

# A Case of Lymphoma Masquerading as Vasculitis

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## BACKGROUND

- A 61-year-old woman previously diagnosed with primary central nervous system (CNS) vasculitis via brain biopsy done for work up of new mass found following presentation in the emergency department for subacute left arm weakness and left-sided facial numbness
- Initial brain biopsy showed “lymphocytic vasculitis with hemorrhage, reactive vascularity, and reactive glial hyperplasia” with negative flow cytometry and culture data most consistent with primary CNS vasculitis
- Treatment consisted of high dose corticosteroids although tapering resulted in worsening left-sided weakness and radiographic worsening of known frontoparietal mass
- She was referred to rheumatology for steroid sparing agent, and she completed five months of oral cyclophosphamide with stabilization of mass lesion. She was then transitioned to mycophenolate for maintenance

## CASE

**Chief Complaint:** Confusion, gait changes, and worsening left-sided weakness

- **VS:** T 36.4C, BP 137/90, HR 89, RR 20, O2 sat 97% on room air
- **Physical exam findings:** Alert and oriented to person, place, and time. Notable for no cranial nerve deficits and normal sensory exam. Motor exam: 5/5 in RUE, RLE, 4+/5 in LUE w/ pronator drift, 5-/5 in LLE proximally, 5/5 distally

### Labs, Imaging, and Pathology:

- Previous laboratory work up for systemic vasculitis included: negative ANA, negative ANCA, negative RF, and normal ESR and CRP

### References:

1. Salvarani C, Brown RD Jr, Calamia KT, et al. Primary central nervous system vasculitis: analysis of 101 patients. *Ann Neurol*. 2007;62(5):442-451. doi:10.1002/ana.21226
2. Bhattacharyya S, Berkowitz AL. Primary angiitis of the central nervous system: avoiding misdiagnosis and missed diagnosis of a rare disease. *Pract Neurol*. 2016;16(3):195-200. doi:10.1136/practneurol-2015-001332

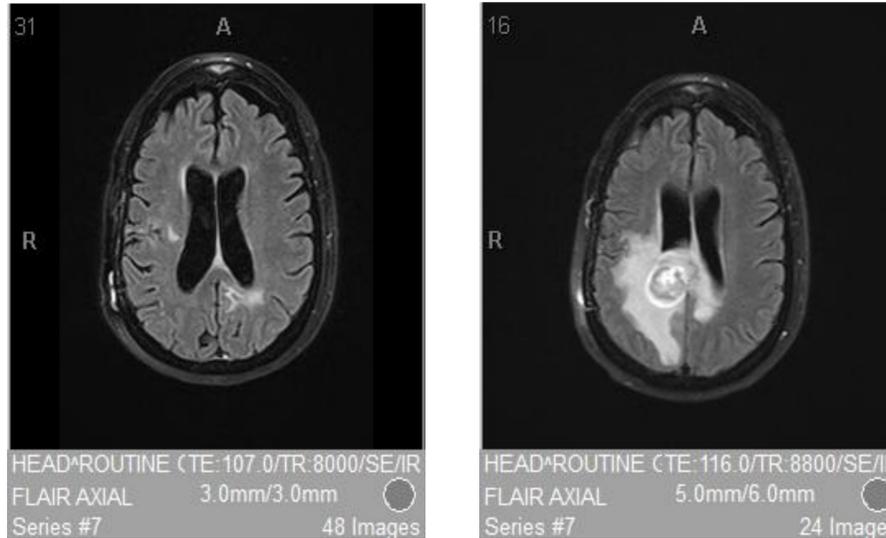


Figure 1. Comparison of MRI brain with IV contrast 5 months apart. MRI brain (left) demonstrates significantly improved left posterior callosal lesion from initial presentation while on cyclophosphamide. On presentation, MRI brain (right) with new enhancing mass-like lesion in the right corpus callosum. Thought to likely be an additional area of vasculitis with associated inflammation, however, CNS lymphoma could have a similar appearance

- **Given worsening mass lesion despite aggressive immunosuppression, repeat brain biopsy was advocated which showed primary diffuse large B-cell lymphoma (DLBCL) of the central nervous system**

## CONCLUSION

- Primary CNS vasculitis is a challenging condition given its rarity with a reported incidence of 2.4 cases per 1 million person-years and similar presentation to malignancy and infections<sup>1</sup>
- MRI is sensitive but not specific, cerebral angiography is neither sensitive nor specific, and CSF findings (typically elevated protein and pleocytosis) are sensitive but not specific<sup>2</sup>
- **Biopsy is the only way to establish a definitive diagnosis**, however, as exemplified in this case, pathologic findings (tissue destruction and necrosis) can be consistent with but not necessarily diagnostic of vasculitis
- **Failure to respond to immunosuppression warrants investigating an alternate diagnosis**

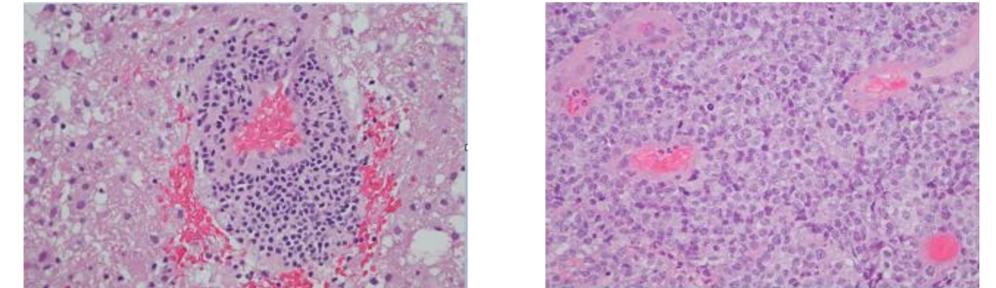


Figure 2. Comparison of brain biopsy histology 1.5 years apart. H&E; 400x objective. Pictured left with findings of lymphocytic vasculitis with hemorrhage, reactive vascularity, and reactive glial hyperplasia. Pictured right with findings of sheets of large B cells, with a perivascular pattern and diffuse growth pattern consistent with DLBCL

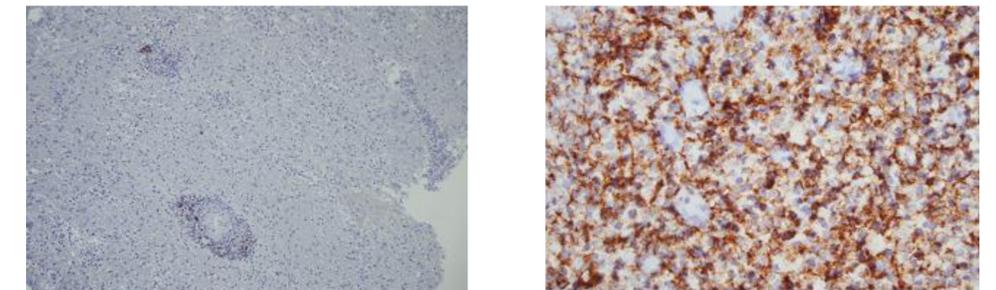


Figure 3. Comparison of brain biopsy histology 1.5 years apart with CD20 stain. Pictured right shows expression of CD20, seen in DLBCL

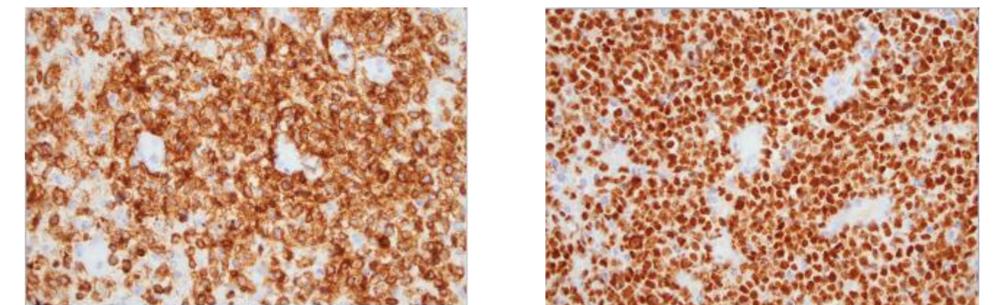


Figure 4. Expression of CD79a (left) and PAX5 (right), both markers of DLBCL