

Young Girl with Weight loss, Prostration and Extreme Fatigue

DR N N Anand^{1*}, DR Karthikeya TM², DR Divya Sriramulu³

¹Professor & HOD, Department of general medicine, Sree Balaji Medical College and Hospital, Chromepet, Tamil Nadu, India.
dmaganand1971@gmail.com

²Assistant Professor, Department of general medicine, Sree Balaji Medical College and Hospital, Chromepet, Tamil Nadu, India

³Senior resident, Department of general medicine, Sree Balaji Medical College and Hospital, Chromepet, Tamil Nadu, India

KEYWORDS

joint pains,
hyperpigmented
rash, SLE, anti ds
DNA, proteinuria,
steroids,
Mycophenolate
mofetil (MMF)

ABSTRACT

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease that can affect multiple organ systems, particularly the kidneys, blood cells, skin, and musculoskeletal system. This report presents a case of a 16-year-old female diagnosed with lupus presenting with major organ involvement.

1. Case Presentation

A 16-year-old female presented to the outpatient department with complaints of fatigue, joint pain, rash over face, neck and arms, she had history of fever 4 months ago which lasted for 1 month low grade, intermittent after which she started developing loss of appetite, loss of hair, loss of weight, joint pains in bilateral knee and hip joints which had worsened over the past few months leading to stiffness and inability to walk and squat. No history of cough, cold, breathlessness. No history of loose stools, constipation. No history of recurrent fever during childhood. No known comorbidities. No significant family history



Figure 1: showing non scarring alopecia

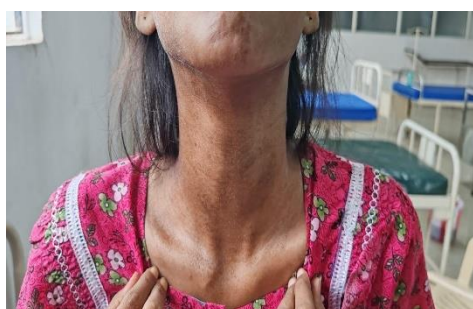


Figure 2: hyperpigmented scaly plaques over forehead, nape of the neck, arms

2. Clinical Examination

Vital signs: Stable.

Skin: hyperpigmented scaly plaques over forehead, nape of the neck, arms with follicular plugs [photosensitivity present], non scarring alopecia. Oral mucosa-normal, palms and soles-normal

Musculoskeletal system: Joint tenderness in the bilateral knees and hips, flexion contracture in bilateral knees

Hematological system: Laboratory tests revealed pancytopenia (anemia, leukopenia, and thrombocytopenia).

Systemic examination was normal

3. Laboratory Findings

Complete blood count (CBC): Hemoglobin 5.4 g/dL, leukocytes $2.4 \times 10^3/\mu\text{L}$, platelets $85 \times 10^3/\mu\text{L}$ (pancytopenia).

Peripheral smear- pancytopenia

LDH: 569 U/L (high)

Direct Coomb's test- positive

ESR >140

Triglycerides - 555 mg/dL

Urinalysis: Significant proteinuria (920 mg/day), urine PCR- 3076 mg/g

Urea- 49 mg/dL, creatinine- 1.5 mg/dL, uric acid- 9.8 mg/dL

Serological markers: ANA-Positive (speckled pattern), ANA profile- nucleosome, RNP 68kD/A/C, Sm/RNP are positive. dsDNA, Histones, Sm are equivocal.

C3 - 0.6g/L and C4 - 0.14g/L (low)

Iron studies - normal, serum ferritin- 621ng/dL elevated

Imaging Studies:

USG abdomen- hepatosplenomegaly, moderate ascites

4. Discussion

Based on the clinical presentation, laboratory findings the patient was diagnosed with Systemic Lupus Erythematosus (SLE) with multi-organ involvement with pancytopenia, dermatological manifestations (rash and alopecia), and musculoskeletal involvement (knee and hip joint involvement). Rheumatology opinion was obtained and advised to ROM exercises {quadriceps -static and dynamic}, Tablet mycophenolate mofetil, Tablet Medrol, Tablet HCQS, Tab Losartan and protein diet. Nephrology opinion taken and advised Inj.EPO and planned for renal biopsy once haemoglobin improves. Dermatology opinion obtained advised topical steroids and sunscreen.

5. Treatment

1. Therapy for SLE:

Oral prednisone (1 mg/kg/day).

Mycophenolate mofetil (MMF) 1.5 g/day as a steroid-sparing agent.

Angiotensin-converting enzyme (ACE) inhibitors for renal protection due to proteinuria.

2. Management of Pancytopenia:

- Careful monitoring of blood counts. Hematology consultations advised supportive care with transfusions as needed.

3. Dermatological and Musculoskeletal Treatment:

-Hydroxychloroquine (200 mg/day) was initiated for skin and joint manifestations.

- NSAIDs for joint pain and inflammation.

4. Preventive Therapy:

- Calcium and Vitamin D supplements were given to reduce steroid-induced osteoporosis risk.
- Proton pump inhibitors to prevent gastric complications due to steroid therapy.

5. Follow-up:

- Close follow-up with regular monitoring of renal function, urine protein, CBC, and complement levels. Patient is planned for renal biopsy.

Outcome:

After 2 weeks of treatment, the patient showed significant improvement. Proteinuria reduced to <0.3 g/day, CBC improved, and the rash resolved. However, she continued to have mild joint pain, managed with NSAIDs and hydroxychloroquine.

6. Conclusion:

This case highlights the complexity of managing SLE in adolescents, where multi-organ involvement such as lupus nephritis and pancytopenia can pose significant treatment challenges. Timely initiation of immunosuppressive therapy, close monitoring of blood counts, and managing side effects of therapy (e.g., steroids) are key aspects of treatment. This case underscores the importance of timely diagnosis and tailored therapeutic interventions in young patients with severe multi-organ SLE, emphasizing renal and hematological monitoring.

References:

- [1] Costenbader, K. H., et al. (2011). "Systemic lupus erythematosus and lupus nephritis." *Lupus Science & Medicine*, 2(1), e000058. [https://doi.org/10.1136/lupus-2021-000058] (https://doi.org/10.1136/lupus-2021-000058)
- [2] Hahn, B. H., et al. (2012). "Management of lupus nephritis: a systematic literature review." *Lupus*, 21(2), 147–156. [https://doi.org/10.1177/0961203311426568] (https://doi.org/10.1177/0961203311426568)
- [3] Bertias, G. K., et al. (2012). "Joint European League Against Rheumatism and European Renal Association guidelines on lupus nephritis." *Annals of the Rheumatic Diseases*, 71(11), 1771–1782. [https://doi.org/10.1136/annrheumdis-2012-201460] (https://doi.org/10.1136/annrheumdis-2012-201460)
- [4] Yap, D. Y. H., & Lai, K. N. (2015). "Cytopenia in Systemic Lupus Erythematosus." *Lupus*, 24(2), 127–134. [https://doi.org/10.1177/0961203314554115] (https://doi.org/10.1177/0961203314554115)
This case report emphasizes the need for a multidisciplinary approach and individualized treatment in lupus patients, especially in adolescents.
- [5] Kaul, A., et al. (2016). "Systemic lupus erythematosus." *Nature Reviews Disease Primers*, 2(16039), 1–21. [https://doi.org/10.1038/nrdp.2016.39] (https://doi.org/10.1038/nrdp.2016.39)
- [6] Gatto, M., et al. (2013). "Lupus nephritis: clinical presentations and outcomes in the 21st century." *Rheumatology (Oxford)*, 52(12), 1933–1943. [https://doi.org/10.1093/rheumatology/ket293] (https://doi.org/10.1093/rheumatology/ket293)
- [7] Tsokos, G. C. (2011). "Systemic lupus erythematosus." *The New England Journal of Medicine*, 365(22), 2110–2121. [https://doi.org/10.1056/NEJMr1100359] (https://doi.org/10.1056/NEJMr1100359)
- [8] Schwartz, N., et al. (2013). "Lupus nephritis: mechanisms, biomarkers, and outcomes." *Nature Reviews Nephrology*, 10(5), 258–268. [https://doi.org/10.1038/nrneph.2014.35] (https://doi.org/10.1038/nrneph.2014.35)
- [9] Mok, C. C. (2016). "Current role of mycophenolate mofetil in the management of lupus nephritis." *Clinical and Experimental Nephrology*, 20(4), 403–411. [https://doi.org/10.1007/s10157-016-1241-1] (https://doi.org/10.1007/s10157-016-1241-1)
- [10] Dooley, M. A., et al. (2010). "Mycophenolate versus azathioprine as maintenance therapy for lupus nephritis." *The New England Journal of Medicine*, 363(20), 1886–1896. [https://doi.org/10.1056/NEJMoa1006543] (https://doi.org/10.1056/NEJMoa1006543)
- [11] Cervera, R., et al. (2009). "Systemic lupus erythematosus: clinical and immunologic patterns of disease expression in a cohort of 1,000 patients." *Medicine (Baltimore)*, 78(3), 167–175. [https://doi.org/10.1097/00005792-199905000-00003] (https://doi.org/10.1097/00005792-199905000-00003)
- [12] Tektonidou, M. G., et al. (2015). "EULAR recommendations for the management of systemic lupus erythematosus with

- neuropsychiatric manifestations and lupus nephritis." *Annals of the Rheumatic Diseases*, 74(2), 245–254. [<https://doi.org/10.1136/annrheumdis-2013-204620>] (<https://doi.org/10.1136/annrheumdis-2013-204620>)
- [13] Petri, M., et al. (2012). "Impact of systemic lupus erythematosus on long-term health outcomes." *Current Opinion in Rheumatology*, 24(5), 505–511. [<https://doi.org/10.1097/BOR.0b013e328356b6bc>] (<https://doi.org/10.1097/BOR.0b013e328356b6bc>)
- [14] Koutsokeras, T., & Healy, T. (2014). "Systemic lupus erythematosus and lupus nephritis." *Nature Reviews Drug Discovery*, 13(3), 173–174. [<https://doi.org/10.1038/nrd4164>] (<https://doi.org/10.1038/nrd4164>)