



Diagnostic challenges in CNS vasculitis: insights from a case report

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OBJECTIVES

To analyse a challenging case of central nervous system vasculitis with a stroke-like clinical onset

MATERIALS & METHODS

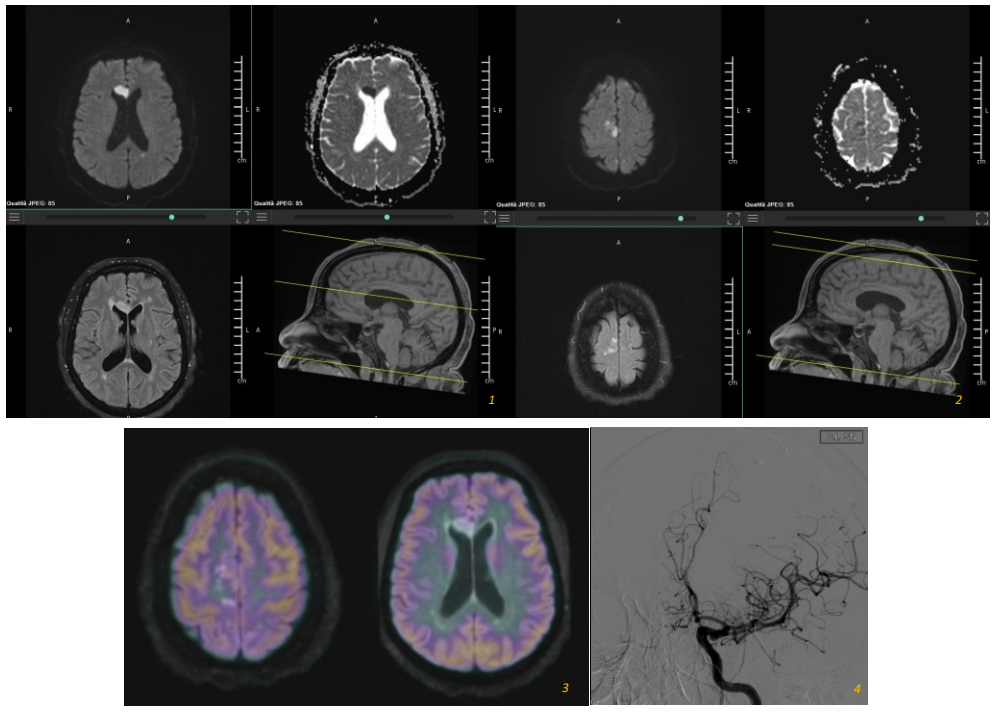
A 62-year-old Nigerian woman presented to the ER with progressive left hemiparesis:

- Recent history: vertigo, mild headache in the previous days.
- On admission: cognitive impairment and herpetic rash.
- PMH: hypertension, pre-diabetes, hypercholesterolemia.
- Diagnostic workup:
 - cardiovascular evaluation
 - serial neuroimaging
 - cerebral angiography
 - CSF analysis
 - infectious, autoimmune, neoplastic screening
 - brain PET.

- Neuroimaging:
- (1) MRI at symptoms onset
 - (2) Control MRI after 3 weeks
 - (3) PET-CT (with MRI reconstruction) after 3 weeks
 - (4) Cerebral Angiography after 3 weeks

RESULTS

- **DWI:** multiple recent lesions, predominantly in the right frontal gyrus and corpus callosum (NO features of demyelination/meningeal enhancement)
 - **Diabetes** was corrected **BUT** no other classic causes of ischemic lesions were found.
 - CSF investigations: elevated protein levels (type 3 OCB pattern), **BUT** no infectious agents nor evidence of systemic autoimmune process
 - **Retinal fluorescein angiography:** normal;
 - **audiometry test:** bilateral neurosensory hearing loss at 2-4kHz
 - Evidence of **pelvic mass + liver nodule with ↑ CA19.9**; preliminary findings suggest benign pathology (awaiting surgery) and negative liver histology
 - **Cerebral AGF:** angiography showed irregular stenosis of the right ACA with pericallosal occlusion and multiple contralateral stenosis.
 - **PET:** hypometabolism in the affected cerebral regions; non-specific findings on the whole-body scans
- Treatment:** : steroid + intensive rehabilitation → sensory-motor and cognitive functions progressively improved



DISCUSSION

- Rapid progression of left hemiparesis and cognitive decline, with MRI revealing **multiple bilateral recent ischemic lesions** (mainly in the **ACA-territory**)
- **Vascular risk factors** were present, including prior zoster infection, but **no** evidence of active viral involvement or systemic inflammatory / neoplastic causes was found.
- **Differential diagnoses** : embolic events, CNS vasculitis, Susac syndrome; brain biopsy was deferred, and ancillary tests excluded retinal and cochlear involvement.
- At the moment the patient is on a **slowly steroid-tapering regimen**, ambulates independently, awaiting gynecological surgery; follow-up MRI demonstrated no new ischemic lesions

CONCLUSION

- Clinical course and imaging supported a working diagnosis of **probable CNS vasculitis**; **current limitations:** absence of pathological confirmation, ongoing testing for secondary causes of vasculitis.
- **Follow-up** is crucial, particularly when biopsy is contraindicated by comorbidities, anatomical constraints, or procedural risks

References:
 M. Kowczyk et al. : Primary CNS vasculitis: A systematic review on clinical characteristics associated with abnormal biopsy and angiography - *Autoimmunity Reviews* - 2021 - Volume 20, Issue 1
 A. P. Shabeeb et al. : Primary angitis of the central nervous system : A challenging diagnosis - *Journal of Neuroimmunology* - 2022 - Volume 365
 G. Vandelli et al. : Intracranial Stenosis Treated with Stenting in Patients with Suspected Cerebral Vasculitis: Two Case Reports - *Case Reports in Neurology* - 2023 - 100-107

Abbreviation list
 ER: emergency department; PMH: past medical history; MRI: Magnetic Resonance Imaging; AGF: angiography; PET: positron-emission tomography; ACA: anterior cerebral artery; CNS: central nervous system; :



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