Teaching NeuroImage: Perivascular radial enhancement in neurosarcoidosis

Author(s):
André A Costa, Medical Doctor¹; Catarina Silva, MD¹; Ricardo Taipa²; João Paulo Gabriel, MD¹; Michel Mendes, MD¹

Corresponding Author:
André A Costa, andre.ja.costa@hotmail.com

Affiliation Information for All Authors: 1. Neurology Department, Trás-os-Montes and Alto Douro Hospital Center, Vila Real, Portugal; 2. Neuropathology Unit, Porto Hospital and University Center, Porto, Portugal

Equal Author Contribution:

Contributions:
André Assunção Costa: Drafting/revision of the manuscript for content; including medical writing for content; Major role in the acquisition of data; Study concept or design; Additional contributions (in addition to one or more of the above criteria)
Catarina Silva: Drafting/revision of the manuscript for content; including medical writing for content; Major role in the acquisition of data; Additional contributions (in addition to one or more of the above criteria)
Ricardo Taipa: Major role in the acquisition of data; Analysis or interpretation of data; Additional contributions (in addition to one or more of the above criteria)
João Paulo Gabriel: Drafting/revision of the manuscript for content; including medical writing for content; Additional contributions (in addition to one or more of the above criteria)
Michel Mendes: Drafting/revision of the manuscript for content; including medical writing for content; Major role in the acquisition of data; Additional contributions (in addition to one or more of the above criteria)

Figure Count:
2

Table Count:
0
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/μL), 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) astrocytopathy\(^1\) and can also be seen with intravascular lymphoma, CNS vasculitis, and neurosarcoidosis\(^2\), which are advisable to include in the differential diagnosis of patients with this MRI pattern, negative GFAP work-up, and no evidence of malignancy.
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
References:


Teaching NeuroImage: Perivascular radial enhancement in neurosarcoidosis
André A Costa, Catarina Silva, Ricardo Taipa, et al.
*Neurology* published online August 29, 2023
DOI 10.1212/WNL.0000000000207830

This information is current as of August 29, 2023

<table>
<thead>
<tr>
<th>Updated Information &amp; Services</th>
<th>including high resolution figures, can be found at: <a href="http://n.neurology.org/content/early/2023/08/29/WNL.0000000000207830.citation.full">http://n.neurology.org/content/early/2023/08/29/WNL.0000000000207830.citation.full</a></th>
</tr>
</thead>
</table>
| Subspecialty Collections      | This article, along with others on similar topics, appears in the following collection(s):  
**All Cognitive Disorders/Dementia** [http://n.neurology.org/cgi/collection/all_cognitive_disorders_dementia](http://n.neurology.org/cgi/collection/all_cognitive_disorders_dementia)  
**MRI** [http://n.neurology.org/cgi/collection/mri](http://n.neurology.org/cgi/collection/mri)  
**Autoimmune diseases** [http://n.neurology.org/cgi/collection/autoimmune_diseases](http://n.neurology.org/cgi/collection/autoimmune_diseases) |
| Permissions & Licensing       | Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: [http://www.neurology.org/about/about_the_journal#permissions](http://www.neurology.org/about/about_the_journal#permissions) |
| Reprints                      | Information about ordering reprints can be found online: [http://n.neurology.org/subscribers/advertise](http://n.neurology.org/subscribers/advertise) |