

Primary Angiitis of the Central Nervous System

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Background:

- Primary angiitis of the central nervous system (PACNS) is a rare condition diagnosed by brain biopsy, as the clinical presentation can mimic several diseases involving the CNS.
- PACNS is a distinct entity, usually in the absence of systemic vasculitis.

Case Presentation:

- A 31-year-old man presented with abnormal behavior, right-sided headache, aphasia, progressive left-sided hemiparesis, and one episode of a generalized tonic-clonic seizure. Agitation and combative behavior followed. He actively used Kush (a preparation of a strain of Cannabis) for several years.
- He appeared somnolent. His physical exam was remarkable for reduced motor strength (1/5 in the LUE and 3/5 in the LLE), without pathological reflexes.
- The patient was treated empirically with intravenous antibiotics, and acyclovir for a presumptive diagnosis of acute viral meningoencephalitis.
- After his brain biopsy, he was started on IV methylprednisolone and cyclophosphamide infusions resulting in significant improvement.

Results:

- Elevated ESR and CRP.
- CSF studies showed lymphocytic pleocytosis.
- Infectious work-up including HIV was negative.
- ANA was 1:40, speckled, P-ANCA/C-ANCA were negative.
- MRI-Brain T2 Flair (**Fig.1**) showed hyperintensity in the right parietal lobe.
- Brain biopsy (**Fig.2**) revealed non-granulomatous small vessel vasculitis with transmural inflammation and partial vascular wall destruction, confirming the diagnosis of PACNS.

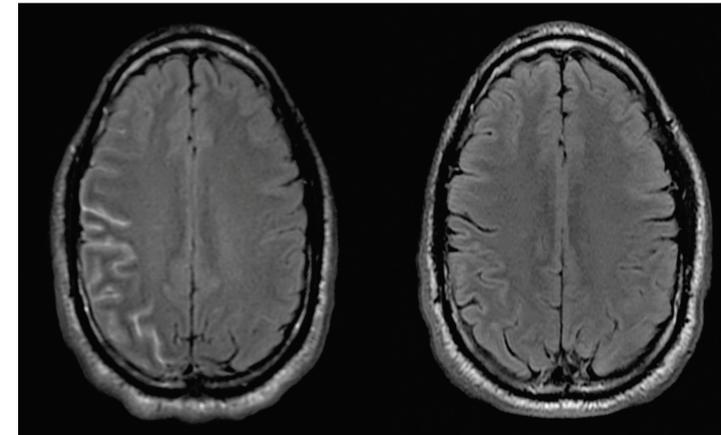


Figure 1: Initial brain MRI (left), axial T2 FLAIR, with hyperintensity evident in the region of the right parietal lobe. Two months post-treatment, the patient's brain MRI (right) demonstrated resolution of corresponding hyperintensity.

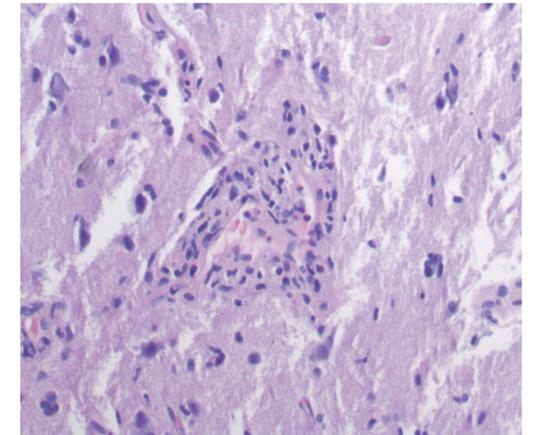


Figure 2: Hematoxylin and eosin (H&E) stain of tissue from brain biopsy, demonstrating a small vessel with transmural inflammatory infiltrate.

Conclusion:

- PACNS is considered a rare cause form of vasculitis in patients, often middle-aged men, without systemic vasculitis. It affects small and medium-sized leptomeningeal and parenchymal arterial vessels.
- Brain MRI findings are considered sensitive but not specific. Thus, the diagnosis is ascertained by brain biopsy.
- Some substances such as cocaine have been implicated in cases of PACNS, although the exact content of this patient's Kush is unknown.
- Standard treatment typically involves administration of glucocorticoids and cyclophosphamide. Early diagnosis and treatment is associated with favorable outcomes.

References:

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