fulfils all the criteria(eleven) given by American College of Rheumatology (Revised Classification Criteria) for diagnosis of Systemic Lupus Erythematosus. It is very rare to find all the features of ACR criteria in a single patient . Thus our case presents as a rare case of disease that presents predominantly in females.

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