Neuroimaging Highlight

Editors: Mark Hudon, Richard Farb

Tumefactive Demyelinating Lesions

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A 40-year-old right-handed female presented with a three-week history of abrupt onset, persistent right-sided sensory alteration. She described a sense of heaviness and numbness in the right face, arm, and part of her right leg. She also noted intermittent expressive speech difficulty. On examination, she was found to have a mild right-sided facial droop and impairment of all sensory modalities including light touch, pinprick, vibration, and proprioception throughout her right side. The remainder of her neurological exam was normal and speech was fluent.

An MRI was obtained, which revealed a solitary 2.4 x 2.6 cm lesion in the deep left parietal lobe with minimal positive mass effect. The lesion had an incomplete periphery or rim of low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. The lesion's central core had slightly greater signal intensity on T1-weighted images and relatively lesser signal intensity on T2-weighted images (Figures 1, 2). There was a small focus of contrast enhancement at the superomedial aspect of the lesion (Figure 3). Diffusion-weighted imaging (DWI) showed increased diffusion at the periphery consistent with a probable cystic nature (Figure 4). No other lesions were identified. The MR angiography was normal. Differential diagnosis for the lesion included neoplastic, infectious, granulomatous, and demyelinating diseases.

Two months following her initial MRI, the patient underwent stereotactic biopsy using an intra-operative MRI scanner. The baseline intra-operative MR images revealed a new lesion in the right posterior frontal white matter measuring 2.4 x 1.7 cm with patchy rim enhancement (Figure 5 and 6), in addition to the known left parietal lesion. Since the new lesion was in the non-dominant hemisphere and relatively superficial, a biopsy targeting this lesion was performed as it was felt to likely represent the same etiology. Pathology revealed sheets of macrophages intermixed with lymphocytes, plasma cells, and reactive astrocytes. In addition, there was perivascular lymphocytic inflammation with sharply defined foci of almost complete loss of myelin and relative preservation of axons. These findings were consistent with the diagnosis of a demyelinating plaque.

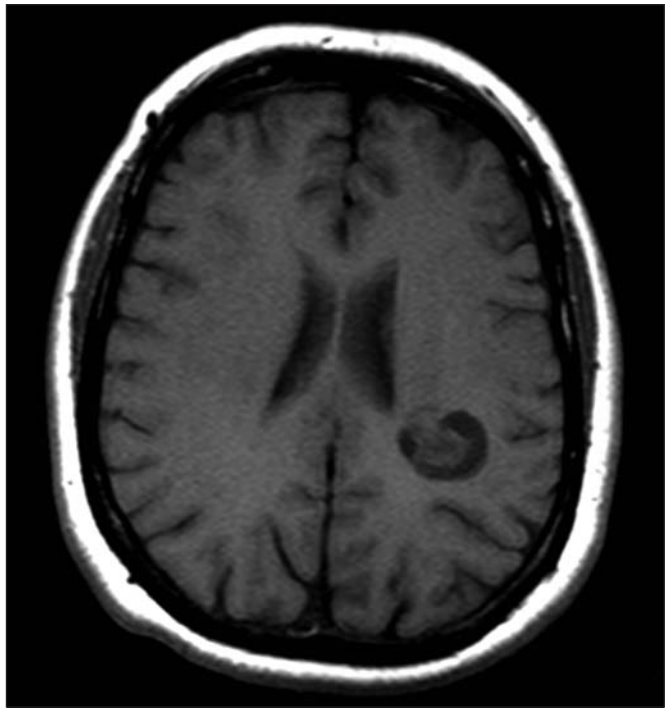

Figure 1: *Axial T1-weighted image showing a deep left parietal lobe white matter lesion with a hypointense periphery surrounding a slightly hyperintense core.*

Inflammatory demyelinting lesions occasionally present as tumefactive lesions that mimic brain tumors or cerebral

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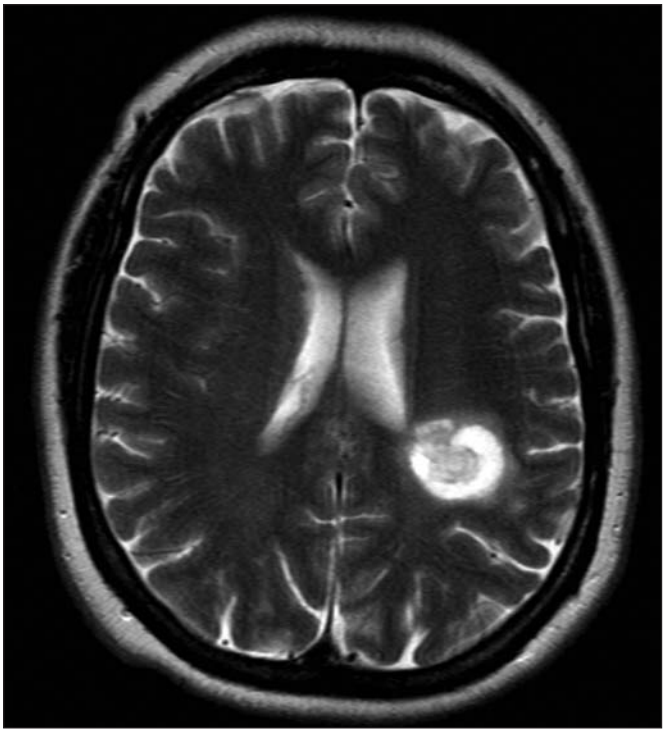

Figure 2: *Axial T2-weighted image showing a rim of high signal intensity and a relative hypointense core.*

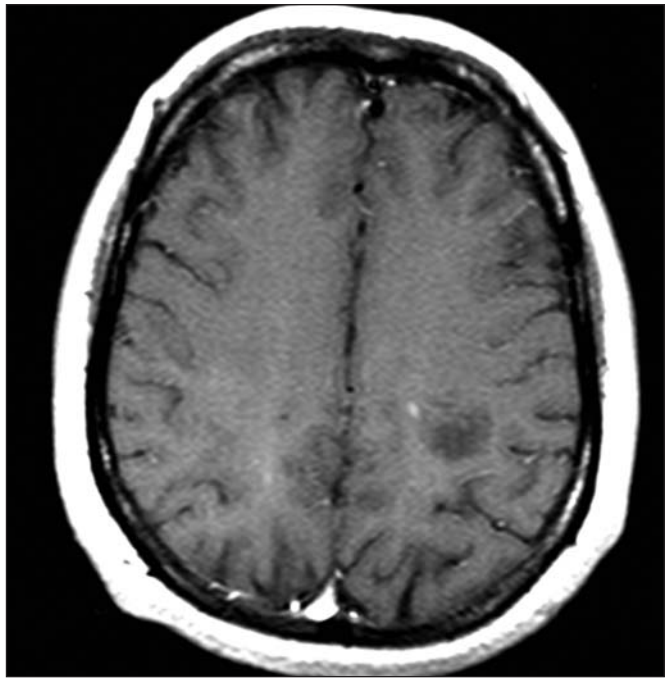

Figure 3: *Axial T1-weighted post-gadolinium image showing a small focus of enhancement at the superomedial aspect of the lesion.*

abscesses. Histopathologically, tumefactive lesions can be confused with high grade neoplasms due to the presence of hypercellularity and atypical reactive astrocytes and mitotic figures.[1-3] The low incidence of oligoclonal protein in the CSF of patients with tumefactive lesions can also obscure the diagnosis. Radiographically, tumefactive demyelinating lesions may resemble tumor or abscess on T1, T2, and DWI images.

Certain imaging characteristics, however, can be helpful in distinguishing tumefactive demyelinating lesions from tumor or abscess. For instance, tumefactive demyelinating lesions are

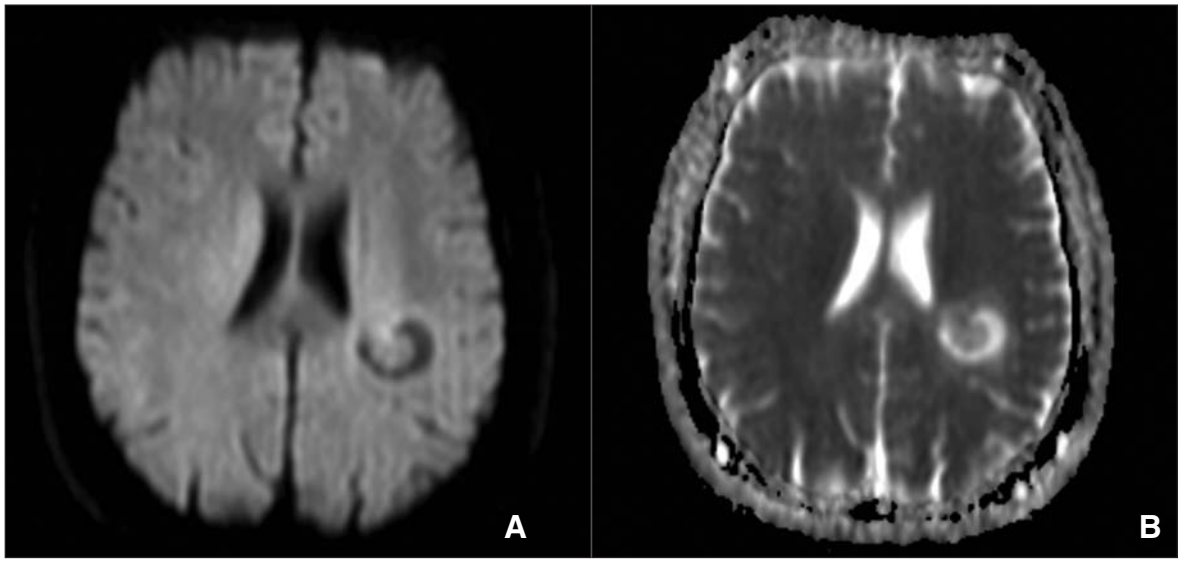

Figure 4: *Axial (A) diffusion-weighted and (B) corrected ADC map images demonstrate the central portion of the lesion to appear isointense with brain, while the peripheral portion shows some increased diffusion, compatible with a cystic lesion.*

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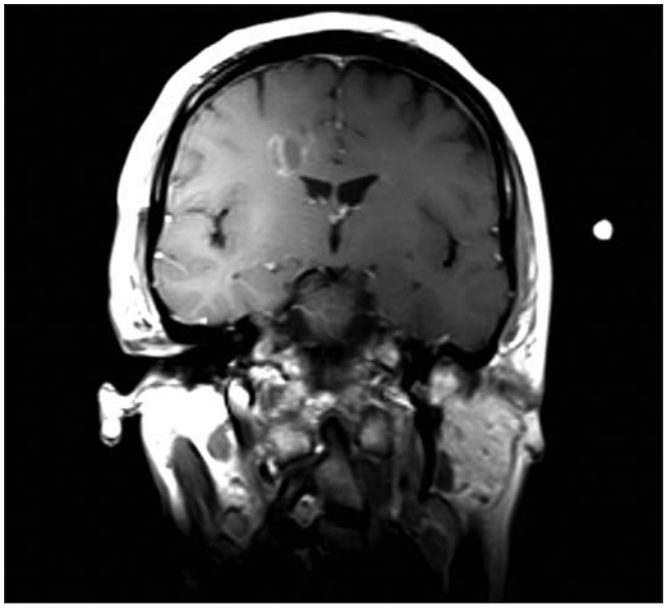

Figure 5: *Intra-operative axial T1-weighted post-gadolinium image demonstrating a new right frontal white matter lesion with patchy rim enhancement.*

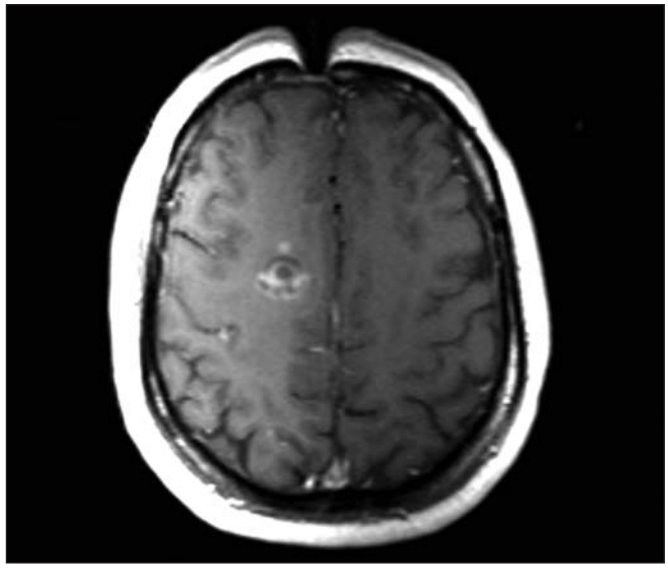

Figure 6: *Intra-operative coronal T1-weighted post-gadolinium image revealing the same right frontal white matter lesion as in Figure 5, with patchy rim enhancement.*

typically found in the periventricular white matter, corpus callosum, brainstem and middle cerebellar peduncles[1] and tend to be circumscribed with little positive mass effect or vasogenic edema.[4] They commonly have an enhancement pattern in the form of an open-ring with the incomplete portion of the ring on the gray matter side of the lesion.[4,5] The enhancing portion of the ring, which corresponds to transient impairment of the blood-brain barrier (BBB) associated with inflammatory infiltration, is believed to be the leading edge of demyelination and is, therefore, found on the white matter, or deeper side of the lesion.[4,6] The central, non-enhancing core represents a more chronic phase of inflammation, with complete or partial repair of the BBB.[6] Despite the presence of these imaging features it often remains difficult to differentiate pathologies by MRI criteria.

Newer physiologic MR imaging sequences can help to differentiate these lesions. Perfusion-weighted MR imaging can be used to calculate relative cerebral blood volume (rCBV) maps which correlate with metabolic activity. Studies have shown rCBV values[7] between tumefactive demyelination and neoplasm to be significantly different.[6] In addition, serial proton MR Spectroscopy has been useful in the diagnosis, given distinct chemical resonance spectra between gliomas and chronic tumefactive plaques.[8,9] The extent that new MR imaging sequences will replace biopsy in the diagnosis of tumefactive appearing lesions remains uncertain.

References

1. Tan HM, Chan LL, Chuah KL, Goh NS, Tang KK. Monophasic, solitary tumefacture demyelinating lesion: neuroimaging features and neuropathological diagnosis. Br J Radiol. 2004; 77(914): 153-6.
2. Tsui EY, Leung WH, Chan JH, Cheung YK, Ng SH. Tumefactive demyelinating lesions by combined perfusion-weighted and diffusion weighted imaging. Comput Med Imaging Graph. 2002; 26(5): 343-6.
3. Zagzag D, Miller DC, Kleinman GM, Abati A, Donnenfeld H, Budzilovich GN. Demyelinating disease versus tumor in surgical neuropathology. Clues to a correct pathological diagnosis. Am J Surg Pathol. 1993; 17(6): 537-45.
4. Given CA, Stevens BS, Lee, C. The MRI appearance of tumefactive demyelinating lesions. AJR Am J Roentgenol. 2004; 182(1): 195-9.
5. Masdeu JC, Moreira J, Trasi S, Visintainer P, Cavaliere R, Grundman M. The open ring. A new imaging sign in demyelinating disease. J Neuroimaging. 1996; 6(2): 104-7.
6. He J, Grossman RI, Ge Y, Mannon LJ. Enhancing patterns in multiple sclerosis: evolution and persistence. AJNR Am J Neuroradiol. 2001; 22(4): 664-9.
7. Cha S, Pierce S, Knopp EA, Johnson G, Yang C, Ton A, et al. Dynamic contrast-enhanced T2*-weighted MR imaging of tumefactive demyelinating lesions. AJNR Am J Neuroradiol. 2001; 22(6): 1109-16.
8. Butteriss DJ, Ismail A, Ellison DW, Birchall D. Use of serial proton magnetic resonance spectroscopy to differentiate low grade glioma from tumefactive plaque in a patient with multiple sclerosis. Br J Radiol. 2003; 76(909): 662-5.
9. Enzinger C, Strasser-Fuchs S, Ropele S, Kapeller P, Kleinert R, Fazekas F. Tumefactive demyelinating lesions: conventional and advanced magnetic resonance imagery. Mult Scler. 2005; 11(2): 135-9.

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