Initial Presentation of Lupus Nephritis complicated by PRES and ITP with Multi-System Involvement

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The simultaneous presentation of lupus nephritis, immune thrombocytopenia (ITP), and posterior reversible encephalopathy syndrome (PRES) as the initial manifestation of systemic lupus erythematosus (SLE) represents a severe, high-activity disease state with significant risk for morbidity and mortality. PRES in SLE is strongly linked to active nephritis and hypertension, while concurrent thrombocytopenia increases the risk of intracranial hemorrhage. This triad reflects profound endothelial dysfunction and systemic disease activity, necessitating early recognition in patients presenting with seizures, renal dysfunction, and cytopenias. Prompt initiation of aggressive immunosuppressive therapy with supportive management for PRES is critical to preventing irreversible organ damage and improving survival.

- Previously healthy young adult woman presented with abdominal pain, nausea, vomiting, and diarrhea. Workup revealed urinary tract infection, acute kidney injury, and anemia that did not improve despite initial therapy, ultimately requiring transfusion of one unit PRBCs.
- Autoimmune evaluation revealed ANA+, anti-dsDNA+, low C4; renal biopsy confirmed class IV/V lupus nephritis.
- During a prolonged admission, the patient developed pneumonia, pleural effusions, worsening renal failure, and seizures secondary to MRI-confirmed posterior reversible encephalopathy syndrome (PRES). She was also diagnosed with immunethrombocytopenic purpura (ITP) secondary to SLE, prompting empiric plasma exchange.
- Treatment included high-dose corticosteroids, mycophenolate, belimumab, and multi-drug antiepileptic therapy. The patient stabilized, was discharged on biologic therapy, but ultimately progressed to end-stage renal disease requiring kidney transplantation.

Overview

Case Presentation

- Previously healthy young adult woman with **two weeks of abdominal pain, nausea, vomiting, and diarrhea**.
- Initial labs: **anemia, urinary tract infection, acute kidney injury (AKI)**.
- **Creatinine and blood pressure remained elevated** despite fluids and antihypertensives.
- **One unit PRBC transfusion** given for worsening anemia.

Autoimmune Workup & Diagnosis

- **Serology:** ANA 1:1640, anti-dsDNA >300 IU/mL, multiple extractable-nuclear antigens, low C4.
- **Kidney biopsy:** Diffuse proliferative and membranous lupus nephritis (class IV/V) with crescents.

Hospital Course

- **Immunosuppressive therapy:** High-dose IV methylprednisolone → oral prednisone, mycophenolate mofetil, and belimumab (cyclophosphamide avoided after fertility counseling).
- Complications: **pneumonia, pleural/pericardial effusions, dialysis-dependent renal failure**.
- **Neurologic event:** Seizures; MRI confirmed PRES. Managed with **levetiracetam, lacosamide, zonisamide**.
- Progressive **thrombocytopenia with schistocytes** → empiric plasma exchange for suspected TTP; ultimately diagnosed as **ITP secondary to SLE**.

Discharge & Outcome

- Discharged on **hydroxychloroquine, mycophenolate, prednisone, and antiepileptics**.
- Ultimately required hemodialysis given progressive renal insufficiency
- **Patient current on renal transplant list awaiting donor**

Discussion

Extraordinary triad presentation: Concurrent lupus nephritis, PRES, and ITP at initial SLE diagnosis is exceptionally rare; literature reports only sporadic combinations of two, with all three together poorly characterized.

Unifying mechanism: Widespread immune-complex–mediated endothelial injury underpins pathogenesis.

- Renal: Complement activation drives proliferative glomerulonephritis and progressive hypertension.
- Cerebral: Hypertension and autoantibody-mediated dysregulation of cerebrovascular autoregulation predispose to vasogenic edema (PRES).
- Hematologic: Dysregulated humoral immunity promotes antibody-mediated platelet destruction (ITP).

Clinical imperatives:

- Rapid renal decline and uncontrolled hypertension mandate early biopsy and potent immunosuppression.
- Therapeutic strategy must balance efficacy with long-term toxicity; fertility considerations can preclude cyclophosphamide, necessitating alternatives (e.g., mycophenolate, belimumab).
- Vigilant multisystem monitoring allows timely detection and treatment of complications (e.g., MRI for PRES to prevent permanent deficits).
- Differentiating ITP from thrombotic thrombocytopenic purpura is challenging; concurrent hematology evaluation and empiric plasma exchange may be warranted until clarity is achieved.

Outcome and significance: Despite multidisciplinary management, the patient progressed to irreversible renal failure requiring transplantation—illustrating both the devastating potential of fulminant SLE and the lifesaving role of transplantation in end-stage disease.

- This patient's presentation with simultaneous fulminant nephritis, extensive PRES, and refractory ITP represents an extremely rare and aggressive form of new-onset SLE.
- The triad reflects severe immune-complex endothelial injury spanning renal, neurologic, and hematologic systems.
- Early interventions—biopsy-guided immunosuppression, fertility-preserving regimens, neurologic monitoring, and empiric plasma exchange—were critical for survival.
- Despite maximal therapy, the patient ultimately required kidney transplantation, underscoring the severity of disease.
- Clinicians should maintain a high index of suspicion for SLE in young patients with multisystem involvement and ensure coordinated multidisciplinary care to prevent irreversible morbidity.

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