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Severe and Rare Cerebrovascular Involvement in Systemic Lupus Erythematosus: Case Report and Literature Review

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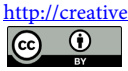

Abstract

Cerebral vasculitis is an uncommon but severe manifestation of systemic lupus erythematosus (SLE), often associated with complex management and unfavorable outcomes due to its neurological impact. We describe the case of a 32-year-old male with SLE who developed recurrent seizures and progressive cognitive decline. Clinical evaluation, laboratory investigations, and neuroimaging supported the diagnosis of lupus-related cerebral vasculitis. Despite aggressive immunosuppressive therapy, the patient experienced worsening neurological impairment, culminating in multiorgan failure and death. This report underscores the critical need for early recognition and prompt, intensive management of cerebral vasculitis in patients with SLE to improve prognosis.

Subject Areas

Neurology, Radiology & Medical Imaging

Keywords

Systemic Lupus Erythematosus (SLE), Cerebral Vasculitis, Neurological Complications, Multiorgan Failure, Immunological Dysfunction

1. Introduction

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by immune system dysfunction, leading to inflammation and damage across various organs and systems in the body. While SLE primarily affects women of childbearing age, clinical manifestations can vary widely, from mild symptoms to life-threatening complications [1]. About half of individuals with SLE experience

neurological symptoms at some point during their disease, with varying severity and clinical forms [2]. Cerebral vasculitis, though uncommon, is one of the most severe neurological complications of SLE, associated with high morbidity and mortality rates [3]. SLE affects multiple organ systems, including muscles as well as the peripheral and central nervous systems. While central nervous system (CNS) involvement is frequent and leads to a variety of neuropsychiatric symptoms in SLE patients, cerebral vasculitis remains a rare manifestation, as confirmed by post-mortem examinations showing a low incidence of this condition. In the context of SLE, vasculitis is characterized by inflammation of cerebral blood vessels, disrupting blood flow and resulting in ischemia and tissue damage. Clinically, this condition can cause seizures, focal neurological deficits, cognitive impairment, and altered consciousness [4]. Diagnosing cerebral vasculitis in SLE patients often requires a thorough clinical evaluation, including laboratory tests and neuroimaging studies. Laboratory results may reveal positive antinuclear antibodies (ANA), elevated inflammatory markers, and evidence of multi-organ dysfunction [5]. Imaging studies, such as MRI, play a key role in detecting abnormalities associated with cerebral vasculitis, including white matter lesions, infarcts, and hemorrhages [6]. Treatment generally relies on powerful immunosuppressants aimed at reducing inflammation and controlling the underlying autoimmune process. Corticosteroids, cyclophosphamide, and other immunomodulatory agents are commonly used to prevent the worsening of neurological symptoms [7]. Despite advancements in the management of SLE and its complications, cerebral vasculitis remains challenging to treat and is often associated with a poor prognosis. Early identification, prompt intervention, and continuous monitoring are crucial to improving outcomes for affected patients. In this context, we present the case of a 32-year-old man with SLE who developed cerebral vasculitis, highlighting clinical signs, diagnostic challenges, and therapeutic approaches for this rare but serious complication.

2. Case Presentation

A 32-year-old patient presented to the emergency room with seizure episodes characterized by abnormal movements of the upper and lower limbs, accompanied by jaw clenching and severe headaches. A year prior, he had exhibited symptoms such as arthralgia, fatigue, skin rashes, photosensitivity, and mouth ulcers, as well as two seizure episodes, with no significant family history of autoimmune or neurological disorders.

Upon physical examination in the emergency room, his blood pressure was 150/85 mm Hg, pulse was 100 bpm, and oxygen saturation was 96%. Neurological evaluation revealed generalized tonic-clonic seizures, limited spontaneous movement, and unreactive pupils. Deep tendon reflexes were diminished or absent. Systemic examinations of the cardiovascular, respiratory, and gastrointestinal systems showed no abnormalities. Additionally, a malar erythematous rash sparing the nasolabial folds, consistent with SLE, was observed.

3. Diagnostic Assessment and Treatment

The patient underwent a comprehensive series of laboratory and imaging investigations, which revealed significant findings. Immunological tests demonstrated the presence of anti-Smith antibodies (Anti-Sm), highly specific for systemic lupus erythematosus (SLE), and antinuclear antibodies (ANA), the most sensitive marker for this diagnosis. Complement levels were reduced (C4 at 7 mg/dL and C3 at 57 mg/dL). Iron deficiency anemia was noted with hemoglobin at 9 g/dL; oral iron protein succinylate was prescribed for one month, but no significant improvement was observed. A blood smear revealed numerous spherocytes, suggesting an autoimmune hemolytic process, which prompted initiation of corticosteroid therapy. Serum creatinine was elevated at 32 mg/L, indicating impaired renal function.

Cerebrospinal fluid (CSF) analysis showed 24 cells/mm^3 (lymphocytic predominance), protein at 1.2 g/L, and normal glucose levels, making infectious meningoencephalitis unlikely. Infectious screening (PCR for HSV, VZV, CMV, HIV, syphilis, and hepatitis serologies) and thrombophilia work-up (protein C, protein S, antithrombin III, antiphospholipid antibodies) were all negative, further supporting an autoimmune etiology.

Radiological studies included chest X-ray, abdominal and pelvic ultrasound, and brain MRI. The chest X-ray was unremarkable, while ultrasound demonstrated cortico-medullary dedifferentiation, suggesting chronic kidney disease. Brain MRI, performed on a 1.5 Tesla scanner, revealed T2/FLAIR hyperintensities in the cerebral cortex, juxtacortical and subcortical white matter of both hemispheres, and in the right cerebellar hemisphere. Multiple small cortical and subcortical ischemic foci were observed on DWI/FLAIR sequences. Susceptibility-weighted imaging (SWI) disclosed diffuse petechial hemorrhages in bilateral cerebral lobes, predominantly involving the cortex and subcortical white matter (**Figure 1**).

The diagnosis of lupus-related cerebral vasculitis was established according to the 2019 EULAR criteria for neuropsychiatric lupus: recurrent seizures, progressive cognitive decline, specific immunological profile (ANA, Anti-Sm positivity, hypocomplementemia), diffuse MRI abnormalities (cortical and subcortical lesions, microhemorrhages), and exclusion of alternative diagnoses. However, no histopathological confirmation by brain biopsy was performed, which remains a diagnostic limitation.

Treatment included high-dose corticosteroids (methylprednisolone 1 g/day IV for 3 days followed by oral prednisone), monthly intravenous cyclophosphamide (750 mg/m^2) according to the 2019 EULAR recommendations, and antiepileptic drugs for seizure control. Despite this aggressive immunosuppressive therapy, the patient's neurological status continued to deteriorate. He developed respiratory failure with pulmonary edema, followed by coma requiring mechanical ventilation. Despite maximal supportive measures, he subsequently progressed to multiorgan failure, leading to death.

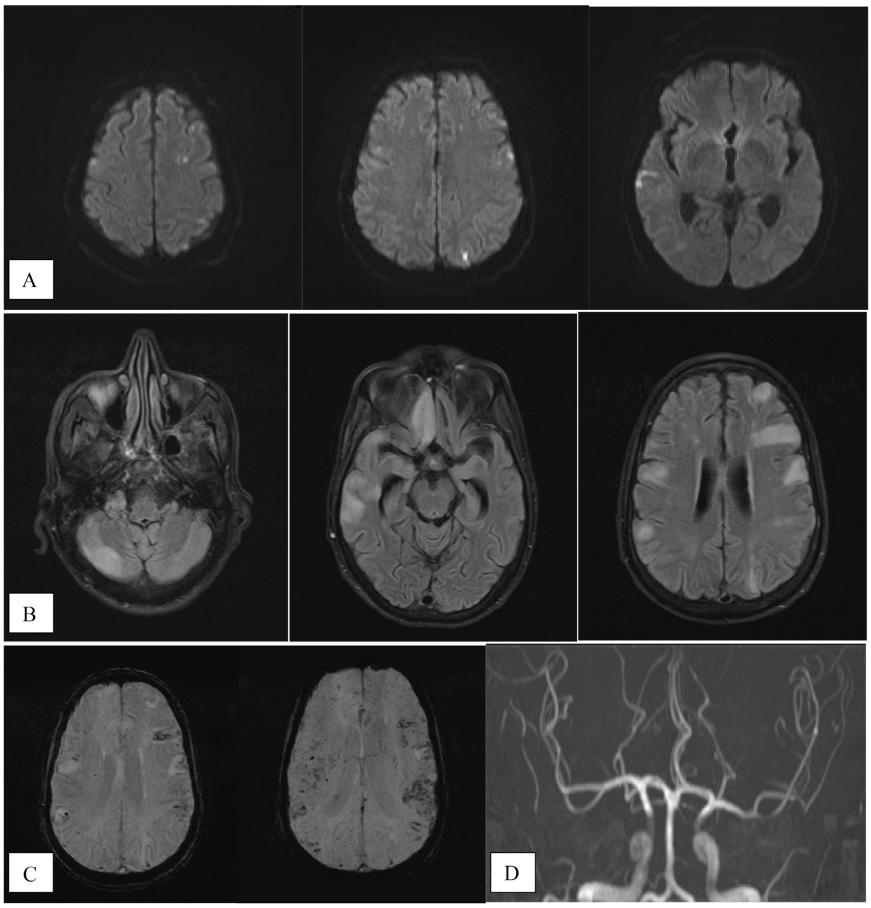

Figure 1. A: Axial slices from the Diffusion sequence. B: Axial slices from the FLAIR sequence. C: SWI sequence. D: 3D TOF sequence. These sequences show multiple cortico-subcortical areas affecting both the posterior fossa and the supratentorial level, appearing on FLAIR with punctate diffusion restriction in some areas, without ADC drop. Presence of cortico-subcortical microbleeds, visible in the SWI sequence. The 3D TOF Angio-MRI sequence does not show any abnormalities in the Circle of Willis.

4. Discussion

Cerebral vasculitis is a rare but serious complication of systemic lupus erythematosus (SLE), characterized by inflammation of the cerebral blood vessels and associated with a high risk of severe neurological complications and poor prognosis [8]. In our patient, multiple neurological manifestations were observed, including seizures, cognitive impairment, and episodes of unresponsiveness, strongly suggesting central nervous system (CNS) involvement related to SLE. The presence of a malar rash, positivity for anti-Smith (Anti-Sm) and antinuclear antibodies (ANA), together with other laboratory findings consistent with SLE, further supported this diagnosis. Imaging findings revealed characteristic features of cerebral vasculitis, such as T2/FLAIR hyperintensities and petechial hemorrhages in the cerebral parenchyma [9].

This case meets several diagnostic criteria for lupus-related cerebral vasculitis. However, the absence of histopathological confirmation by brain biopsy remains

a diagnostic limitation. Furthermore, vascular wall MRI (VW-MRI) or conventional angiography were not performed, which reduces diagnostic certainty, as these modalities could have distinguished vasculitis from mimics such as reversible cerebral vasoconstriction syndrome, intracranial atherosclerosis, Moyamoya disease, or dissection.

The treatment of cerebral vasculitis in the context of SLE typically requires aggressive immunosuppressive therapy aimed at controlling the autoimmune process and reducing CNS inflammation. In this patient, high-dose corticosteroids combined with cyclophosphamide were administered to limit disease activity and prevent further neurological deterioration [10]. Despite these interventions, the patient's neurological status progressively worsened, underscoring the therapeutic challenges in managing this severe manifestation of SLE. The development of respiratory failure and multiorgan dysfunction further complicated the clinical course. Ultimately, despite intensive supportive care, including mechanical ventilation and hemodynamic management, the patient succumbed to multiorgan failure, attributable to CNS vasculitis and its complications.

5. Conclusion

In conclusion, this case highlights the major challenges posed by cerebral vasculitis in the context of systemic lupus erythematosus (SLE). Despite aggressive management, including immunosuppressive therapy and supportive care, the patient's neurological condition worsened, ultimately leading to multiorgan failure and a tragic outcome. This situation underscores the limitations of current therapeutic approaches and the urgent need to pursue more effective strategies for treating cerebral vasculitis in SLE. It also emphasizes the importance of early detection and rapid treatment to improve clinical outcomes. Finally, the complexity and severity of this condition necessitate a multidisciplinary approach, involving rheumatologists, neurologists, and intensive care specialists, to optimize care and reduce the potentially devastating impact of this disease.

Author Contributions

Dr. AJERTIL is the primary author, Professor Kabbaj and Professor Abdeljalil EL QUESSAR contributed to the development of this work by providing her expertise in writing.

Conflicts of Interest

The authors declare no conflicts of interest.

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