

Rare Presentation of SLE-Convulsion without Stroke before the Appearance of Malar Rash: A Case Study

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Abstract: We report a case of Systemic Lupus Erythematosus presenting with generalized tonic clonic convulsion without stroke before the appearance of malar rash which is indeed a very rare presentation. She recovered very well with DMARD's, anti epileptics, antibiotics, corticosteroids, immunosuppressants and Platelet transfusions. Systemic lupus erythematosus (SLE) is a heterogeneous, inflammatory, multisystem autoimmune disease in which antinuclear antibodies appear in serum often years before clinical symptoms. Lupus erythematosus describes the typical rash of SLE and the term systemic emphasizes the potential for multi-organ involvement. Diagnosing this disease early & providing appropriate treatment is important to achieve an optimal clinical outcome.

Key words Convulsion, Malar Rash, SLE

1. Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease in which organs and cells undergo damage initially mediated by tissue binding autoantibodies and immune complexes. Multiple autoantibodies are present. Prevalence of SLE in United States is 10 to 400 per 1,00,000 population¹. There is paucity of data in India. In India, SLE constitutes 1-2% of major rheumatological problems.

Kidney is involved in 30-50 % cases with asymptomatic proteinuria being the commonest feature. CNS involvement occurs in 10 % cases with stroke secondary to vasculitis being the commonest feature². However, a convulsion at presentation without stroke even before onset of malar rash is a rare presentation.

2. Case Report

A 28 years old female patient presented to emergency room with complaints of fever since 15 days and thrombocytopenia. Patient had three episodes of generalized tonic-clonic convulsions in emergency room.

On clinical examination, the patient was poorly built and nourished, with severe pallor. Patient was drowsy, febrile, responding to painful stimuli pulse was 96/min, regular. Blood pressure was 100/70mm of Hg. Cardiovascular

examination revealed tachycardia. Respiratory system examination showed right based crepitations. Per abdomen examination revealed mild splenomegaly (2cm below the left costal margin) but no hepatomegaly. Patient was intubated and mechanically ventilated in view of aspiration and repeated convulsions (status epilepticus). She was treated as a case of viral fever with thrombocytopenia with aspiration pneumonitis as a provisional diagnosis. After 2 days of admission, patient developed maculopapular erythematous rash over both cheeks.(Fig. 1)

Investigation revealed anaemia with pancytopenia. Details of investigations done during hospitalization are described in the table below (TABLE 1). Patient was treated as a case of SLE with CNS lupus with lupus nephritis with intravenous antibiotics, platelet transfusion, antiepileptics, DMARD's (Hydroxychloroquine for 4months) and pulse methylprednisolone (1 gm/day for 3 days) therapy followed by oral prednisolone(1mg/kg/day) and methotrexate (15mg weekly) for 6 months according to weight. Intravenous cyclophosphamide (500mg monthly) was given for 6 months.

Patients made a good recovery after immunosuppression with improvement in hematological profile. There was significant reduction in 24 hours proteinurea after 15 days. Patient was on ventilator for 20 days. Patient made a good recovery and was subsequently discharged home 30 days, post admission.

Patient was discharged on oral immunosuppression drugs and to be follow up for every month for intravenous cyclophosphamide (500mg monthly) for 6 months.

3. Discussion

The presentation of SLE in the form of convulsion without stroke before malar rash is indeed a very rare presentation which we have discussed in this case report. Systemic lupus erythematosus (SLE) is a heterogeneous, inflammatory, multisystem autoimmune disease in which antinuclear antibodies appear in serum often years before clinical symptoms³. SLE is rare in India. A prevalence study in India (carried out in a rural population near Delhi) found a point prevalence of 3 per 100,000⁴. It affects predominantly women in their reproductive years. The median age of onset in Indian SLE is 24.5 years and the sex ratio (F:M) is 11:1⁵. The prognosis of SLE is quite grim with more than half of the patients developing irreversible organ damage over time. Although the survival has improved in the west with modern treatment to the tune of 80% at 10 years after diagnosis⁶, the Indian figures are not so good (50%-60% survival at 10 years)^{7,8}. Possible reasons for poor survival in Indian SLE include delay in diagnosis, referral bias (only the most serious cases are referred by practitioners), suboptimal health care facilities and an inherently more severe disease (genetic factors?) and endemic tuberculosis to which the lupus patients are more susceptible. The major causes of death in the first few years after diagnosis include disease activity and infections. Late mortality i.e. 10 years after diagnosis, on the other hand, is mainly attributed to atherosclerotic vascular disease⁹.

Systemic lupus erythematosus (SLE) is a remitting and relapsing illness¹⁰. Symptoms and signs are often nonspecific, eg fatigue (can be severe and debilitating), malaise, fever, splenomegaly, lymphadenopathy, weight loss, arthralgia and fatigue, oral ulcers, photosensitive skin rashes, pleuritic chest pains, headache, paraesthesiae, dry

eyes and mouth. Raynaud's phenomenon, mild hair loss and myalgia^{11,12}

The CNS lupus nomenclature has been revised to catalog many manifestations. Because of the difficulty distinguishing causal SLE associations with some neurological symptoms, only seizure and psychosis were typically included in the diagnostic criteria. Seizures related to SLE may be generalized or partial and may precipitate status epilepticus. Psychosis may manifest as paranoia or hallucinations.^{13,14,15}

However, the American College of Rheumatology (ACR) created standardized case definitions and diagnostic testing recommendations for 19 neuropsychiatric syndromes in SLE, including seizures/seizure disorders and psychosis.¹⁶

Delirium represents a spectrum of fluctuating altered consciousness characteristic of SLE. Delirium may be caused by CNS vasculitis, encephalopathy, cerebritis, or the manifestations previously called organic brain syndrome. Aseptic meningitis, myelopathy, optic neuropathy or other demyelinating disorders may also require urgent evaluation.¹⁶

Migraine headaches may be linked to antiphospholipid syndrome. Headache and mood disorders may be the most commonly reported neurologic manifestation of SLE, but cause and effect may be difficult to distinguish.

There is a study done describing the cumulative percentage frequency of clinical manifestations in patients with SLE from different regions in India. It reveals a mean of 9 % & 11 % presenting as seizure and thrombocytopenia respectively in a case of SLE.¹⁷⁻²². Another study done by Wang et al in 2002²³ in Asia, revealed 22.8% & 22.9 % as neurological manifestation and thrombocytopenia respectively in patients diagnosed as SLE in comparison with 76.1% presenting with malar rash, 74% with nephropathy & 50.5% presenting with arthritis in the SLE patients.

Table 1

Date	19/10/13	26/10/13	28/10/13	29/10/13	30/10/13	5/11/13	8/11/13
Haemoglobin(gm%)	9.6	6.2	9.2	10.4	9.9	9.9	
Total Leucocyte Count(mm ³)	1700	1920	6030	3900	5300	9020	
Platelet(mm ³)	74000	70000	152000	220000	458000	628000	
Sodium(Meq/L)	150	148				133	
Potassium(Meq/L)	3.1	3.3				4.3	
Blood Urea(mg/dl)	25	42				35	
Creatinine(mg/dl)	1.2	1				0.7	
Bilirubin(total)(IU/L)	2.4	1.4					
Bilirubin(direct)(IU/L)	1.8	0.7					
SGOT(IU/L)	2648	166					
SGPT(IU/L)	519	141					
Alkaline Phosphatase(U/L)	199	121					
Urine-							
Albumin	2+			3+			
Sugar	Nil			Nil			
M/E(cells/hpf)	5-6 pus cells			3-4 pus cells			
INR	1.25						
24 hrs urinary protein(mg/24hrs)					2950		507

ANA(ratio)	2.32(ratio)						
ds DNA(IU/ml)	60 IU/ml						
CRP	Positive						
MRI Brain & Venogram	Normal Study						
2Decho		Normal					
Rapid malarial test	Negative	Negative					
Dengue(IgG,IgM,NS1Ag)	Negative	Negative					
Widal(Slide method)	Negative						
ESR(mm/hr)	40					18	

Figures



Day 2 after admission(ICU)

Figure 1



Shifted from ICU to Ward

Figure 2



Discharged from Ward & Hospital

Figure 3



During followup

Figure 4

4. Conclusion

Systemic Lupus Erythematosus(SLE) presenting as convulsion without stroke is a rare presentation and should be evaluated meticulously by early diagnosis and treatment. Hence, every young patient specially female in the reproductive age group presenting as fever,convulsion,malar rash with significant proteinuria / microalbuminuria should be evaluated for Systemic Lupus Erythematosus(SLE).

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