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Teaching NeuroImage: Perivascular radial enhancement in neurosarcoidosis

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A 78-year-old healthy female presented with abulia, aphasia, and echopraxia for one week. Brain MRI with contrast (Figure 1) revealed radial perivascular enhancement perpendicular to the ventricles. CSF showed 16 WBC (normal 0-5/ μ L), 1.18 g/L proteins (normal <0.45 g/L), CD4/CD8 ratio of 14.7 and no neoplastic cells. Extensive autoimmune, infectious, and neoplastic work-up was unremarkable. CSF and serum GFAP-IgG were negative. Brain biopsy (Figure 2) confirmed neurosarcoidosis. After corticosteroid treatment, MRI normalized (Figure 1) and patient substantially improved however remain disabled with a multi-domain cognitive impairment.

This linear radial periventricular enhancement MRI brain pattern is a hallmark of autoimmune glial fibrillary acidic protein (GFAP) astrocytopathy¹ and can also be seen with intravascular lymphoma, CNS vasculitis, and neurosarcoidosis², which are advisable to include in the differential diagnosis of patients with this MRI pattern, negative GFAP work-up, and no evidence of malignancy.

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Legends:

Figure 1. Post-gadolinium axial and sagittal T1-weighted images at baseline reveal linear perivascular enhancement (A, B), while post-gadolinium axial and sagittal T1-weighted images at 3 months after corticosteroid treatment show significant improvement of the previous MRI pattern (C, D).

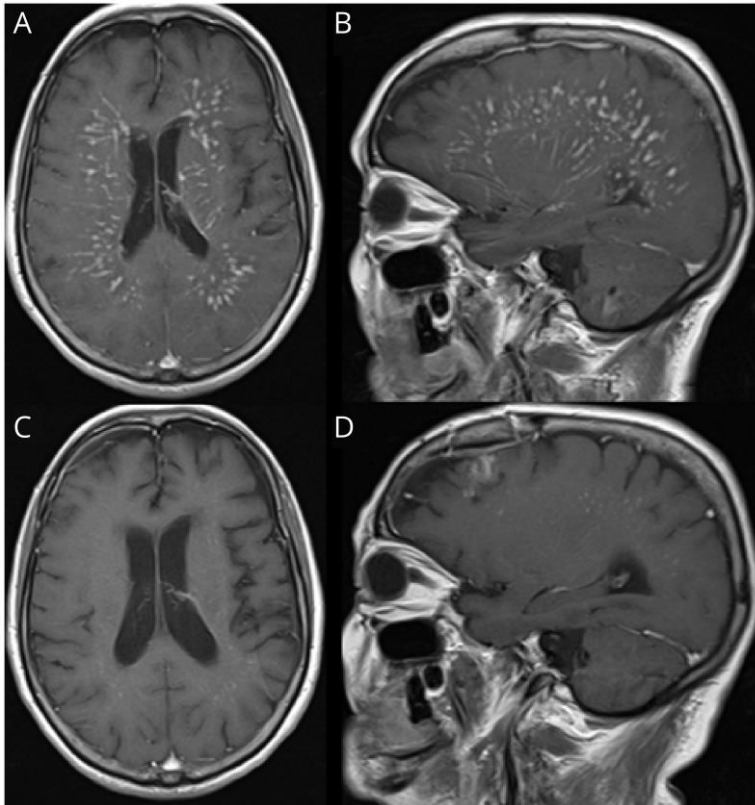
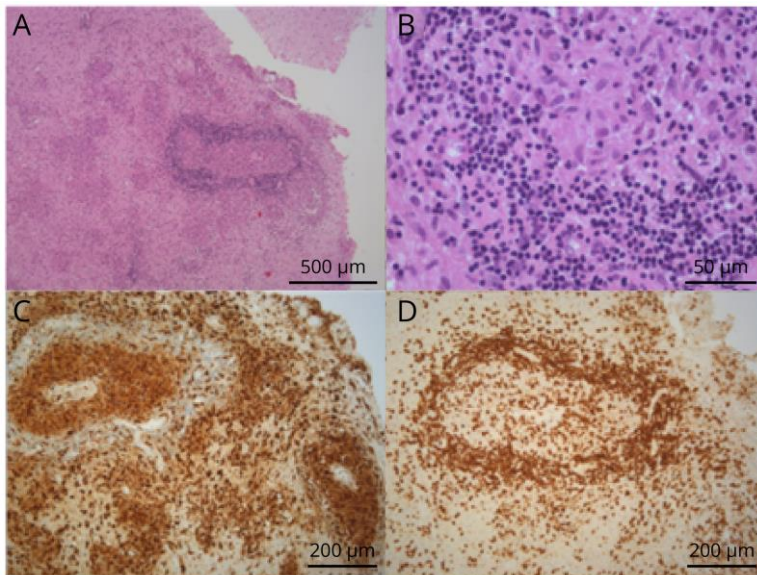


Figure 2. Brain biopsy in the right frontal lobe reveal white matter inflammatory granulomatous lesion with a histiocytic center and a ring of T lymphocytes (predominantly CD4+), associated with small inflammatory macrophagic lesions centered to white matter vessels. No fibrinoid necrosis or microorganisms (A and B – H&E; C – CD68; D – CD3).



Research ethics and informed consent:

This study was exempt from ethics review board approval.

Permission for publication was obtain from the legally authorized representative since the patient was unable to provide written informed consent.

WNL-2023-001259 slides --- <http://links.lww.com/WNL/D68>

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