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Teaching NeuroImage: Primary Central Nervous System Vasculitis Mimicking Intracranial Tumor

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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

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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 lesions¹. It can mimic glioblastoma², CNS lymphoma and tumefactive multiple sclerosis¹.

Appendix 1: Authors

Name	Location	Contribution
Hanlin Sun ,M.D.	West China Hospital , Sichuan University	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
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Zhang ,M.D.	Sichuan University	Analysis or interpretation of data
Tianping Yu ,M.D.	West China Hospital , Sichuan University	Analysis or interpretation of data
Dong Zhou, Ph.D.	West China Hospital , Sichuan University	Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data
Jinmei Li, Ph.D.	West China Hospital , Sichuan University	Drafting/revision of the manuscript for content, including 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)

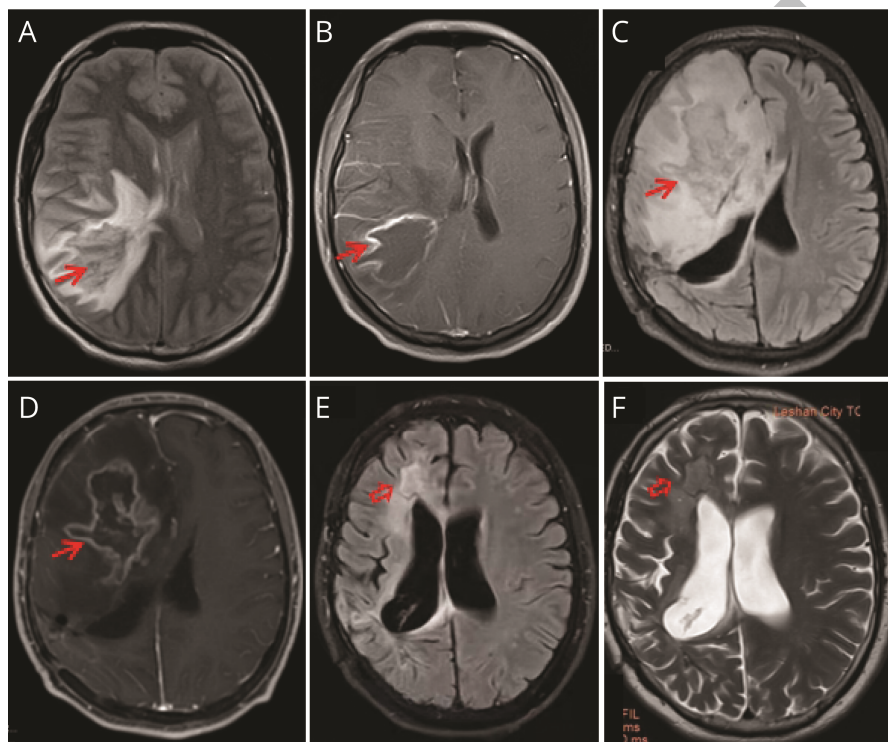
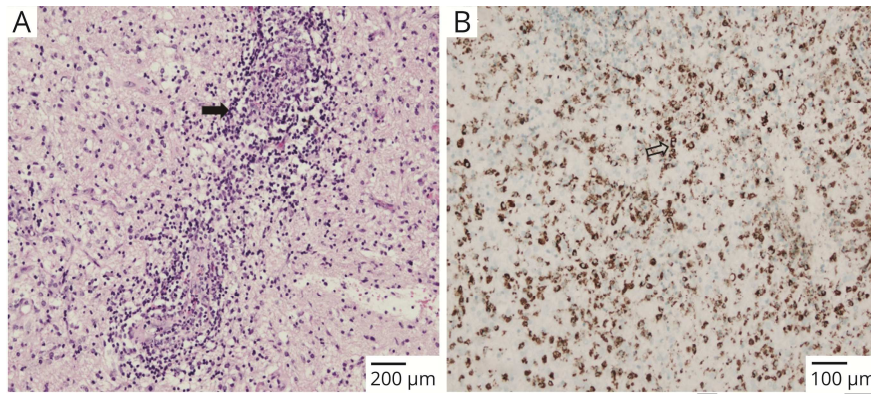


Figure 2. The pathology slide

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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