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Reversible Cerebral Vasoconstriction Syndrome (RCVS) in Systemic Lupus Erythematosus: A Case Report

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Abstract

Reversible cerebral vasoconstriction syndrome (RCVS) is a neurovascular condition characterized by episodes of sudden and severe headache secondary to transient angiographic changes in the cerebral arteries, which may be accompanied by neurological deficits. Differentiating RCVS from cerebral vasculitis associated with systemic lupus erythematosus (SLE) represents a clinical and radiological challenge due to overlapping manifestations, but it has major therapeutic and prognostic implications. We report the case of a 32-year-old female patient with SLE who developed a thunderclap headache and mild motor deficit. Cranial computed tomography angiography revealed a pattern of arterial irregularity compatible with RCVS, later confirmed by magnetic resonance angiography, which demonstrated complete reversal of the vascular abnormalities. The patient showed favorable clinical evolution with conservative management, avoiding the need for intensive immunosuppressive therapy. This case highlights the importance of early recognition of RCVS in patients with SLE, preventing potentially harmful treatments and enabling a targeted approach for improved clinical outcomes.

Keywords

Cerebral Vasculitis, Reversible Cerebral Vasoconstriction Syndrome, Systemic Lupus Erythematosus

1. Introduction

Considered a neurovascular disorder, reversible cerebral vasoconstriction syndrome (RCVS) is characterized by episodes of sudden-onset, severe headache, also known as thunderclap headache, associated with neurological deficits in 8% - 43% of cases, resulting from multifocal and segmental vasoconstriction of the cerebral arteries [1]. This entity typically shows clinical reversibility within a 12-week period; however, it may be complicated by cerebral ischemic and hemorrhagic events in up to 10% of cases, with a reported mortality rate of 1% - 5% [2] [3].

In patients with systemic lupus erythematosus (SLE), distinguishing RCVS from other clinical entities—particularly lupus-associated cerebral vasculitis—is of paramount importance due to differences in clinical prognosis and therapeutic strategies. While cerebral vasculitis benefits from immunosuppressive therapy, RCVS management relies mainly on calcium channel blockers and the removal of precipitating factors [2] [3].

Individuals with SLE, whether with active disease or low activity/remission, may develop episodes of RCVS, which do not necessarily depend on classical immunological activity of lupus [2]. Precipitating factors such as exposure to vasoactive drugs (including certain immunosuppressants such as tacrolimus and cyclophosphamide), blood transfusions, hypertensive states, endothelial dysfunction, and other lupus-directed therapeutic interventions have been described as potential triggers in the literature [4].

2. Case Description and Discussion

I.S.O., a 32-year-old female patient, smoker, with a diagnosis of SLE established approximately two years earlier, was referred to a tertiary care hospital, presenting signs of disease activity. She reported moderate to severe headache, alopecia, hyperchromic skin lesions, symmetric arthritis of small joints, and laboratory abnormalities, including anemia, thrombocytopenia, and proteinuria. Neurological examination at admission was unremarkable, and a non-contrast cranial computed tomography scan showed no abnormalities.

After five days of hospitalization, the patient developed a new episode of headache with peak intensity reached within minutes, described as unprecedented by the patient, associated with mild, symmetric reduction in lower limb strength, without other accompanying symptoms. Further laboratory evaluation, cerebrospinal fluid analysis, and cranial computed tomography angiography (CTA) were performed for diagnostic clarification. CTA demonstrated globally reduced cerebral blood flow with focal arterial irregularities, particularly involving the middle cerebral arteries, resembling a "string-of-beads" pattern. These findings raised the suspicion of RCVS, with cerebral vasculitis associated with lupus considered as a differential diagnosis, as both conditions may present with similar imaging features (**Figure 1** and **Figure 2**).

During the subsequent days of hospitalization, conservative therapeutic measures were adopted, consisting of simple analgesics and close clinical observation. The

patient evolved with improvement of headache, without alarm signs or significant neurological deterioration. Laboratory test results did not reveal relevant abnormalities.

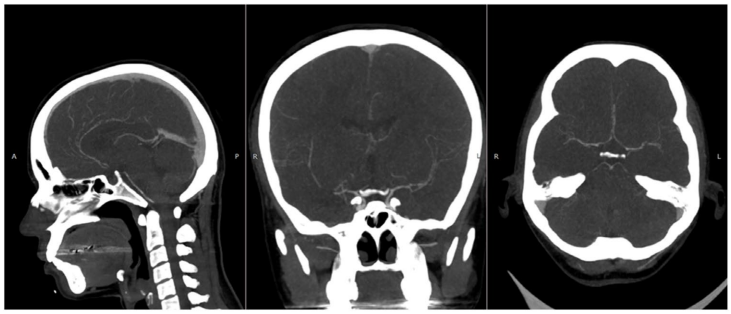

Figure 1. Cranial CT angiography in sagittal (a), coronal (b), and axial (c) views, arterial phase with intravenous contrast, showing global reduction of cerebral circulation.

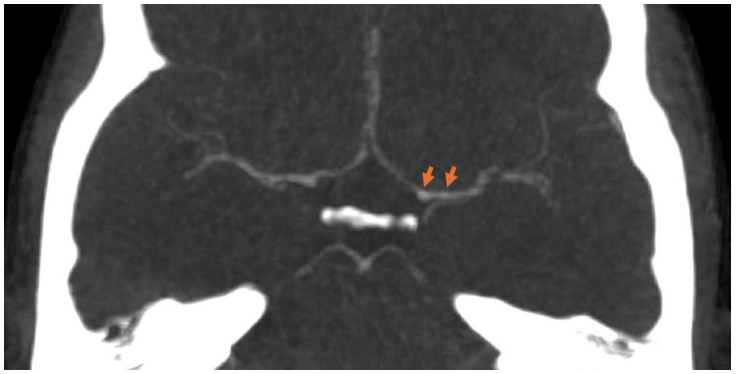

Figure 2. Cranial CT angiography, arterial phase, axial section, showing areas of flow narrowing alternating with preserved-caliber segments in the anterior, middle, and posterior cerebral arteries, resembling a "string-of-beads" pattern.

On the twenty-first day of hospitalization, the patient reported complete resolution of headache and underwent follow-up neuroimaging with magnetic resonance angiography (MRA). The examination demonstrated adequate cerebral blood flow with complete regression of the previously observed focal intracranial arterial irregularities, confirming the diagnosis of RCVS (**Figure 3** and **Figure 4**).

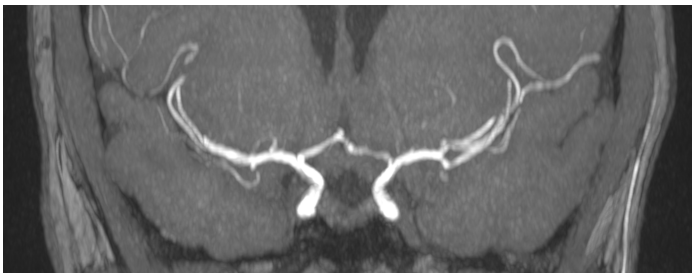

Figure 3. Brain MR angiography (3D-TOF sequence) showing preserved flow signal and normal calibers in the intracranial segments of the internal carotid and middle cerebral arteries.

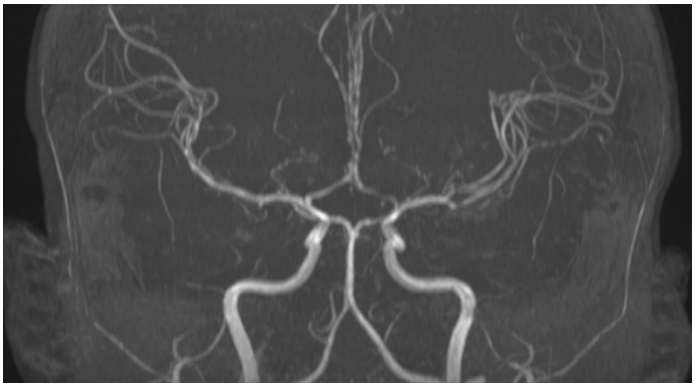

Figure 4. Brain MR angiography with 3D MIP reconstruction demonstrates preserved flow signal and arterial calibers in intracranial segments.

3. Conclusions

Headache is a frequent clinical complaint in patients with systemic lupus erythematosus, particularly in those with central nervous system (CNS) involvement, with or without associated cerebral vasculitis [5]. RCVS represents an important differential diagnosis in this population, as it shares several overlapping clinical and imaging features that may hinder accurate diagnosis and appropriate management [6].

Given the divergent therapeutic approaches between cerebral vasculitis and RCVS—and the potential harm and clinical deterioration associated with the use of immunosuppressive medications in patients with RCVS—adequate knowledge of neuroradiological characteristics is essential. The use of imaging modalities such as CT angiography and MR angiography in SLE patients with compatible clinical presentations is crucial to guide appropriate therapeutic decisions and achieve favorable clinical outcomes.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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