



## A Rare Case of Systemic Lupus Erythematosus with Multiorgan Involvement in A Young Pregnant Women – A Case Report

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**Abstract:** Systemic Lupus erythematosus (SLE) is an autoimmune disease that notoriously varies in presentation, course, and outcome. Active Nephritis, especially if the neuropsychiatry component is involved, is associated with adverse pregnancy outcomes such as Gestational hypertension, pre-eclampsia, and renal failure. Our aim is to elaborate on the case report of young pregnant women diagnosed with SLE with multiorgan involvement about its course and management and the need for a multidisciplinary approach. A 20-year-old primigravida at 33 weeks of gestation with no known co-morbidities previously came with a history of fever for 2 days, abdominal pain, and bleeding per vagina. Given placental abruption, the patient underwent an emergency lower segment cesarean section. The patient developed postpartum hemorrhage (PPH), which was managed medically. On POD#8 (postoperative day 8), the patient had generalized tonic-clonic seizures (GTCS) and developed hematuria. On evaluation, it revealed abnormal renal parameters suggestive of Pregnancy-related Acute Kidney Injury (PRAKI). The patient's condition did not improve with conservative management. After immunological investigations, a renal biopsy was done, and the diagnosis was confirmed as SLE-LN III (Systemic lupus erythematosus-Lupus nephritis) with Neuropsychiatry component involvement. Hence, the patient was started on Hydroxychloroquine based on the National Institute of Health protocol, and her condition improved. Hence given the relatively high number of maternal and neonatal deaths, lupus pregnancies should be followed by a multidisciplinary team, including nephrologists, rheumatologists, and obstetricians experienced in high-risk pregnancies, and deliveries should be planned in tertiary-care settings provided with neonatology intensive care units.

**Keywords:** Systemic lupus erythematosus, Pre-eclampsia, Pregnancy induced acute kidney injury, autoimmune disease, Lupus Nephritis, Neuropsychiatry component.

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**I. INTRODUCTION**

Systemic Lupus erythematosus is an autoimmune disease with a complex pathogenesis resulting from interactions between susceptibility genes and environmental factors. SLE and pregnancy exert mutual effects; pregnancy and labor harbor a significant risk for exacerbation of SLE<sup>1</sup>. Its course is characterized by periods of remission and relapse. However, the exact influence of pregnancy on SLE is still unclear, with reported rates of lupus flare in recent studies ranging from 13.5% to 65%<sup>1</sup>. Findings may be confined initially to one organ system; others become involved later. Instead, the disease may first be multisystem. Frequent findings are malaise, fever, arthritis, rash, pleuro-pericarditis, photosensitivity, anemia, and cognitive dysfunction. At least half of patients have renal involvement. SLE may also be associated with decreased attention, memory, and reasoning. Identification of antinuclear antibodies (ANA) is the best screening test. However, a positive result is not specific to SLE. Antibodies to double-stranded DNA (dsDNA) and Smith (Sm) antigens are relatively specific for SLE, whereas other antibodies are not. Women with SLE are at greater risk for thrombotic complications, infection, postpartum hemorrhage, and blood transfusion. Higher fetal loss rates, preterm delivery, and fetal growth restriction<sup>2</sup> characterize fetal outcomes in lupus pregnancy. Newly diagnosed SLE in pregnancy tends to be even more severe. Here is a case report of SLE, diagnosed for the first time in pregnancy.

**2. CASE REPORT**

20-year-old primigravida, immunized, on irregular follow-up, presented at 33 weeks of gestation with no known medical history previously, with complaints of fever for 2 days, sudden onset abdominal pain, and bleeding per vaginum. The patient had no surgical interventions in the past and no history of known drug allergies. On Examination: General condition fair, Afebrile, Pallor ++, No Icterus, Bilateral pitting pedal edema (grade 2). On Systemic examination: Per abdominal examination (P/A): Uterus corresponding to 32-34 weeks, hypertonic, tense and tender, cephalic, Fetal heart rate (FHR) was good. Per speculum examination (P/S): Bleeding through os+. On per vaginal examination (P/V): Cervix soft, Uneffaced, os admits tip of a finger. Vitals: BP: 140/100mmHg (Tab.Labetalol 100mg stat given) Pulse rate:90bpm, regular. Respiratory rate:18/min. After placental abruption, the patient underwent an emergency lower segment cesarean section under general anesthesia. Bleeding time and clotting time - preoperatively was normal, Urine protein 3+.

**2.1. Intraoperatively**

Baby delivered in cephalic presentation, faint cry+, with an APGAR score of 3/10 and 6/10 at 1 min and 5 mins, respectively.180 grams of retroplacental clots +, same removed. 1-unit packed red blood cells (PRBC) and 1-unit fresh frozen plasma (FFP) transfusion done.

**2.2. Postoperatively**

- 1-unit PRBC and 1-unit FFP were transfused postoperatively.
- Then, 2 hours after the surgery, the patient went into PPH and was managed medically along with balloon catheter tamponade.
- Blood Pressure charting showed elevated BP (160/110mmHg- 150/100mmHg). Hence MgSO4 was given as per the Zuspan regimen, and the patient was also started on anti-hypertensives.
- Serial BP monitoring was done, and blood pressure was maintained—input-output charting done—Adequate.
- Patients' hematological and biochemical parameters are shown in Table I
- On POD#6, a Fundus examination revealed no evidence of hypertensive retinopathy.
- POD#8: The patient had one episode of GTCS, managed with Magnesium sulfate according to the Zuspan regimen. Vigilant monitoring of vitals and urine output was done.
- MRI brain - Normal.
- POD#9: The patient had frank hematuria. Ultrasound abdomen was suggestive of bilateral renal parenchymal disease.
- Serial monitoring revealed abnormal renal parameters suggestive of Pregnancy-related Acute kidney Injury (PRAKI). As a result, a nephrology opinion was obtained, and appropriate treatment was administered.
- On POD#13, the patient had high-grade fever associated with chills, rigor, and vomiting (Non-Bilious/Non-blood stained). 24hr Urine protein: 4241 mg/day, Anti-nuclear antibody (ANA) -Positive, showed a speckled pattern, LIA-17 (line-immunoassay) showed smith antigen positive, RNP (Ribonucleoprotein)- Positive and SSA (Sjogren syndrome antibody) - Positive.
- Antiphospholipid antibody profile (APLA) showed negative for anti-cardiolipin antibodies and beta 2 glycoproteins.
- The patient underwent renal biopsy, suggesting SLE-Lupus Nephritis III with neuropsychiatry component (NP) involvement. The immunofluorescence report is shown in Table 2.
- Patient was advised Tab.Hydroxychloroquine, Tab. Enalapril, Inj.Cyclophosphamide according to National Institute of health (NIH) protocol.
- Treatment options for SLE-LN III with NP manifestations were explained:
- Mycophenolate mofetil (MMF) Vs. Cyclophosphamide
- B) Costs/Risks and benefits of each explained.
- As the patient opted for MMF, Tab.MMF 500MG twice daily was advised.
- Need for contraception explained. An iron-rich diet with a fluid restriction of 2-2.5L/day was advised.
- The patient was followed up regularly, and her condition improved.

Table I: Hematological and biochemical parameters in the postoperative period							
	POD 1	POD 3	POD 7	POD 9	POD 10	POD 11	POD 12
Hemoglobin	9.2	9		9			8.4
Platelet	1.67		2.89	2.76			2.71
Peripheral Smear	MCHC			MCHC			
Bt			2'30"	2'33"			

Ct		4'42"	4'35"		
Sgot/Sgpt	NORMAL	NORMAL		NORMAL	NORMAL
Serum Electrolytes	NORMAL	NORMAL		NORMAL	NORMAL
Urine Albumin	1 +	2+		3+	3+
Serum Creatinine	0.6	1.2	1.4	1.2	1.7

GTCS	Frank hematuria	Pregnancy-related acute kidney injury
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**Fig 1: Intraoperative finding of placental abruption**



**Fig 2: Patient presented with frank hematuria on postoperative day 9**

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<b>IMPRESSION:</b> IgM, IgG, IgA, C1q, C3 shows positivity in capillary wall and mesangium. Kappa and lambda shows positivity					
CAPILLARY WALL					
CONJUGATES	CAPILLARY WALL		MESANGIUM	TUBULES	BLOOD VESSELS
	GRANULAR	LINEAR			
IgM	Positive	Negative	Positive	Negative	Negative
IgG	Positive	Negative	Positive	Negative	Negative
IgA	Positive	Negative	Positive	Negative	Negative
C1q	Positive	Negative	Positive	Negative	Negative
C3	Positive	Negative	Positive	Negative	Negative
Fibrinogen	Negative	Negative	Negative	Negative	Negative
Kappa	Positive	Negative	Negative	Negative	Negative
Lambda	Positive	Negative	Negative	Negative	Negative
Albumin	Negative	Negative	Negative	Negative	Negative

Pathologist: DR. BARATHI DR. SUBALAKSHMI DR. PONNEYIN CHELVI

**Fig 3: Immunofluorescence report IgM, IgG, IgA, C1q, C3 shows positivity in capillary wall and mesangium.**

Kappa and Lambda show positivity only in the granular capillary wall.

**Table 2: Clinical and laboratory manifestations seen in this patient<sup>2</sup>.**

Criteria	Definition
1. Malar rash	Fixed erythema, flat or raised over the malar eminences
2. Discoid rash	Erythematous raised patches with adherent keratotic scaling and follicular plugging
3. Photosensitivity	Skin rash as a result of unusual reaction to sunlight
4. Oral ulcers	Oral or nasopharyngeal ulceration, usually painless
5. Non-erosive arthritis	Involving 2 or more peripheral joints, characterized by tenderness, swelling or effusion
6. Pleuritis or Pericarditis	Pleuritis- convincing history of pleuritic pain Pericarditis- documented by ECG
7. Renal disorder ★	a) Persistent proteinuria >0.5g/day or >3 + or b) Cellular casts
8. Neurological disorder ★	a) Seizures or b) Psychosis
9. Hematological disorder ★	a) Hemolytic anemia with reticulocytosis or b) Leucopenia or c) Lymphopenia or d) Thrombocytopenia
10) Immunological disorder ★	a) Anti-DNA Ab or b) Anti-Sm Ab or c) Positive finding for APLA
11) Positive Antinuclear antibody ★	Abnormal ANA titre by immunofluorescence

In our case, the patient presented with 5/11 criteria for diagnosing systemic lupus erythematosus, which includes renal, neurological, hematological, immunological manifestations, and positive anti-nuclear antibody.

### 3. DISCUSSION

Systemic Lupus Erythematosus is an autoimmune connective tissue disorder that affects various organs and systems diagnosed between 20 and 40 years of age. Most commonly encountered during pregnancy and puerperium<sup>1</sup>. It is suspected in the presence of clinical characteristics and

confirmed by laboratory detection of immunological abnormalities. It is considered a Th2 cytokine-driven disease due to the overexpression of Th2-type cytokines<sup>3</sup>. The incidence of SLE flare in pregnancy is 10-58%<sup>1</sup>. Pregnancy loss is more likely if SLE is diagnosed during the index pregnancy. Four risk factors for pregnancy loss can be determined by routine testing early in pregnancy by using the acronym PATH

(Proteinuria, Antiphospholipid syndrome, Thrombocytopenia, and Hypertension) to remember these important risk factors easily. These data suggest that these risk factors independently increase the risk of pregnancy loss severalfold over other pregnant women with lupus<sup>4</sup>. Pre-pregnancy counseling enables women to commence pregnancies with quiescent disease<sup>5</sup>. Lupus nephritis is due to renal deposition of Immune complexes like Immunoglobulin G antinuclear autoantibodies against DNA and nucleoprotein in glomeruli leading to complement activation and inflammatory tissue damage in the kidney. It has been strongly associated with Interferon-gamma overproduction<sup>6</sup>. The most common presentation is Proteinuria, hematuria, pyuria, and urinary casts; this may help distinguish it from pre-eclampsia<sup>7</sup>. Renal biopsy is necessary to confirm the diagnosis of LN, and determining renal histology is key to understanding prognosis and providing direction for appropriate treatment<sup>6</sup>. Women with lupus nephropathy face severe pregnancy challenges, including deterioration of renal function and progression of underlying renal diseases. Predictors of renal flare: Plasma creatinine >1.2 mg/dl Proteinuria >= 500mg in 24 hrs. The presence of red blood cell casts is attributable to active nephritis. There should be accompanying signs suggesting glomerulonephritis or interstitial nephritis; hence in the complete absence of Proteinuria, hematuria solely related to active nephritis is unlikely<sup>8,9</sup>. In the presence of LN, in past studies, fetal losses ranged from 8 to 36%, miscarriages between 4 and 31%, and stillbirths or neonatal deaths between 4 and 23%<sup>[10-12]</sup>.

### 3.2. Management

Ist line therapy: Glucocorticoids, Specific immunosuppressive treatment regimens according to renal biopsy results. Cytotoxic agents such as Cyclophosphamide, Mycophenolate mofetil in moderate to severe Degenerative glomerulonephritis<sup>13</sup>. Refractory or relapse to standard therapy- Biological agents such as Rituximab can be used<sup>14</sup>. SLE patients with active lupus nephritis are at higher risk for pregnancy complications than those without renal disease. Therefore, they should be advised against pregnancy until a renal remission of at least 6 months, if not 12– 18 months<sup>15</sup>. Lupus cerebritis with an incidence of 10-35% in SLE patients with neurologic and psychiatric manifestations. These manifestations should be considered in patients with altered mental state (AMS), headache, anxiety, sadness, or unexplained psychosis. These can happen shortly after or even before the diagnosis of SLE. The variety of lupus cerebritis symptoms makes diagnosis difficult. Establishing a diagnosis of lupus cerebritis requires the identification of SLE using serologic markers and biopsy, the exclusion of other potential causes of nervous system dysfunction, and other factors<sup>16</sup>.

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## 4. CONCLUSION

Pre-eclampsia and SLE flares share hypertension, Proteinuria, edema, and renal function deterioration features. However, the management is distinct. Renal flares during pregnancy and postpartum are not uncommon and should be recognized and treated adequately. It may be difficult, if not impossible, to discriminate lupus nephropathy from severe preeclampsia if the kidney is the only involved organ. Pre-eclampsia is more likely in patients with antiphospholipid syndrome, a history of pre-eclampsia, or those with hypertension, LN, or diabetes mellitus. Both conditions can present with hypertension, Proteinuria, edema, and renal function deterioration and may co-exist in the same patient. The distinction is important as management is vastly different. While treatment with steroids is mandatory in active LN, steroids will typically aggravate pre-eclampsia. In contrast to active lupus, Proteinuria also declines rapidly with the baby's delivery in pre-eclampsia. In the most severe and confusing cases, a correct diagnosis may be made only by renal biopsy. Unfortunately, maternal and fetal well-being concerns may have prompted delivery by that time. Hence given the relatively high number of maternal and neonatal deaths, lupus pregnancies should be followed by a multidisciplinary team, including nephrologists, rheumatologists, and obstetricians experienced in high-risk pregnancies, and deliveries should be planned in tertiary-care settings provided with neonatology intensive care units.

## 5. ETHICAL APPROVAL STATEMENT

The patient obtained consent for this case report. Additionally, written informed consent was obtained from the individual to publish any potentially identifiable images or data in this article.

## 6. AUTHORS CONTRIBUTION STATEMENT

Dr. Nithya.R, Assistant Professor, Department of OBG, and Dr. Idhalya Ravi, Junior resident, were on duty when the patient presented to us in the OBG outpatient department. Dr. Idhalya investigated, followed the case, collected data, and discussed it with Dr. Nithya. Dr. Nithya performed an emergency cesarean section for this patient. Postoperatively, they analyzed and interpreted patient data and were major contributors to writing the manuscript. Dr. Bharathi and Dr. Subulakshmi performed an interpretation of the pathological data. All authors contributed to the article and approved the submitted version.

## 7. CONFLICT OF INTEREST

Conflict of interest declared none.

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