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Primary Angiitis of the Central Nervous System: Exclusion of Differentials and Long-Term Follow-Up

Josef Finsterer, MD*

Department of Neurology, Neurology & Neurophysiology Center, Vienna, Austria

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***Corresponding author**
Josef Finsterer, MD
Department of Neurology, Neurology & Neurophysiology Center, Postfach 20, 1180 Vienna, Austria.

Tel 43-1-5861075
Fax 43-1-5861075
E-mail fifigs1@yahoo.de

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With interest we read the article by Park et al. (1) on a 19-year-old male patient with primary angiitis of the central nervous system (PACNS), characterized by an irregular, rim-enhancing, necrotic mass with central diffusion-weighted imaging (DWI) hyperintensity in the corpus callosum and a peripheral DWI-hyperintense lesion in the left parieto-temporal periventricular area. Susceptibility-weighted imaging revealed multiple punctate bleedings within these lesions. A stereotactic biopsy confirmed the biopsy. The patient responded well to high-dose corticosteroids (1). While an impressive study, some points warrant further discussion.

Notably, long-term follow-up data is absent. While lesions reportedly reduced after two months of steroid therapy, what happened afterward? We need to know if glucocorticoids provided long-term efficacy or if additional treatment was necessary. Steroid treatment alone often is insufficient for PACNS. Was immune-modulating or immunosuppressive therapy, such as cyclophosphamide, rituximab, or mycophenolate mofetil, ever employed (2)? Given PACNS' potential to relapse and its 10% fatality rate (3), regular disease progression monitoring is crucial. What was the outcome after two years?

Adequately excluding other CNS vasculitides, such as giant-cell arteritis, Takayasu's arteritis, polyarteritis nodosa, anti-neutrophil cytoplasm antibody (ANCA)-associated vasculitis, cryoglobulinemic vasculitis, IgA vasculitis, and Behçet's disease is crucial (4). Did evidence suggest any of these types of CNS vasculitis?

Vasculitis parameters like anti-nuclear antibodies, ANCA, or circulating immune complexes were not determined in either serum or cerebrospinal fluid.

PACNS complications include ischemia and cerebral bleeding (5). Did imaging reveal any evidence of stroke or intracerebral bleeding in the patient? Additionally, PACNS can induce symptomatic epilepsy (6). Was his medical history positive for seizures? Did the electroencephalography show any epileptiform discharges?

Organ involvement beyond the brain has not been definitively ruled out. Before diagnosing PACNS, excluding extra-cerebral manifestations is essential.

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The patient did not undergo digital subtraction angiography, as recommended in the European Stroke Organization guidelines.

In conclusion, the excellent study has limitations that merit consideration before drawing conclusions. Addressing these weaknesses would strengthen the conclusions and potentially enhance the study. Confirming the absence of extra-cerebral manifestations is crucial before diagnosing PACNS. Patients with PACNS require close, long-term follow-up to avoid missing relapses.

Conflicts of Interest

The author has no potential conflicts of interest to disclose.

ORCID iD

Josef Finsterer https://orcid.org/0000-0003-2839-7305

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