Neuropsychiatric SLE (NPSLE) with Cerebral Vasculitis-like Tumor

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Background

Systemic lupus erythematosus (SLE) is a chronic, multisystem autoimmune disease. **Neuropsychiatric SLE (NPSLE)** refers to a spectrum of **neurological and psychiatric manifestations** directly related to SLE.

Female predominance: Prevalence is higher in women, with a female-to-male ratio of approximately 9:1.

Cerebral vasculitis is rare in SLE, reported in **<10% of postmortem studies.** While **small-vessel vasculitis of the brain** is relatively well recognized**, large-vessel vasculitis** producing a **tumor-like appearance** is extremely uncommon.

Rapid diagnosis and prompt management are essential to improve outcomes in such rare presentations.

Case Report

Mrs. I, a **30-year-old woman** with a **history of SLE for 4 years,** presented with a **sudden onset throbbing headache on the right side (NPRS 7–8)** accompanied by **weakness of the right hand.** She reported a **history of frequent right-hand spasms and involuntary movements for the past 3 months,** along with **frequent auditory hallucinations** and **anxiety since being diagnosed with SLE. Vital signs were normal.** On physical examination, a **malar rash (+) was noted. Neurological examination revealed monoparesis of the right hand with spastic tone, disuse atrophy, and motor strength 3/5. Pathological reflexes were positive for Hoffman and Tromner in the right hand,** while other neurological findings were within normal limits.

Conclusion

NPSLE should always be considered in the differential diagnosis of SLE patients with CNS lesions resembling a mass (tumor). Prompt recognition of this atypical presentation and prompt initiation of immunosuppressive therapy are crucial to limit permanent nerve tissue damage and improve patient prognosis.

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Diagnostic Findings

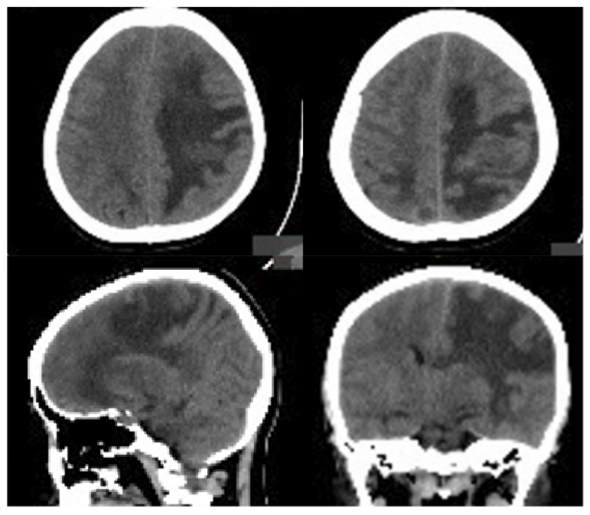

Figure 1. A slight iso-dense lesion measuring approximately 5x4 cm in the left basal ganglia accompanied by surrounding edema that causes displacement of the left lateral ventricle with a midline shift to the right of 0.5 cm, suggestive of a cerebral mass.

Parameter	Result	Normal Range
Hb	8,9	12 – 16 g/dl
Ureum	55	10 – 50 mg/dl
Kreatinin	2,0	0,6 - 1,1 mg/dl

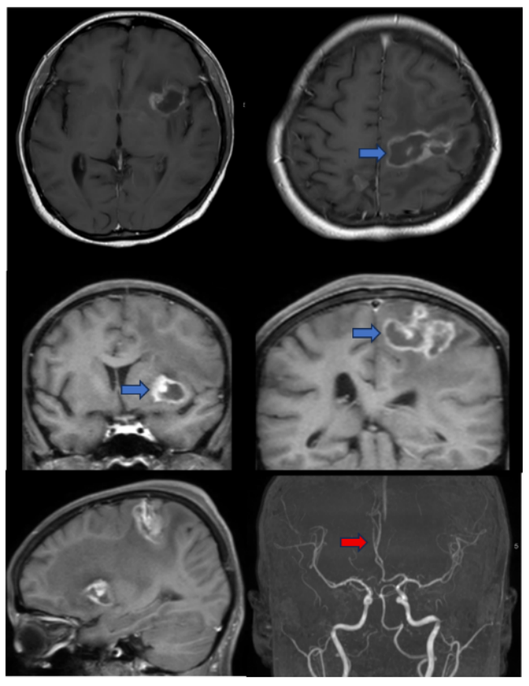

Figure 2. Blue Arrow: Rim enhancing lesion, irregular shape in the external capsule – left lentiform nucleus and left parietal cortex-subcortex with hemorrhagic components, extensive perifocal edema in the fronto-parietal subcortex. Suggestion of demyelinating process (SLE CNS Manifest). Red Arrow: Right A1 ACA irregularity can be DD: 1. Stenosis, 2. Vasculitis

Discussion

This patient meets the diagnostic criteria for **neuropsychiatric SLE (NPSLE)**, namely patients who have been diagnosed with SLE who experience neurologicaland psychiatric symptoms. CNS symptoms in SLE patients can be caused by **cerebral vasculitis** and cause **thromboembolic processes** in the cerebral blood vessels and cause CNS manifestations resembling stroke. When vasculitis affects **large blood vessels**, it may appear tumor-like on imaging, with a **CT scan showing vasogenic edema and mass effect**. The diagnostic challenge in mass-lesion cases is that **lesions that mimic tumors** (cerebral vasculitis-like tumors) **must be distinguished from primary neoplasms/metastases and brain infections** (abscesses). Diagnosis of **NPSLE is made by exclusion** after ruling out other causes and is supported by features of vasculitis. **Management must immediately involve intensive immunosuppression** to stop the autoimmune-inflammatory process in the CNS. Initial therapy with **high-dose methylprednisolone, followed by maintenance therapy with hydroxychloroquine and mycophenolate mofetil,** and **symptomatic therapy with antiepileptic drugs and antiplatelets** to control seizures and reduce thrombotic risk. After treatment, the patient's clinical condition improved.