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Case Report

Cerebral Vasculitis in Systemic Lupus Erythematosus (SLE) : Case Report and Literature Review

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Abstract

Cerebral vasculitis, though rare, represents a serious complication of systemic lupus erythematosus (SLE) and poses numerous challenges in terms of management due to its potential for severe neurological consequences and often poor prognosis. We report the case of a 32-year-old man with SLE who presented with three seizure episodes and cognitive deterioration. Neurological examinations, laboratory analyses, and imaging findings led to a diagnosis of cerebral vasculitis secondary to SLE. Despite intensive immunosuppressive treatment, the patient's neurological condition deteriorated, resulting in multiorgan failure. Ultimately, the patient died due to multiorgan failure linked to severe central nervous system vasculitis and associated complications. This case highlights the importance of early detection and aggressive treatment of cerebral vasculitis in the context of SLE.

Keywords: Systemic Lupus Erythematosus (SLE); Cerebral Vasculitis; Neurological Complications; Multiorgan Failure; Immunological Dysfunction

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.

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.

Diagnostic assessment and treatment

The patient underwent a comprehensive series of laboratory and imaging tests, revealing significant findings. Notably, a positive anti-Smith antibody (Anti-Sm), highly specific for SLE, and an antinuclear antibody (ANA), the most sensitive for this diagnosis, were identified. The tests also showed reduced complement levels (C4 at 7 mg/dL and C3 at 57 mg/dL). Iron deficiency anemia was detected with a hemoglobin level of 9g/L, and oral iron protein succinylate was prescribed for one month; however, no notable improvement was observed. A blood smear revealed a high presence of spherocytes, suggesting an autoimmune hemolytic process, necessitating the initiation of corticosteroid therapy.

The patient also had an elevated serum creatinine level of 32 mg/L, indicating impaired renal function. Finally, cerebrospinal fluid (CSF) analysis showed elevated protein levels and pleocytosis, indicating central nervous system (CNS) inflammation.

Radiological examinations included chest X-ray, abdominal and pelvic ultrasound, and brain MRI. The chest X-ray was normal, the ultrasound revealed cortico-medullary dedifferentiation, suggesting chronic kidney disease. The brain MRI, performed on a 1.5 Tesla MRI machine, showed T2/FLAIR hyperintensities in the cerebral cortex and juxtacortical and subcortical white matter of both cerebral hemispheres as well as in the right cerebellar hemisphere. Additionally, multiple small cortical and subcortical ischemic foci were visible on DWI/FLAIR sequences. The susceptibility-weighted imaging (SWI) sequence revealed diffuse petechial hemorrhages in the bilateral cerebral lobes, primarily involving the cerebral cortex and subcortical white matter, as shown in figure 1.

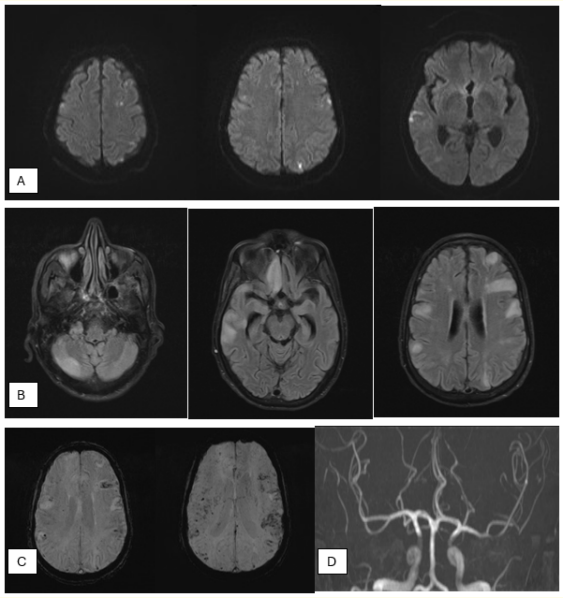

Figure 1: A: Axial slices from the Diffusion sequence. B: Axial slices from the FLAIR sequence. C: SWI sequence. D: 3D TOF sequence. These sequences show multiple cortico-subcortical areas affecting both the posterior fossa and the supratentorial level, appearing on FLAIR with punctate diffusion restriction 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.

The patient received treatment including corticosteroids (Methylprednisolone 1g IV) to control inflammation, antiepileptics to control seizures, and cyclophosphamide to address the ongoing autoimmune process. Despite aggressive medical intervention for SLE, the patient's neurological status continued to deteriorate. He developed respiratory failure with pulmonary edema, followed by coma, requiring respiratory support. Despite all supportive care provided, the patient subsequently developed multiorgan failure, leading to his death.

Discussion

Cerebral vasculitis is a rare but serious complication of systemic lupus erythematosus (SLE), involving inflammation of the cerebral blood vessels and presenting a significant risk of severe neurological complications, along with a poor prognosis [8]. In our patient, he showed various neurological symptoms such as seizures, altered cognitive functions, and unresponsiveness, indicating central nervous system (CNS) involvement related to SLE. The presence of a malar rash, positive anti-Smith (Anti-Sm) and antinuclear (ANA) antibodies, along with other laboratory findings consistent with SLE, supported this diagnosis. Moreover, imaging tests revealed signs of cerebral vasculitis, including T2/FLAIR hyperintensities and petechial hemorrhages in the cerebral parenchyma [9]. To exclude secondary infections, contrast-enhanced MRI is necessary. In addition to standard MRI, vascular wall MRI (VW-MRI) can help differentiate vascular narrowing related to intracranial atherosclerotic plaque, reversible cerebral vasoconstriction syndrome, dissection, Moyamoya disease, as well as vasculitis, which presents as enhancement of the affected arterial wall.

The treatment of cerebral vasculitis in the context of SLE generally relies on an aggressive immunosuppressive protocol aimed at controlling the underlying autoimmune process and reducing inflammation. In this case, the patient received a combination of corticosteroids and cyclophosphamide to limit disease activity and prevent neurological worsening [10]. Despite these interventions, the patient's neurological condition continued to deteriorate, illustrating the challenges in managing this SLE complication. The onset of respiratory failure and multiorgan dysfunction further complicated the situation. Despite intensive supportive care, including respiratory assistance and hemodynamic management, the patient ultimately succumbed to multiorgan failure, attributable to CNS vasculitis and its complications [11].

Conclusions

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.

Conflicts of Interest

The authors declare no conflicts of interest.

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.

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