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Letter to the Editor

Postpartum Psychosis Masking the Diagnosis of Neuropsychiatric Lupus

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Dear Sir/Madam,

Systemic Lupus Erythematosus (SLE) is a chronic, relapsing, and remitting systemic disease. Autoimmunity and protean clinical manifestations affecting almost all the systems are the hallmarks of SLE. Systemic lupus can affect the central nervous system. The severity of CNS manifestations varies from less severe subclinical neurocognitive dysfunction affecting memory, intellect, and learning to more severe manifestations such as seizure, stroke, or transverse myelitis [1]. The psychotic features of primary psychiatric disorders overlap with Neuropsychiatric SLE (NPSLE) and often mask and delay the diagnosis of NPSLE.

We present a 27-year-old female who presented 3 weeks after her delivery with decreased sleep, slurring of speech and increased anxiety. She had associated bilateral leg pain and disturbed memory. She was evaluated and treated with postpartum psychosis. During the course of treatment, she developed persistent fever, generalized weakness, and vomiting followed by an erythematous rash over the lip and multiple petechiae over the left elbow and forearm. The rashes extended to the neck and breast along with new scalp lesions. There was associated significant hair loss. In spite of treatment, the symptoms worsened day by day. She had paranoid thoughts and an episode of violent behavior with irrelevant speech and fecal incontinence. She had typical psychotic features like paranoid delusions, auditory hallucinations along persistent insomnia. Later on, she developed multiple joint pains along with lower-limb edema. She denied any significant past or family history of rashes and joint pains.

General examination showed mild pallor along with significant alopecia. There were healed hyper-pigmented macules seen in elbows, scalp, and both knees. She was febrile without any obvious synovitis in any of the joints. The chest was clear without any added sounds. Heart sounds were regular without any murmurs. The abdomen was soft without any organomegaly. She was drowsy but there were no focal neurological deficits.

Hemogram revealed mild anemia (Hemoglobin 9.4 gm/dl) with elevated erythrocyte sedimentation rate (ESR 67mm/hr) and positive C-reactive protein. Antinuclear antibody (ANA 6.6) and Anti double-stranded DNA (Anti ds DNA 1.8) were positive. She had low complement levels [C3 level was 0.126 gm/dl (0.17-0.38); C4 level was 0.416gm/dl (0.89-1.87)]. 24-hour urine protein estimation was within normal range (80mg/day).

The patient was treated with intravenous methylprednisolone and initiated on Mycophenolate Mofetil (MMF) along with psychiatric medications. She survived the acute attack and maintained in remission with MMF and steroids along with antipsychotics (trifluoperazine, trihexyphenidyl, escitalopram).

It is always a diagnostic dilemma to distinct between NPSLE (formerly known as lupus cerebritis) from primary psychiatric disorders. American College of Rheumatology (ACR) has classified neuropsychiatric manifestations of SLE (Table 1 and Table 2) into cognitive dysfunction, cerebrovascular disease, seizures, psychosis, and peripheral nervous system disorders [2,3]. NPSLE can manifest in the absence of either serologic activity or other metabolic disorders. Cognitive decline in SLE is attributed to blood-brain barrier integrity dysfunction due to auto-antibodies [4].

Risk factors for NPSLE include [5]

Table 1: Case definitions-central nervous system manifestations of NPSLE [3].

1	Aseptic Meningitis
2	Cerebrovascular disease
3	Demyelinating syndrome
4	Headaches [migraines and benign intracranial hypertension]
5	Movement disorders [chorea]
6	Myelopathy
7	Sseizure disorders
8	Acute confusional states
9	Anxiety disorders
10	Cognitive dysfunction
11	Mood disorders
12	Psychosis

Table 2: Case definitions-peripheral nervous system manifestations of NPSLE [3].

1	Acute Inflammatory Demyelinating Polyneuropathy (Guillaine Barre Syndrome)
2	Autoimmune disorder
3	Mononeuropathy
4	Myasthenia gravis
5	Neuropathy, cranial
6	Plexopathy
7	Polyneuropathy

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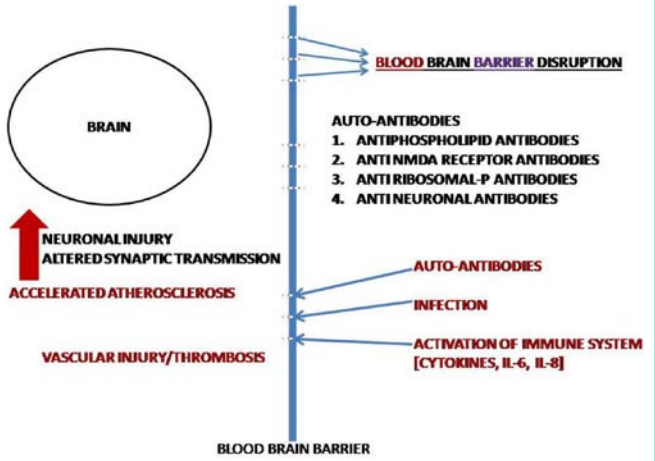

Figure 1: Pathogenesis of NPSLE.

- Persistently positive, moderate-to-high titers of antiphospholipid antibodies.
- Previous or concurrent major NPSLE events.
- Increased SLE activity or damage.

Auto-antibodies are involved in the pathogenesis of neuropsychiatric lupus (Figure 1). Auto-antibodies like antiphospholipid antibodies, anti-ribosomal-P antibodies, anti-NMDA receptor antibodies, and anti-neuronal antibodies enter the brain through the disrupted blood-brain barrier [4]. The blood-brain barrier disruption maybe by an external trigger like infection or by internal triggers like cytokines. Auto-antibodies cause neuronal injury and altered synaptic transmission. Vascular injury mediated by antibodies or by accelerated atherosclerosis can also cause neuronal damage [4].

The diagnosis of NPSLE is like solving a puzzle. The clinical manifestations of NPSLE might overlap with the neuropsychiatric manifestations of Sjögren's syndrome and antiphospholipid antibody syndrome and other autoimmune diseases. Autoantibodies are the key to the diagnosis of SLE. Several autoantibodies are found to correlate with neuropsychiatric symptoms even though they are not specific markers for NPSLE [4].

- Antiphospholipid antibodies-stroke and vascular dementia, seizures, chorea, headache, and transverse myelitis.
- Anti-ribosomal-P antibodies-depression or psychosis.
- Anti-neuronal antibodies-cognitive dysfunction and depression.
- Anti-NMDA receptor antibodies-migraine, acute confusional state, depression, and peripheral neuropathy.

Initial diagnostic evaluation should be similar to that in non-SLE patients, aiming to exclude secondary causes of neuropsychiatric events. MRI is the preferred imaging test for visualizing brain and spinal pathologies [5]. Neuroimaging is helpful mostly to rule out other possible causes of the presentation but cannot be the sole diagnostic tool for NPSLE, which greatly relies on clinical evaluation.

Management involves symptomatic treatment like correction of hypertension and metabolic parameters; anti-epileptics for seizures; and anxiolytics, antidepressants, mood-stabilizers, or antipsychotics for psychiatric symptoms. Immunosuppressive therapy is indicated for inflammatory NPSLE or when generalized SLE activity is present. Anticoagulation/antiplatelet therapy is recommended for antiphospholipid antibody syndrome-associated manifestations [5]. Glucocorticoids may be considered in severe cases of acute confusional state or SLE-associated psychosis, especially in the presence of generalized SLE disease activity [5].

The clinical presentation of our patient was mistaken as that of postpartum psychosis since she was 3 weeks postpartum and her symptoms overlapped with that of psychosis. Later on, she developed a skin rash, scalp lesions, hair loss, joint pains, and psychotic features. She had low hemoglobin, high ESR, low complement levels, and positive ANA and Anti ds DNA. Thus she fulfilled more than 5 criteria [as per SLICC criteria] and had more than 10 points for the diagnosis of systemic lupus erythematosus by the American College of Rheumatology. Her acute confusional state, anxiety, and psychosis [paranoid delusions, auditory hallucinations] along with persistent insomnia were the central nervous system manifestations of NPSLE.

This case clearly illustrates the relevance of the diagnosis of SLE in our patient. It highlights the neuropsychiatric manifestations of SLE can often overlap with other primary psychiatric disorders. Early diagnosis and initiation of immunosuppressive therapy will not only lead to better control of CNS symptoms but also defer the progression of the underlying autoimmune disease.

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Keywords: Neuropsychiatric systemic lupus erythematosus; NPSLE; Lupus; Postpartum psychosis; Lupus cerebritis

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