SVOA Neurology

ISSN: 2753-9180

ScienceVolks Open Access

Case Report

Primary Vasculitis of the Central Nervous System: A Case Report with Various Clinical and Semiological Spectra to Consider

Farid Abedrabbo, MD[1]*; Pablo Granda[1]; Galo Arcentales, MD[1]; Rocio Álvarez, MD[1]; Helder Peñaloza, MD[1]; Carolina Aulestia, MD[1]

[1]Hospital Vozandes Quito, Ecuador.

***Corresponding Author:** Farid Abedrabbo, MD, Hospital Vozandes Quito, Ecuador.

https://doi.org/10.58624/SVOANE.2025.06.016

Received: June 04, 2025

Published: June 25, 2025

Citation: Abedrabbo F, Granda P, Arcentales G, Álvarez R, Peñaloza H, Aulestia C. Primary Vasculitis of the Central Nervous System: A Case Report with Various Clinical and Semiological Spectra to Consider. *SVOA Neurology* 2025, 6:3, 83-85. doi. 10.58624/SVOANE.2025.06.016

Abstract

We report the case of a 47-year-old Ecuadorian woman with no significant medical history who presented with sentinel headache associated with homonymous hemianopia visual deficit. Initial imaging studies showed signs of bilateral occipital parenchymal hemorrhage and superimposed subarachnoid hemorrhage. Initial management focused on hemorrhagic lesions with adequate evolution without a clear identified etiology. On the 15th day of hospitalization, one day before discharge, she presented with multifocal neurological deficits associated with Balint's syndrome and the presence of multifocal ischemic strokes on MRI with suggestive signs of vasculitis in neurovascular study. After excluding infectious, primary vascular, autoimmune, and neoplastic causes, a diagnosis of primary central nervous system vasculitis was established. Following the initiation of immunosuppressive treatment, her clinical evolution was favorable, with no recurrence of events during follow-up.

Keywords: *Central Nervous System Vasculitis, Multifocal Neurological Deficit, Clinical Diagnosis.*

Introduction

Primary central nervous system vasculitis is a rare disease, with an incidence of 2.4 cases per 1,000,000 people per year. It is a medium and small vessel vasculitis that specifically affects the brain and spinal cord in the absence of signs of systemic vasculitis. The variety of symptoms with which it can present is extensive, overlapping several probable etiologies in the context of a critical patient where considering this entity and performing rapid diagnosis and treatment is vital, as the reported mortality range is between 8 to 23%, with at least a quarter of patients suffering severe disability despite treatment; 40% have an unfavorable evolution, and 5% do not survive to be discharged. Although the gold standard for diagnosis is brain biopsy, it is not always available, especially in countries like Ecuador. (1-4)

Case Presentation

A 47-year-old woman, residing in a rural area, with no significant medical history, was admitted to our institution due to progressive onset of severe headache and nonspecific visual disturbances. In the initial interrogation, she did not report the use of drugs or medications. In the systems review, she was asymptomatic. On physical examination, the only notable finding was a right homonymous hemianopsia.

As the first diagnostic study, a plain brain CT scan (Image A-B) was performed, which showed two intraparenchymal occipital hematomas, with the right being larger and extending to the temporal and posterior parietal lobes, signs of hemispheric perilesional edema, and slight midline deviation.

Additionally, there was bilateral convexity subarachnoid hemorrhage (more pronounced in the right hemisphere) and a small right frontotemporal laminar subdural hematoma. Due to the multifocal bleeding findings and three different types of hemorrhages, a diagnostic angiography study was performed, which reported signs of vasospasm in the left middle cerebral artery. She was admitted to the ICU with a diagnosis of multifocal cerebral hemorrhage of unknown etiology, and treatment with nimodipine and supportive care was initiated, with adequate clinical evolution but with recurrent and difficult-to-control headaches.

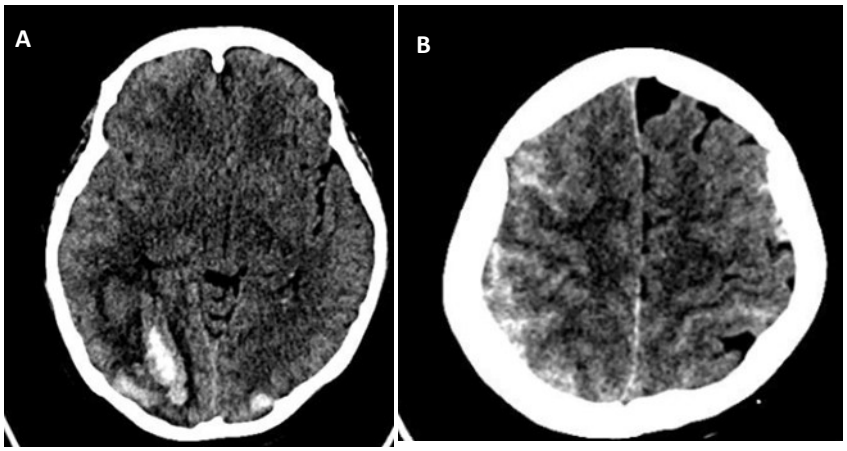
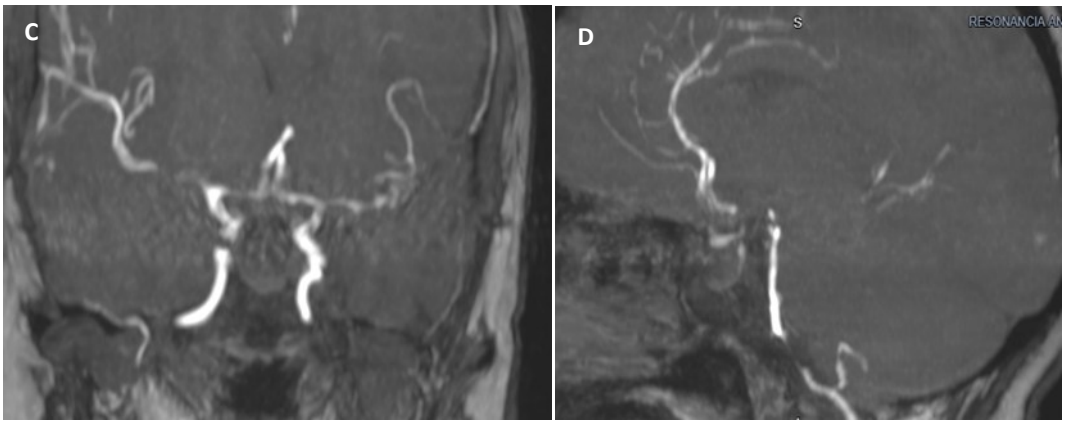


Figure 1. *A: Bilateral lobar hemorrhage. B: bilateral convexity subarachnoid hemorrhage (more pronounced in the right hemisphere).*

Step to the ward on her sixth day of admission**.** She remained asymptomatic until the 10th day of admission, when she presented with a confusional episode and multifocal neurological deficits with optic apraxia, ocular ataxia, and simultagnosia (Balint's syndrome), conjugate gaze deviation to the left, which could cross the midline with oculocephalic impulse maneuvers. Right arm and left leg were plegic. Left arm showed no deficit, and the right leg was paretic. MRI of the brain (Image 3) showed the presence of multifocal ischemic infarcts in both hemispheres, in both anterior and posterior circulation, with signs of irregularity of the walls in the middle cerebral arteries, anterior, and left posterior cerebral artery from segment P1, with decreased flow in segment P3 of the right posterior cerebral artery. The irregularity of the wall of the basilar artery in the distal tract was noted. No abnormalities were found in the extracranial vessels.



C-D: *Irregularity of the walls with diffuse narrowing in multiple vessels of the anterior and posterior circulation.*

Due to the multifocal deficit and alteration in the morphology of the wall in different arterial territories, a diagnosis of probable vasculitis affecting the central nervous system was proposed. Cardioembolic sources and systemic vessel involvement were excluded, and extensive studies of both autoimmune and infectious origins in serum and CSF were conducted, both showing no alterations.

Due to the multifocal neurological deficit without an identified cause, signs of angiitis on angio-MRI, and the absence of signs of systemic vasculitis or any other entity that could explain the clinical picture, a diagnosis of primary angiitis of the central nervous system was established, initiating treatment with Methylprednisolone 1g IV for 5 days and Cyclophosphamide. After the initiation of treatment, the clinical evolution was favorable. In follow-up angiography studies, improvement in the pattern of stenosis and multisegmented beading in the cerebral arteries was noted.

Currently, she has remained asymptomatic from a neurological standpoint, recovering nearly all motor and visual function, becoming independent in all activities of daily living.

Discussion

This case illustrates the various ways primary angiitis of the central nervous system can manifest, a condition that often escapes diagnosis. In this patient's context, simultaneous and diverse hemorrhagic manifestations (intraparenchymal hemorrhage, subarachnoid hemorrhage, subdural hematoma) were evident, followed by the appearance of multifocal ischemic lesions. clinically and syndromically the case shows how diverse this entity can be when presenting and why it should always be included in the differential diagnosis in similar clinical situations. (1-2)

We consider that the particularities of our case are the young age of the patient (average age of diagnosis is 50 years) and the predominance of multiple hemorrhagic pathology at presentation, being documented in two cohort studies by Salvarani and colleagues (Mayo Clinic cohort) that only 8 to 10% of patients have this as the initial clinical manifestation, and even rarer is bilateral subarachnoid hemorrhage. (3-4)

Conclusion

In conclusion, primary CNS angiitis constitutes a diagnostic challenge that every clinical neurologist must consider when addressing a patient with multifocal, complex, and varied neurovascular pathology; both ischemic and hemorrhagic. We believe that our case demonstrates several aspects of the spectrum of this entity that can guide clinical diagnosis and, primarily, timely treatment.

Conflict of Interest

The author declare no conflict of interest.

References

1. Salvarani C, Brown RD Jr, Calamia KT, et al. Primary central nervous system vasculitis: analysis of 101 patients. Ann Neurol 2007; 62:442.
2. Hajj-Ali RA, Singhal AB, Benseler S, Molloy E, Calabrese LH. Primary angiitis of the CNS. Lancet Neurol 2011;10:561–572.
3. Salvarani C, Hunder GG, Brown RD Jr. Primary Central Nervous System Vasculitis. N Engl J Med. 2024;391(11):1028-1037. doi:10.1056/NEJMra2314942.
4. Torres J, Loomis C, Cucchiara B, Smith M, Messé S. Diagnostic yield and safety of brain biopsy for suspected primary central nervous system angiitis. Stroke 2016;47:2127–2129.