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**Neurology Publish Ahead of Print** 

DOI: 10.1212/WNL.0000000000206889

Teaching NeuroImage: Primary Central Nervous System Vasculitis Mimicking Intracranial Tumor

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*Neurology*® Published Ahead of Print articles have been peer reviewed and accepted for publication. This manuscript will be published in its final form after copyediting, page composition, and review of proofs. Errors that could affect the content may be corrected during these processes.

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**Contributions:** 

Hanlin Sun: Drafting/revision of the manuscript for content, including medical writing for content; Major role

in the acquisition of data; Study concept or design; Analysis or interpretation of data

Shujiang Zhang: Analysis or interpretation of data

Tianping Yu: Analysis or interpretation of data

Dong Zhou: Drafting/revision of the manuscript for content, including medical writing for content; Major role

in the acquisition of data

Jinmei Li: Drafting/revision of the manuscript for content, including medical writing for content; Major role in

the acquisition of data; Analysis or interpretation of data; Additional contributions:

Figure Count: 2

**Table Count:** 0

**Search Terms:** 

[ 120 ] MRI, [ 134 ] Vasculitis

Acknowledgment:

Study Funding: The authors report no targeted funding

**Disclosures:** The authors report no disclosures relevant to the manuscript.

**Preprint DOI:** 

**Received Date: 2022-07-22** 

**Accepted Date:** 

2022-12-20

### **Handling Editor Statement:**

Submitted and externally peer reviewed. The handling editor was Resident and Fellow Section Editor Whitley Aamodt, MD, MPH.

A 21-year-old man with headache, vomiting, and limb weakness presented to the clinic in two years ago. Examination showed paresthesia and weakness in left upper and lower limbs. Brain MRI demonstrated a large space-occupying lesion with ring enhancement and compression of the right fronto-tempo-parietal lobes (Figure 1, A - B). The patient underwent surgery for a presumed glioblastoma. Pathological examination revealed primary central vasculitis (PCNSV) without neoplasm (Figure 2). His screening workup for systemic vasculitis were negative. Symptoms improved after a corticosteroid taper. After stopping immunosuppressive therapy for one year, new lesions were found again in the right frontotemporal lobe (Figure 1, C - D). Corticosteroids and mycophenolate mofetil were given, and the patient's symptoms significantly improved and lesions on MRI had subsided significantly (Figure 1, E - F). MRI findings of PCNSV frequently present as nonspecific white matter lesions<sup>1</sup>. It can mimic glioblastoma<sup>2</sup>, CNS lymphoma and tumefactive multiple sclerosis<sup>1</sup>.

## **Appendix 1: Authors**

Name	Location	Contribution
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	Sichuan University	content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data
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	Sichuan University	medical writing for content; Major role in the acquisition of data
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	Sichuan University	medical writing for content; Major role in the acquisition of data; Analysis or
		interpretation of data

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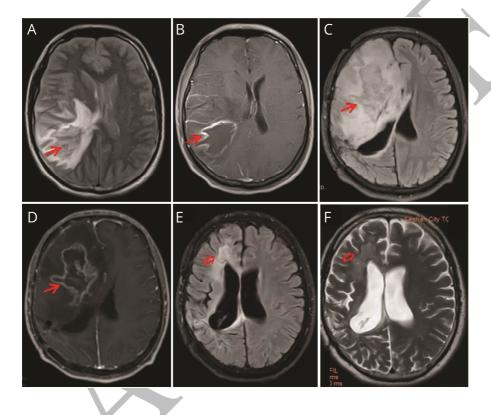
## **Reference:**

- 1. Hajj-Ali RA, Calabrese LH. Central nervous system vasculitis: advances in diagnosis. Curr Opin Rheumatol 2020;32:41-46.
  2. Jin H, Qu Y, Guo Z-N, Cui G-Z, Zhang F-L, Yang Y. Primary Angiitis of the Central Nervous System Mimicking Glioblastoma: A Case Report and Literature Review. Front Neurol 2019;10:1208.

## Figure legends:

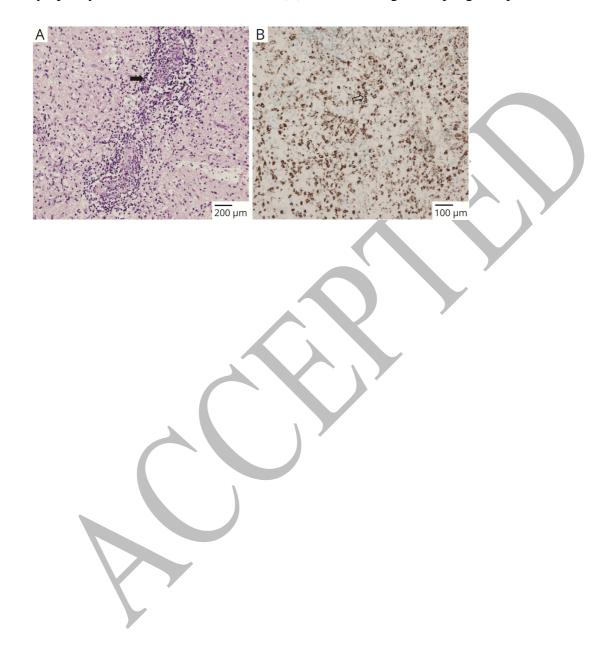
Figure 1 .MRI of Brain

Brain MRI demonstrates hyperintense irregular mass within the right frontotemporal and parietal lobes with perifocal edema and ring enhancement (A-B). A lesion appeared in the right frontotemporal lobe and corpus callosum, the anterior horn of the right ventricle was significantly compressed (C-D). MRI lesion largely disappeared after immunosuppressant therapy (E-F)



## Figure 2. The pathology slide

H&E (A) showed necrosis of small blood vessels with perivascular infiltrates of lymphocytes. Anti-CD68 immunostain (B) demonstrating macrophages expression.





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Hanlin Sun, Shujiang Zhang, Tianping Yu, et al. *Neurology* published online February 7, 2023 DOI 10.1212/WNL.000000000206889

## This information is current as of February 7, 2023

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