Multicentric Benign Demyelinating Pseudotumor: The Great Masquerade

Joshua A. Cuoco, DO, MS^{1,2,3} , Evin L. Guilliams, DO^{1,2,3},
Cara M. Rogers, DO⁴, John J. Entwistle, DO^{1,2,3},
and Adeolu L. Olasunkanmi, MD^{1,2,3}

The Neurohospitalist
2020, Vol. 10(4) 326-328
© The Author(s) 2020
Article reuse guidelines:
sagepub.com/journals-permissions
DOI: 10.1177/1941874420923539
journals.sagepub.com/home/NHO

Keywords

demyelinating pseudotumor, inflammatory pseudotumor, lymphoma, metastasis, steroids

Figure 1. A-B, Initial MRI magnetization-prepared rapid acquisition gradient-echo (MP-RAGE) sequence with a large, vividly enhancing lesion within the body of the corpus callosum crossing midline with mass effect on the lateral ventricles and cingulate gyrus and a second lesion within the left middle cerebellar peduncle. C-D, Follow-up MRI MP-RAGE at 3 months post-methylprednisolone treatment with near resolution of both lesions.

¹ Carilion Clinic, Section of Neurosurgery, Roanoke, VA, USA

² Virginia Tech Carilion School of Medicine, Roanoke, VA, USA

³ Virginia Tech School of Neuroscience, Blacksburg, VA, USA

⁴ The University of Texas MD Anderson Cancer Center, Department of Neurosurgery, Houston, TX, USA

Corresponding Author:

Joshua A. Cuoco, Carilion Clinic, Section of Neurosurgery, 2331 Franklin Road, Roanoke, VA 24014, USA.
Email: jacuoco@carilionclinic.org

Figure 2. Histopathology consistent with benign demyelinating pseudotumor. (A) Hematoxylin and eosin stain at $\times 400$ magnification with hypercellular white matter and an abundance of cells with foamy cytoplasm. (B) Glial fibrillary acidic protein (GFAP) stains at $\times 400$ magnification with large stellate cells with processes consistent with reactive astrocytosis. Immunostains at $\times 400$ magnification with a high population of CD45 positive-cells (C) and CD68 positive-cells arranged in sheets (D).

A 72-year-old female presented to the emergency department with 1 week of progressively worsening nausea and vomiting. On examination, she was without neurologic deficit. Computer tomography of the head was without evidence of acute pathology and a basic workup was unrevealing for a source of her symptomatology. One week later, she represented to the emergency department with worsening complaints and overall fatigue. Given the progressive nature of her symptomatology, a magnetic resonance imaging (MRI) of the brain was ordered by the emergency department physician to evaluate for intracranial pathology. Magnetic resonance imaging demonstrated a large, vividly enhancing lesion within the body of the corpus callosum crossing midline with mass effect on the lateral ventricles and cingulate gyrus and a second vividly enhancing lesion within the left middle cerebellar peduncle with surrounding vasogenic edema (Figure 1A and B). Metastatic workup revealed a large mass involving the mid-to-distal sigmoid colon with regional lymphadenopathy consistent with primary colonic adenocarcinoma on biopsy. The intracranial lesions were suspected to be metastases versus primary central nervous system lymphoma.

Stereotactic biopsy of the periventricular lesion was performed to provide definitive tissue diagnosis and guide

treatment. Permanent sections revealed hypercellular white matter, an abundance of cells with foamy cytoplasm, and reactive astrocytosis with an inflammatory infiltrate (Figure 2). Immunostains demonstrated a high population of cluster of differentiation (CD) 68 positive-cells arranged in sheets consistent with histiocytes and CD45 positive-cells consistent with leukocytes. Many of the inflammatory cells were CD3 positive; however, only a few stained positive for CD20. Glial fibrillary acidic protein (GFAP) stains revealed large stellate cells with processes consistent with reactive astrocytosis: cells between astrocytes were negative for GFAP. Tests for other autoimmune and infectious etiologies, including cerebrospinal fluid analysis, were negative. These observations supported a diagnosis of benign demyelinating pseudotumor. Treatment consisted of intravenous methylprednisolone 1-gram daily for 7 days followed by a steroid taper. Follow-up MRI 3 months after initial diagnosis revealed near resolution of the intracranial lesions (Figure 1C and D).

Benign demyelinating pseudotumor of the central nervous system is a rare entity describing a reactive, inflammatory nonmalignant process that can mimic an intracranial neoplasm both clinically and radiographically.¹⁻³ Although there are no specific clinical or radiographic manifestations of these

lesions, in the majority of cases, subtle differences are known to exist between these pathologies. Intracranial demyelinating pseudotumor is often found in young and middle-aged individuals, presents acutely with a short duration of symptomatology, and typically exhibits minimal vasogenic edema and mass effect with lesion enhancement perpendicular to the surface of the lateral ventricle.^{1,2} However, there are no characteristic locations within the intracranial compartment where pseudotumors are known to occur.^{1,2} Comparatively, intracranial neoplasms, such as astrocytomas, generally present in middle-aged to older individuals, exhibit slower progression with a longer duration of symptomatology and demonstrate a significant amount of vasogenic edema surrounding the lesion with varying degrees of enhancement.^{2,4}

As depicted in the present case, benign demyelinating pseudotumors are often sensitive to a short course of steroid treatment. Kepes described the outcomes of 31 patients with biopsy-confirmed benign demyelinating pseudotumor treated with corticosteroid therapy.⁴ Of the 31 cases treated with corticosteroids, 28 cases showed no recurrence at 12-year follow-up.⁴ There remains no consensus regarding a standard steroid regimen for the treatment of these lesions with regimens remaining prescriber dependent.^{1,2} This is contrast to the majority of brain neoplasms that do not respond to corticosteroids.

Here, we presented an unusual and exceedingly rare case of multicentric benign demyelinating pseudotumor. Certainly, metastases and primary central nervous system lymphoma were highly considered in the differential diagnosis given the identified sigmoid colon mass on metastatic workup and vividly enhancing periventricular lesions, respectively. However, neither diagnosis was confirmed on histopathology. Although subtle clinical and radiographic differences exist

between pseudotumor and intracranial neoplasms, the present case highlights that such distinctions are not absolute. Indeed, histopathology and immunohistochemical staining are obligatory to reliably distinguish between demyelinating pseudotumor and neoplastic tissue as treatment modalities for these pathologies differ significantly.

Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

ORCID iD

Joshua A. Cuoco, DO, MS <https://orcid.org/0000-0002-7118-3890>

References

1. Ning X, Zhao C, Wang C, Zhang D, Luo Q. Intracranial demyelinating pseudotumor: a case report and review of the literature. *Turk Neurosurg*. 2017;27(1):146-150. doi:10.5137/1019-5149.JTN.10920-14.0
2. Ma Y, Mou H, Cai X, Ma J, Lu G. Primary inflammatory demyelinating pseudotumor in the left frontal lobe with meningeal involvement presenting as malignant neoplasms. *Chinese Neurosurg J*. 2016;2(1):5. doi:10.1186/s41016-016-0024-9
3. Jaffe SL, Minagar A. Demyelinating pseudotumor. *Arch Neurol*. 2005;62(9):1466-1467. doi:10.1001/archneur.62.9.1466
4. Kepes JJ. Large focal tumor-like demyelinating lesions of the brain: intermediate entity between multiple sclerosis and acute disseminated encephalomyelitis? A study of 31 patients. *Ann Neurol*. 1993;33(1):18-27. doi:10.1002/ana.410330105