Radiology Case Reports 20 (2025) 3221–3226

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Case Report

Status epilepticus complicating a hemorrhagic and ischemic stroke due to primary cerebral angiitis: A case report ☆

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ARTICLE INFO

Article history:
Received 27 November 2024
Revised 9 March 2025
Accepted 11 March 2025

Keywords:
Angiitis
Stroke
Status epilepticus
MRI
Angiography

ABSTRACT

Primary angiitis of the central nervous system (CNS), also known as "isolated" or "idiopathic" angiitis, is a rare condition characterized by inflammation of cerebral vessels in the absence of an identifiable cause. This contrasts with secondary angiitis, which is associated with systemic diseases, infections, or toxic exposures. The clinical presentation of primary angiitis is highly variable and, in severe cases, may include ischemic or hemorrhagic strokes or refractory status epilepticus (SE).

We present a unique case of a 45-year-old female patient with a history of primary CNS angiitis, admitted to the intensive care unit for refractory SE secondary to concurrent ischemic and hemorrhagic strokes. This rare combination of SE, stroke, and CNS angiitis is exceptionally uncommon in the literature and underscores the diagnostic and therapeutic challenges of this condition. This case highlights potential pathophysiological mechanisms, diagnostic difficulties, and management strategies in a highly complex clinical scenario.

Introduction

Primary angiitis of the central nervous system (CNS), also referred to as "isolated" or "idiopathic" angiitis [1], is a rare disease [2] that belongs to the spectrum of cerebral vasculitides [3], a group of conditions characterized by inflammation of the cerebral blood vessels.

Unlike secondary angiitis, which is associated with systemic diseases, toxic exposures, infections, or neoplastic conditions [4], primary angiitis is defined by the absence of any identifiable cause.

The clinical presentation of primary angiitis is highly variable [5], with signs and symptoms that often mimic other neurological disorders. In severe cases, primary CNS angiitis

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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https://doi.org/10.1016/j.radcr.2025.03.038

may manifest as ischemic strokes (IS) and, more rarely, hemorrhagic strokes (HS) [5].

We present the case of a 45-year-old female patient with a known history of cerebral angiitis, admitted to the intensive care unit for management of status epilepticus (SE) secondary to concurrent IS and HS in the context of cerebral angiitis. This simultaneous triple association of SE, ischemic and hemorrhagic strokes, and cerebral angiitis is exceptionally rare.

This article aims to present a clinical case illustrating this pathological combination and to discuss the potential pathophysiological mechanisms, diagnostic challenges, and therapeutic strategies involved in this complex scenario.

Patient observation

A 45-year-old female patient presented to the emergency department with status epilepticus, manifesting as a continuous convulsive seizure lasting 15 minutes. According to her family, she had been on long-term treatment with azathioprine for primary cerebral vasculitis, diagnosed in 2014 via leptomeningeal biopsy (unfortunately, no prior imaging studies were available for review).

Upon initial evaluation, the patient was actively seizing, exhibiting ocular revulsion, frothing at the mouth, and urinary incontinence. Her vital signs included a blood pressure of 140/90 mmHg, a heart rate of 130 beats per minute (without signs of peripheral hypoperfusion), and tachypnea at 33 breaths per minute. Her oxygen saturation was 89% on room air, which improved to 98% with supplemental oxygen at 5 liters per minute. Pulmonary auscultation revealed bilateral wheezing. Capillary blood glucose was measured at 1.1 g/L, and her body temperature was 37.5°C.

Two initial doses of midazolam (0.015 mg/kg) were administered 5 minutes apart, but the seizures persisted. A subsequent infusion of phenobarbital (30 mg/kg over 30 minutes) also failed to control the seizures. Due to the refractory nature of the status epilepticus, orotracheal intubation and general anesthesia were initiated for seizure management.

Intubation was performed using a crash-induction protocol with 200 mg of fentanyl, 150 mg of titrated propofol, and 80 mg of rocuronium. After securing the airway, sedation was maintained, and a brain CT scan was performed (Fig. 1**).** The imaging revealed ischemic lesions with hemorrhagic transformation, as well as hemorrhagic lesions, consistent with both acute and chronic ischemic and hemorrhagic strokes. No abnormalities were observed in the branches of the circle of Willis. These findings were likely related to her history of primary cerebral angiitis, reflecting involvement of the cerebral microvasculature**.**

An additional imaging assessment with MR angiography was subsequently requested (Fig. 2).

The patient was subsequently admitted to the intensive care unit (ICU) for further management. Additional intravenous access was established, and a urinary catheter and nasogastric tube were inserted. Sedation was maintained using fentanyl and midazolam administered via a syringe pump. The patient was positioned semi-recumbent at 30 degrees, with proper alignment of the head and torso. To manage the refractory status epilepticus, 2 anticonvulsants, Urbanyl (clobazam) and Keppra (levetiracetam), were administered. Cerebral hemodynamic monitoring was conducted using repeated transcranial Doppler ultrasounds.

Two days later, the patient developed bilateral mydriasis. Transcranial Doppler findings suggested a no-flow state, indicative of the absence of cerebral perfusion. A brain CT angiography was performed following a brain death protocol, which revealed no contrast opacification of the intracranial arteries or the internal cerebral veins, confirming the absence of cerebral blood flow and supporting the diagnosis of brain death. An organ donation protocol was considered; however, the patient succumbed to cardiorespiratory arrest the following day.

Discussion

Cerebral angiitis encompasses a group of disorders characterized by inflammation of the cerebral blood vessels [3]. This condition primarily affects small leptomeningeal and cortical arteries (200 to 500 μm in diameter) [6] and, less frequently, medium-sized arteries, capillaries, and veins. The inflammation is often segmental, which may account for the potential of negative biopsy results [6]. Histopathological findings may include vascular necrosis or granulomatous formations [6,7].

A clear distinction is made between primary and secondary angiitis [3]. Secondary angiitis is associated with identifiable causes, such as systemic diseases, toxins, infections, or neoplasms. In contrast, primary central nervous system (CNS) angiitis occurs without an identifiable underlying cause, making its diagnosis particularly challenging.

The etiopathogenesis of primary central nervous system (CNS) angiitis remains poorly understood [8]. It is hypothesized to result from a chronic immune response triggered by an infectious agent, either viral or bacterial, leading to sustained inflammation mediated by the immune system. Viruses such as herpes-zoster, HIV, and cytomegalovirus, as well as bacterial agents like Mycoplasma, have been implicated in this process [8–10].

The clinical presentation of primary CNS angiitis is highly variable [5]. Symptoms may include headaches, focal neurological deficits, seizures, language disturbances, and other neurological manifestations. The onset can be acute, mimicking a stroke, or chronic, resembling a pseudo-tumoral process. The classic presentation often involves diffuse encephalopathy, characterized by persistent headaches, dementia-like symptoms, and seizures. The absence of extra-neurological symptoms, despite a wide range of neurological findings, may suggest this diagnosis.

Primary CNS angiitis can lead to severe complications, most notably ischemic strokes (IS) and, more rarely, hemorrhagic strokes (HS). In the presented case, the clinical manifestation involved status epilepticus as a complication of both IS and HS in the context of primary CNS angiitis.

Complementary investigations play a crucial role in the diagnostic process. CT-angiography is usually the first imaging method prescribed, providing rapid, high-resolution imaging of larger vessels but limited sensitivity for small vessel disease [11]. MR-angiography [12] is instrumental in identifying vascular anomalies, multiple infarcts of varying ages, or inflam-

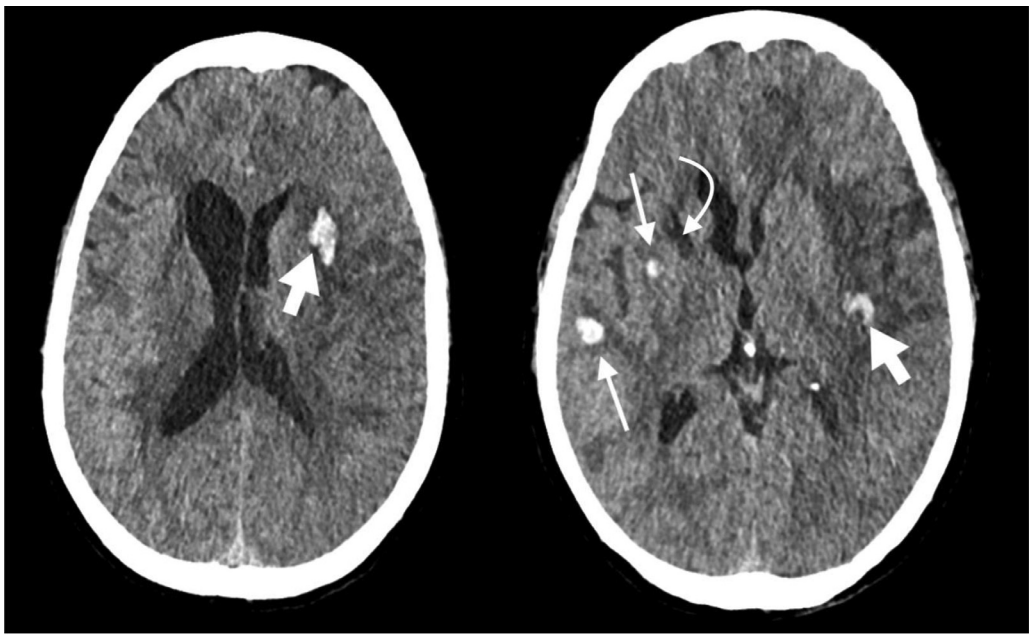

Fig. 1 – CT images showing a hypodense area in the left parieto-temporo-insular and lenticular regions, consistent with an ischemic stroke (IS) with signs of hemorrhagic infarction (thick arrows), along with contralateral intraparenchymal hematomas (thin arrows) and chronic infarct in the right internal capsule (bended arrow).

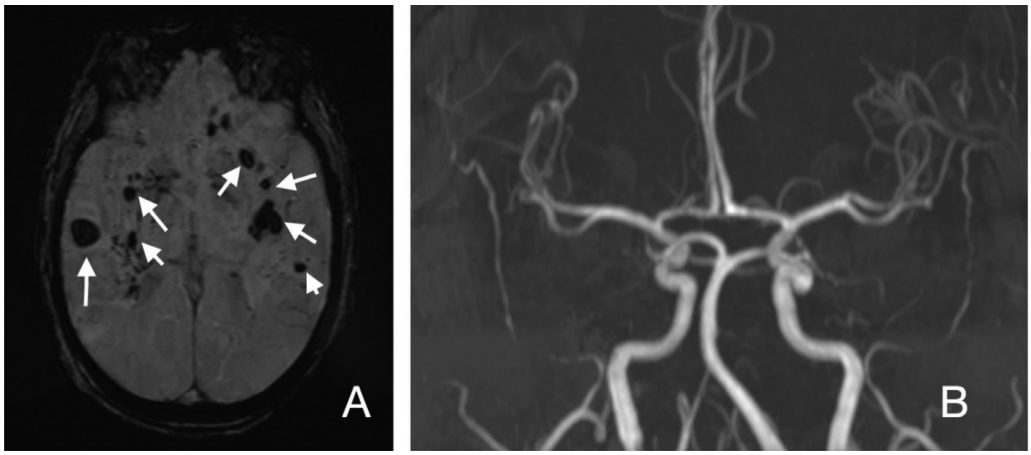

Fig. 2 – SWI sequence which is highly sensitive to molecules with high magnetic susceptibility, such as calcium and blood, showing the presence of several areas of signal void, corresponding to intraparenchymal hemorrhagic foci of varying sizes (white arrows) (A). TOF sequence, which is a noncontrast angiography technique used to assess cerebral circulation, showing no flow abnormalities in the branches of the circle of Willis (B).

matory lesions. It may also reveal leptomeningeal enhancement or signs of vasogenic edema. However, it may also miss small vessel disease or subtle changes, leading to false negatives. Therefore, CTA and MRA are mainly used as initial non-invasive screening tools, and further investigations may prove necessary.

Cerebral angiography [13], is considered the gold standard for diagnosing cerebral angiitis due to its high resolution and ability to detect subtle vascular abnormalities. It can show characteristic findings such as segmental narrowing, beading, and vessel occlusion. However, it is invasive, carries risks (e.g., stroke, contrast reactions), and may miss small vessel involvement, which is common in cerebral angiitis.

Lumbar puncture may be performed to analyze cerebrospinal fluid (CSF), with findings such as pleocytosis, elevated protein levels, or oligoclonal bands suggesting inflammation. Laboratory tests, including inflammatory markers such as C-reactive protein (CRP) and erythrocyte sedimenta-

I. Secondary angiitis

1. Cerebral Angiitis Associated with Systemic Disease

Systemic Vasculitides and Connective Tissue Diseases

2. Cerebral Angiitis Associated with Infectious Pathology

- **Bacterial**: Bacterial meningitis, bacterial endocarditis, Lyme disease, chlamydia, tuberculosis, Mycoplasma infection, syphilis, cat scratch disease (Bartonella), rickettsioses (Rocky Mountain spotted fever, typhus).
- **Viral**: Herpes, cytomegalovirus, shingles, HIV, chickenpox.
- **Parasitic**: Toxoplasmosis, protozoan infections, cysticercosis.
- **Fungal**: Aspergillosis, coccidioidomycosis, candidiasis, histoplasmosis, cryptococcosis, mucormycosis

3. Cerebral Angiitis Associated with Neoplastic Disease

- Carcinomatous meningitis (all causes)
- Hodgkin and non-Hodgkin lymphomas

4. Drug-Induced, Toxic, or Iatrogenic Cerebral Angiitis

- **Sympathomimetic substances:** Crack, amphetamines
- **Medications and treatments:** Carbamazepine, radiation therapy

5. Post-Transplant: Immunosuppression and organ rejection

II. Primary angiitis of the central nervous system

Fig. 3 – Angiitis classification.

1. **Neurological deficit suggestive of CNS angiitis.**
   - Headaches, focal neurological deficits, impaired consciousness, seizures, and dementia syndrome
   - Rapidly progressive onset, with the association of multiple symptoms indicating diffuse cerebral and/or spinal cord involvement
2. **Absence of systemic signs.**
3. **Biological tests, CT scan, brain MRI, and CSF analysis: exclude systemic conditions that could lead to secondary CNS angiitis.**
4. **Demonstration of CNS angiitis, either through angiography or leptomeningeal and cortical biopsy :**
   - Cerebral angiography: shows an alternation of segmental vessel narrowing and dilations of the intracranial vessels, suggestive of angiitis
   - Leptomeningeal and cortical biopsy: performed if no other cause is found; confirms the diagnosis of vasculitis

Fig. 4 – Diagnostic criteria for primary cerebral angiitis according to Calabrese and Mallek (1988) [16].

1. Headaches and multiple focal neurological deficits lasting for at least 6 months (or diffuse and severe neurological involvement, rapidly progressive over several days or weeks).

2. Cerebral angiography showing alternating segmental narrowing and dilation of intracranial vessels, suggestive of angiitis.

3. Exclusion of any systemic disease.

4. Leptomeningeal and cortical biopsy confirming vasculitis and ruling out other causes of vasculopathy (infections, neoplasms, atherosclerosis).

Fig. 5 – Diagnostic criteria for primary cerebral angiitis according to Moore (1989) [17].

tion rate (ESR), as well as immunological studies, can help exclude systemic vasculitis or connective tissue diseases.

The definitive diagnosis of primary CNS angiitis is established through leptomeningeal or brain biopsy [14]. However, this approach has limited sensitivity due to the segmental nature of the disease, which can result in false-negative findings.

The rarity of primary CNS angiitis, combined with the absence of standardized diagnostic criteria, makes its diagnosis particularly challenging. The diagnostic approach typically involves a 3-step process:

1. Exclusion of nonvascular differential diagnoses.
2. Elimination of secondary causes of angiitis.
3. Confirmation of primary CNS angiitis after ruling out all identifiable causes.

Diagnostic criteria, such as those proposed by Calabrese and Malek [15] or Moore [16], provide a framework for navigating this complex diagnostic process. These criteria emphasize a multidisciplinary approach, combining clinical, radiological, and pathological findings to achieve an accurate diagnosis.

Therapeutic management of cerebral angiitis involves both foundational and symptomatic treatments [17]. The foundational treatment primarily consists of corticosteroid therapy, typically initiated with high doses administered either orally or intravenously in severe cases, followed by a gradual taper based on clinical and radiological response. In severe or refractory forms, immunosuppressive agents such as cyclophosphamide, azathioprine, or methotrexate may be added to the regimen (Figs. 3–5).

Symptomatic treatment focuses on managing complications and includes anticonvulsants for seizure control, antithrombotic agents in cases with an increased thrombotic risk, and admission to intensive care or resuscitation units for patients requiring advanced monitoring and support

Conclusion

This case highlights the complexity and diagnostic challenge of primary cerebral angiitis, particularly when complicated by ischemic and hemorrhagic strokes and refractory status epilepticus. Given the rarity of such presentations, the clinical course can be difficult to differentiate from other more common neurological conditions. Imaging plays a crucial role in diagnosis. While conventional cerebral angiography is superior for definitive diagnosis, CTA and MRA are valuable noninvasive alternatives, often used in conjunction with MRI for initial evaluation. It also aids in the differentiation between primary and secondary angiitis. Treatment, which includes corticosteroids, immunosuppressive agents and symptomatic measures, must be tailored to the severity and progression of the disease.

Author contributions

Each author contributed to imaging interpretation or the work done in intensive care unit.

Patient consent

The written consent of the family of the patient was obtained before submission.

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