



SYSTEMIC LUPUS ERYTHEMATOSUS WITH CNS LUPUS IN THE FORM OF VASCULITIS, GROSS ASCITES, PLEURAL EFFUSION, ANEMIA OF CHRONIC DISEASE AND UTI

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ABSTRACT

Systemic lupus erythematosus (SLE) is a complex autoimmune disease with a varying clinical phenotype and prognosis from mild to life threatening. SLE needs to be differentiated from its mimickers sharing similar clinical and laboratory features, as Early management is crucial to prevent involvement of various organs. Also, it is important to establish the severity and potential reversibility of the illness and to estimate the possible consequences of various therapeutic interventions. Interventions including steroids, immunosuppressants, plasmapheresis, some biologics, and vaccination have shown efficacy in reducing mortality rates, though still higher than general population.

KEYWORDS: Lupus, autoimmune, prognosis, SLE, steroids, mortality.

CASE REPORT

A 20 year female presented with on and off fever for 2 months associated with headache, abdominal distension for one month without pain or diarrhea, dyspnea for ten days and right sided leg weakness for four days. The patient was previously fit without any history of any chronic disease or any ongoing long term treatment. Vitals were normal except for tachycardia. On general examination, pallor was found. Per abdominal examination showed abdominal distension and shifting dullness without any tenderness. USG abdomen confirmed the above findings. Chest X-ray showed small bilateral pleural effusion. Pleural Tap was exudative

while ascitic tap was low SAAG. Neurological examination was normal with no motor and cranial nerve deficit. MRI brain revealed Multiple patchy area of acute/early subacute lacunar infarct at bilateral cerebral hemisphere. Fundus was within normal limit. 2D echo and Color Doppler bilateral carotids were normal. TB IGRA test was negative. CBC showed normocytic normochromic anemia. Iron studies showed Anemia of Chronic Disease. Bilateral Color Doppler, done for right sided lower limb weakness was suggestive of subcutaneous edema. Further blood Investigations revealed:

PARAMETER	Patient value	NORMAL RANGE
Hb	7.5 gm/dl	11.0-15.0
TLC	10,510 /µl	4000-10000
DLC (N/L/E/M)	90/5	
PLATELET	165000/µl	150000-450000
MCV	97 fl	80-100
LDH	423 U/L	230-460
IRON/TIBC	45/ 129 µg/dl	58-158/250-425
Reticulocyte count	2.6%	0.5-2.5
RPI	1.6	
CREATININE	1.1 mg/dl	0.5-1.4
UREA	42 mg/dl	15-45 mg/dl
SGOT / SGPT	19/14 U/L	9-39/10-40
TOTAL PROTEIN/ ALBUMIN	6.2/2.08 g/dl	6.4-8.5/3.2-5.5

Urine		
ALBUMIN	+	Nil
SUGAR	Nil	Nil
RBC	Nil	NIL
WBC	40-60/ hpf	<5 / hpf
KETONE	Negative	Negative
24 Hour Urinary Protein	0.63mg/d	0.04-0.15
CSF Tap		
TLC	9	<5
RBC	0-1	
SUGAR	70	40-80
PROTEIN	39	10-40
ADA	2	<10
Autoimmune Panel		
ANA	183.48	<20
Anti-dsDNA	>150	<10
APLA Profile	Negative	
NMDA receptor/anti glutamate	Negative	
Anti GBM	2.73	<20
C3	25.7	90-180
C4	<8.00	10-40
p-ANCA	Negative	
c-ANCA	Negative	

A diagnosis of Systemic Lupus Erythematosus with CNS Lupus in the form of Vasculitis, gross ascites, pleural effusion, anemia of chronic disease and UTI was made. The patient was managed conservatively with immunosuppressants, anticoagulants, antibiotics and other supportive treatment. She is now asymptomatic in follow ups with fair general condition on mycophenolate.

DISCUSSION

Systemic lupus erythematosus (SLE) is an autoimmune disease with tissue-binding autoantibodies and immune complexes mediated damage. In most patients, even before the first clinical symptom appears, autoantibodies can be present for few years. Female sex is permissive for SLE with evidence of hormone effects, genes on the X chromosome and epigenetic differences between genders playing a role. Autoimmunity results due to complex interplay between genetic susceptibility, environment, gender, race and abnormal immune responses. Titres of anti-dsDNA vary over time. In some patients, increase in quantities of anti-dsDNA herald a flare, particularly of nephritis or vasculitis, especially when associated with declining levels of C3 and C4 complement. The prevalence of Neurological manifestations accounts for 60% of lupus cases. CNS lupus is generally seen as being angiopathic in origin, although other mechanisms can also contribute to CNS disease. Cerebral ischemia and infarction in SLE patients may result from (1) coagulopathy secondary to APLA syndrome, (2) Vasculitis (3) embolization from carotid artery plaque or (4) cardiogenic thromboemboli in Libman-Sacks disease. Two process can occur at once-vasculitis plus bland vascular occlusions in which it is

appropriate to treat with anticoagulation plus immunosuppression. Anti neuronal antibodies are detected in only 60% of CNS lupus. Mortality causes have been mainly grouped in disease activity, renal failure, infections in first decade and subsequently thromboembolic events. Of all deaths in SLE patients, 30–60% are due to infections.^[2,3] Risk factors for severe infections in SLE include low socioeconomic status, race, nephritis, anti-phospholipid syndrome, a high SLE disease activity (SLEDAI) score and the degree of immunosuppression.^[1-7] Interventions including steroids, immunosuppressants, plasmapheresis, some biologics, and vaccination have shown efficacy in reducing mortality rates, though still higher than general population.

CONCLUSION

Systemic lupus erythematosus (SLE) is a complex autoimmune disease with a varying clinical phenotype and prognosis from mild to life threatening. SLE needs to be differentiated from its mimickers sharing similar clinical and laboratory features. Timely intervention has led to significant reduction in mortality rates and end organ damage. Also, it is important to establish the severity and potential reversibility of the illness and to estimate the possible consequences of various therapeutic interventions.

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