Case Report

NRJ

The Neuroradiology Journal
2025, Vol. 0(0) 1–5

DOI: 10.1177/19714009251356989
journals.sagepub.com/home/neu
Sage

Association of a giant developmental venous anomaly and acute disseminated encephalomyelitis: A case report and magnetic resonance perfusion study

Louis Deprez[1] and Emilie Lommers[2]

Abstract

We describe the first reported association between acute disseminated encephalomyelitis (ADEM) and giant developmental venous anomaly (DVA) in the context of myelin oligodendrocyte associated glycoprotein (MOG) associated disorder (MOGAD). Patient was a young woman presenting with headache, bradypsychia and tetrapyramidal syndrome. Imaging showed disseminated tumefactive inflammatory lesions in the brain and spinal cord, with a massive right frontal lobe lesion centred around a giant DVA. Demyelinating inflammatory lesions are known to occur in a perivenular pattern, and the association between some inflammatory diseases such as multiple sclerosis (MS) and DVA has already been described. Developmental venous anomalies are variant of the normal venous drainage of the brain, responsible of a local alteration of the venular network, and micro-perfusion anomalies as well as possible increased of blood–brain barrier permeability. As such, they might be responsible for a favourable environment for pathogenic auto-antibodies penetrance in such region, potentializing the inflammatory lesion size. Perfusion imaging showed a significant increase in regional blood volume and blood transit time in the DVA and the surrounding brain tissue, which regressed in the follow-up imaging studies after the acute stage. This case illustrates the potential role of DVA in the setting of demyelinating diseases, and its consequences on the local micro-perfusion of the brain, evolving between the acute and chronic phase of the illness.

Keywords

ADEM, MOGAD, developmental venous anomaly, perfusion imaging, demyelination

Case report

A 31-year-old female patient was admitted to the emergency room for headache, rated as 8–9 out of 10. She described a frontal and occipital pulsatile headache with bilateral facial pain in the last 2 days. Pain was worsened by eye movement, while lying down and while bending down. It did not respond to classic antalgic medication (paracetamol and NSAID). She experienced nausea and vomiting on the day of admission. She had a recent history of rhino-sinusitis. She had no medical history.

Clinical examination demonstrated a tetrapyramidal syndrome with left proportional hemiparesis as well as bradypsychia. She had no diplopia or visual acuity impairment. Fundus examination was normal.

Preliminary biological analyses including serologies were unremarkable. Cerebrospinal fluid (CSF) analyses demonstrated lymphocytic pleocytosis (59/mm^3) and a few oligoclonal bands within serum and CSF indicating blood–brain barrier (BBB) permeability. Electro-encephalogram showed right frontal slow waves focalization without epilepsy.

A computed tomography (CT) brain scanner, without and with iodinated contrast injection, was acquired and showed a right frontal lobe subcortical hypodensity, localized around a developmental venous anomaly (DVA) (Figure 1).

Complementary emergent brain magnetic resonance imaging scan (MRI) was realized revealing disseminated subcortical round-shaped lesions in hyper fluid attenuated inversion recovery (FLAIR) and T2 signal, with a massive lesion in the right frontal lobe (Figure 2) of 50 mm of transverse axis. Inside this parenchymal lesion was a huge DVA, appearing in hyposignal on the FLAIR, T2 and susceptibility weighted imaging (SWI) sequences, and vividly enhancing after gadolinium intravenous injection. Gradient-echo T1 sequence after gadolinium injection showed numerous hyper-intense vascular formations inside the area of the FLAIR lesion, corresponding to a huge abnormal venous network related to the DVA.

This pattern was recognized as acute demyelinating encephalomyelitis (ADEM), with a massive frontal lobe lesion in the context of a pre-existing lobar developmental venous anomaly (DVA). Fixed cell-based assay turned out to be positive for anti-MOG antibodies, thus confirming the diagnosis of MOGAD with ADEM phenotype at onset.

During her hospitalization, she had multiple further imaging examinations, including spine MRI (showing multiple demyelinating lesions, Figure 2), dynamic susceptibility

[1]Departement of Radiology, CHU de Liège, Belgium
[2]Department of Neurology, CHU de Liège, Belgium

Corresponding author:
Louis Deprez, Departement of Radiology, CHU de Liège, Service de Radiodiagnostic, 1 Avenue de l'Hôpital, Liège 4000, Belgium.
Email: louis.deprez@chuliege.be

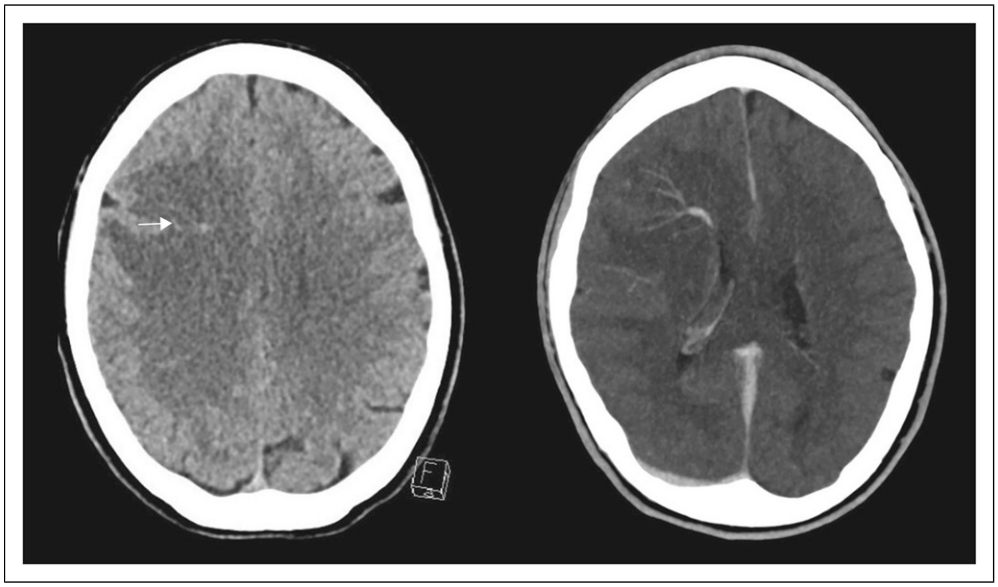

Figure 1. Left panel: emergent non-contrast-enhanced computed tomography (CT) brain scan, showing a massive hypodense lesion in the right frontal lobe. Right panel: contrast-enhanced CT, showing a giant DVA in the right frontal lobe.

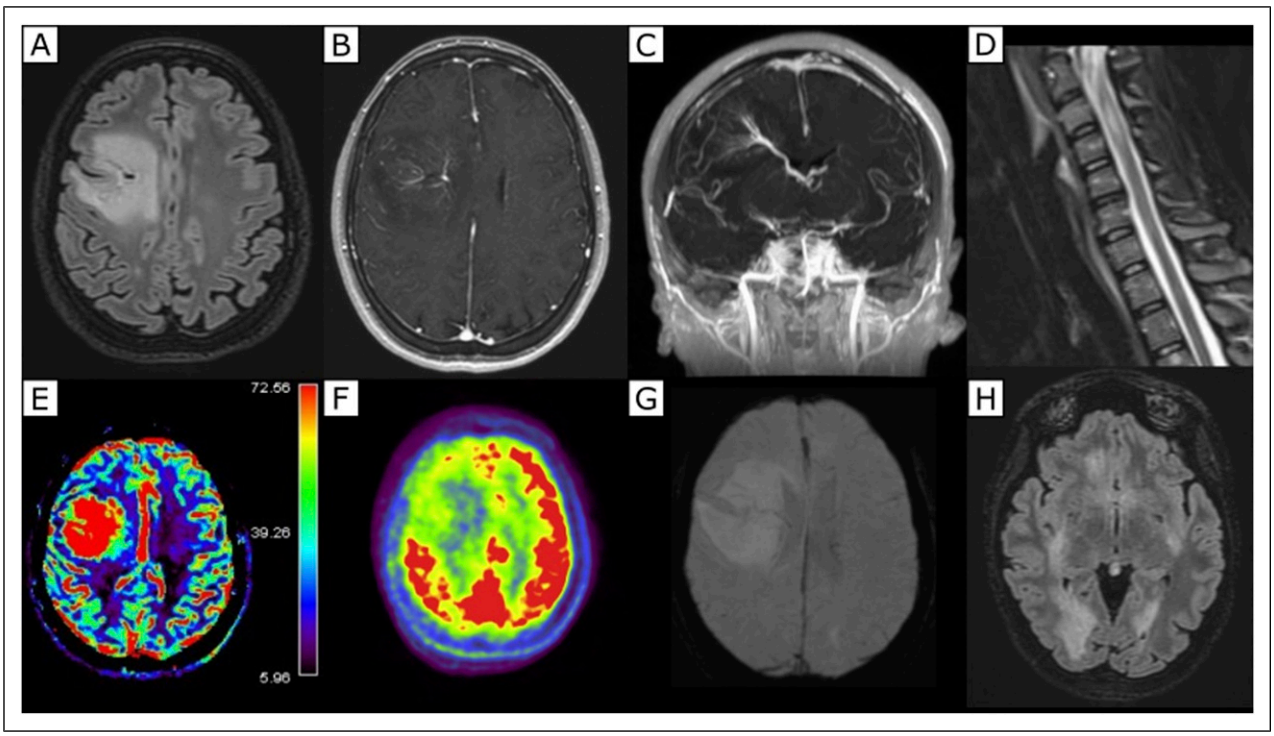

Figure 2. (A) FLAIR ponderated sequence, showing a massive tumefactive inflammatory lesion in the right frontal lobe. (B) Gradient-echo T1 ponderated sequence with gadolinium, showing the right frontal lobe DVA. (C) Frontal maximum intensity projection (MIP) showing the same DVA. (D) Short TI Inversion recovery sagittal sequence, showing an hyper-T2 lesion in the medullar cord at C3–C4 level. (E) rCBV map, showing a massive increase in relative blood volume in the area of the DVA and inflammatory lesion. (F) FDG-PET scan showing a decrease in brain glucose intake in the right frontal lobe. (G) SWI MIP sequence, showing a relatively low vascular hyposignal in the area of the DVA. (H) FLAIR ponderated sequence, showing multiples other ADEM lesions in hypersignal trough the brain.

contrast (DSC) perfusion imaging and positron-emitted tomography with computed tomography (PET-CT) (Figure 2).

Patient was treated with high-dose intravenous (IV) corticosteroids (1 gram once daily for 5 days) with slow oral tapering. She also underwent six plasma exchanges quickly followed by a first infusion of B cell depletion therapy (rituximab 1 g two weeks apart).

Clinical recovery was almost complete at 6 months, with persistence of slight attention deficit.

Follow-up imaging showed a near-disparition of the frontal lobe lesion and a decrease of its perfusion parameters (Figure 3). Arterial spin labelling (ASL) imaging was normal (not shown, not done prior in the acute phase).

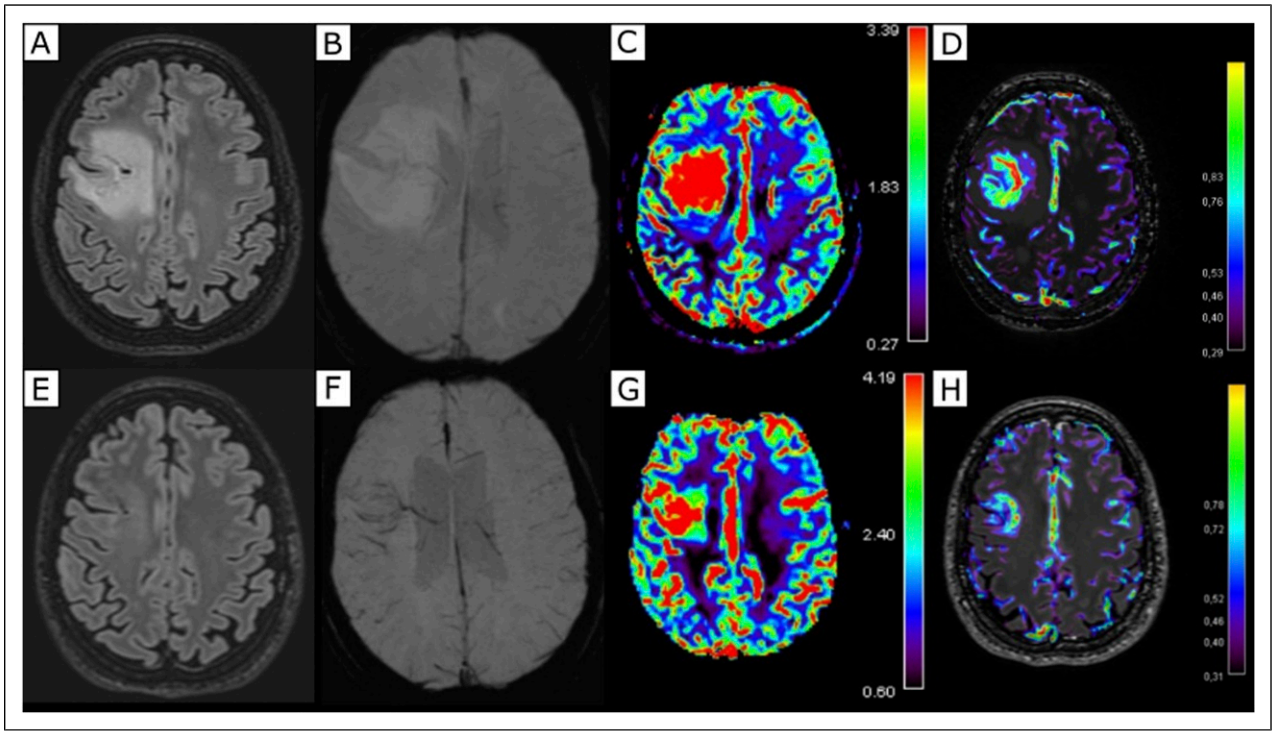

Figure 3. Top row: acute magnetic resonance examination. Bottom row: 3 months follow-up examination. (A and E) FLAIR ponderated sequence, showing the regression of oedema between the acute (A) and follow-up (E) examinations. (B and F) SWI MIP sequence, showing the relative increase in hyposignal the DVA venular network and main collector between acute (B) and follow-up (F) exams. (C and G) rCBV maps, showing a relative decrease of regional blood volume in the DVA between acute and follow-up exam. (D and H) tMIP maps, showing relative decrease in the slowness of the regional blood drainage in the DVA area between acute and follow-up exams.

Discussion

Physiopathology

Developmental venous anomalies (DVAs) are variants of normal transmedullary veins that drain the normal brain parenchyma.[1,2] They are organized around a main collector body, with a corolla of small draining vessels disposed around the distal extremity of the main collector, responsible for its 'caput medusa' appearance. They are a frequent incidental finding on imaging brain studies, with a supposed incidence of 2.6% on imaging and autopsy findings.[3] They are the result of incomplete regression of foetal temporary veins in the foetus, and modify the local venous drainage of the part of the brain they are in.

They are mostly benign and asymptomatic findings, even though some symptomatic cases have been reported, mostly by thrombosis[4] and haemorrhage.[5] Some rare cases report of headache, seizure, focal neurological deficits, dizziness and ataxia.[6] Their association with brain cavernomas has been extensively described.[7]

Although mostly benign, some studies showed that brain tissue surrounding DVA is abnormal in most cases with patients demonstrated surrounding white matter abnormalities, evident on brain MRI as T2/FLAIR hyperintensities[8] as well as PET-CT hypometabolism in 75% of cases.[9] The mechanisms underlying these changes are various and include venous congestion (due to a modification of the normal venous drainage of the brain parenchyma) and vasogenic oedema, which can cause chronical micro-ischaemic and haemorrhagic injuries of the surrounding brain.[10] Those chronic injuries may promote neoendothelial development, white matter gliosis and demyelination.[11]

The predisposition for demyelination to occur around normally distributed cerebral veins has been extensively described,[12,13] particularly in multiple sclerosis (MS).[14] Perivenular inflammation represents a hallmark of MS pathology, as confirmed by the central vein sign on susceptibility weighted imaging.[15,16] The association between DVA and MS lesions has been reported multiple times,[17,18] suggesting that DVA may create a compelling milieu increasing the propensity to lymphocytic infiltration and so, demyelination.

In contrast to MS, acute disseminated encephalomyelitis is a usually monophasic acute immune-mediated disease of the central nervous system, characterized in imaging by multiple white matter tumefactive lesions.[19–21] It is most often seen in the paediatric population,[20] but can happen in young adults.[22,23] Differential diagnosis of tumefactive lesions in demyelinating disorders may be difficult on imaging only,[24,25] and lab tests are required to confirm the diagnosis.[26] It is a classical first clinical manifestation of MOG associated disorder[27–29] and needs to be treated with high-dose corticosteroid therapy and plasma exchange.

To our knowledge, this report is the first published association of cerebral DVA and tumefactive demyelinating lesion in the context of positive MOG antibodies. Different from what is observed in MS, MOG associated WM demyelinated lesions exhibit a confluent pattern around small veins undetectable by MRI.[30] In the context of DVA, we can hypothesize that abnormal venous drainage may favour both lymphocytic infiltration and MOG antibodies

penetrance thus increasing the risk to develop ADEM-like phenotype with perivenous demyelination as suggested by Lassmann et al.[31]

Imaging features

We performed a dynamic susceptibility contrast (DSC) perfusion study in both the initial MRI and in the follow-up MRI, after the resolution of the initial oedematous lesion (as shown by the reduction in FLAIR hypersignal between both examinations). On the initial examination, the DVA and its neighbouring region were associated with a massive increase in corrected regional blood volume (rCBV), and an increase of temporal parameters (as shown in Figure 3 with the tMIP map).

Those findings are similar to those reported in the literature[32–34] and can correspond to venous congestion venular network draining in the main collector of the DVA. Some studies have demonstrated that the vessels of the DVA have different vessel wall characteristics than normal vessels, with thickened and hyalinized vessel walls, lacking a smooth-muscle layer and/or elastic lamina (ref), and thus may be partially responsible of the abnormal drainage of the brain parenchyma in the DVA area.

Interestingly, the follow-up imaging after oedema resolution showed a relative decrease of both those parameters (rCBV and tMIP), while being still elevated comparatively to normal brain parenchyma (Figure 3). This suggests that the initial inflammatory episode was associated with an increase in the venous congestion in the DVA that may be accounted for the size and severity of the main frontal lobe demyelinating lesion.

Those perfusion imaging features correlate well with the aforementioned tendency of demyelinating lesions to le localized in the perivenular brain parenchyma and the severity of the main demyelinating lesion in our case. The histologic characteristics of the DVA, whose consequences appear macroscopically in perfusion imaging, are associated with the preferential localization of the disease area.

The susceptibility weighting imaging (SWI) sequence comparison between the acute and follow-up exam shows an increase of venous structures size (including the DVA) on the follow-up exam. It could be hypothesized that the acute phase of the disease was associated with a lower level of deoxyhaemoglobin in venous blood, explaining the weaker signal on SWI.

In the light of those findings, we could suggest that in some rare cases, as the one presented here, DVA can potentialize the gravity of inflammatory brain disease, in providing a very favourable environment for demyelination to occur. Due to the scarcity of such reports, there is however no definite proof of this hypothesis, and the association observed here could be incidental. More data are needed in order to determine correlation between DVAs and ADEM lesions.

Conclusion

Developmental venous anomaly's repercussion on the surrounding brain and venular network seem to provide a favourable environment for the inflammation and demyelination process associated with acute disseminated encephalomyelopathy. Our case illustrates this phenomenon in a patient bearing a massive frontal lobe inflammatory lesion centred around a giant DVA. Magnetic resonance perfusion imaging shows increased regional blood volume and temporal parameters in the DVA network in the acute phase, possibly explaining the local severity of the inflammatory lesions. Those perfusion parameters tend to decrease in the months after acute inflammatory process regression, suggesting a specific role of the DVA in the acute phase of the disease.

Author contributions

Louis Deprez: conceptualization, investigation, visualization, and writing – original draft. Emilie Lommers: conceptualization and writing – review and editing.

Declaration of conflicting interests

The authors declare 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.

Ethical statement

Informed consent

The requirement for informed consent to this case report has been waived by the local Universitary Ethics Committee.

ORCID iD

Louis Deprez https://orcid.org/0000-0002-8162-8793

Data Availability Statement

The datasets generated during and/or analysed during the current study are available from the corresponding author on reasonable request.

References

1. Rammos SK, Maina R and Lanzino G. Developmental venous anomalies: current concepts and implications for management. *Neurosurgery* 2009; 65(1): 20–29.
2. San Millán Ruíz D and Gailloud P. Cerebral developmental venous anomalies. *Childs Nerv Syst* 2010; 26(10): 1395–1406.
3. Magyar M, Gattringer T, Enzinger C, et al. Incidence of developmental venous anomalies in patients with multiple sclerosis: a 3 Tesla MRI study. *Front Neurol* 2022; 13: 824347.
4. Amuluru K, Al-Mufti F, Hannaford S, et al. Symptomatic infratentorial thrombosed developmental venous anomaly: case report and review of the literature. *Interv Neurol* 2016; 4(3–4): 130–137.
5. Nagatani K, Osada H, Takeuchi S, et al. Surgical resection of developmental venous anomaly causing massive intracerebral haemorrhage: a case report. *Br J Neurosurg* 2014; 28(1): 116–118.
6. Pereira VM, Geibprasert S, Krings T, et al. Pathomechanisms of symptomatic developmental venous anomalies. *Stroke* 2008; 39(12): 3201–3215.
7. Topper R, Jurgens E, Reul J, et al. Clinical significance of intracranial developmental venous anomalies. *J Neurol Neurosurg Psychiatry* 1999; 67(2): 234–238.

8. Santucci GM, Leach JL, Ying J, et al. Brain parenchymal signal abnormalities associated with developmental venous anomalies: detailed MR imaging assessment. *Am J Neuroradiol* 2008; 29(7): 1317–1323.
9. Larvie M, Timerman D and Thum JA. Brain metabolic abnormalities associated with developmental venous anomalies. *Am J Neuroradiol* 2015; 36(3): 475–480.
10. Alexander JS, Prouty L, Tsunoda I, et al. Venous endothelial injury in central nervous system diseases. *BMC Med* 2013; 11(1): 219.
11. San Millán Ruíz D, Delavelle J, Yilmaz H, et al. Parenchymal abnormalities associated with developmental venous anomalies. *Neuroradiology* 2007; 49(12): 987–995.
12. Haacke EM, Beggs CB and Habib C. The role of venous abnormalities in neurological disease. *Rev Recent Clin Trials* 2012; 7(2): 100–116.
13. Tallantyre EC, Brookes MJ, Dixon JE, et al. Demonstrating the perivascular distribution of MS lesions in vivo with 7-Tesla MRI. *Neurology* 2008; 70(22): 2076–2078.
14. Zhang Y and Metz LM. Dominant perivenular enhancement of tumefactive demyelinating lesions in multiple sclerosis. *Neurology* 2010; 75(15): 1396.
15. Maggi P, Absinta M, Grammatico M, et al. Central vein sign differentiates multiple sclerosis from central nervous system inflammatory vasculopathies. *Ann Neurol* 2018; 83(2): 283–294.
16. Sati P, Oh J, Constable RT, et al. The central vein sign and its clinical evaluation for the diagnosis of multiple sclerosis: a consensus statement from the North American imaging in multiple sclerosis cooperative. *Nat Rev Neurol* 2016; 12(12): 714–722.
17. Rogers DM, Peckham ME, Shah LM, et al. Association of developmental venous anomalies with demyelinating lesions in patients with multiple sclerosis. *Am J Neuroradiol* 2018; 39(1): 97–101.
18. Ma M, Chen JY, Plowey ED, et al. Tumefactive demyelination associated with developmental venous anomaly: report of two cases. *Clin Imag* 2017; 43: 194–198.
19. Steiner I and Kennedy PGE. Acute disseminated encephalomyelitis: current knowledge and open questions. *J Neurovirol* 2015; 21(5): 473–479.
20. Singh S, Alexander M and Korah IP. Acute disseminated encephalomyelitis: MR imaging features. *Am J Roentgenol* 1999; 173(4): 1101–1107.
21. Algahtani H, Shirah B and Alassiri A. Tumefactive demyelinating lesions: a comprehensive review. *Mult Scler Relat Disord* 2017; 14: 72–79.
22. Li K, Li M, Wen L, et al. Clinical presentation and outcomes of acute disseminated encephalomyelitis in adults worldwide: systematic review and meta-analysis. *Front Immunol* 2022; 13: 870867.
23. Höllinger P, Sturzenegger M, Mathis J, et al. Acute disseminated encephalomyelitis in adults: a reappraisal of clinical, CSF, EEG, and MRI findings. *J Neurol* 2002; 249(3): 320–329.
24. Nakayama M, Naganawa S, Ouyang M, et al. A review of clinical and imaging findings in tumefactive demyelination. *Am J Roentgenol* 2021; 217(1): 186–197.
25. Sechi E, Krecke KN, Messina SA, et al. Comparison of MRI lesion evolution in different central nervous system demyelinating disorders. *Neurology* 2021; 97(11): e1097–e1109.
26. Kaneko K, Sato DK, Nakashima I, et al. Myelin injury without astrocytopathy in neuroinflammatory disorders with MOG antibodies. *J Neurol Neurosurg Psychiatry* 2016; 87(11): 1257–1259.
27. Salama S, Khan M, Pardo S, et al. MOG antibody–associated encephalomyelitis/encephalitis. *Mult Scler* 2019; 25(11): 1427–1433.
28. Santoro JD and Chitnis T. Diagnostic considerations in acute disseminated encephalomyelitis and the interface with MOG antibody. *Neuropediatrics* 2019; 50(05): 273–279.
29. Körtvélyessy P, Breu M, Pawlitzki M, et al. ADEM-like presentation, anti-MOG antibodies, and MS pathology: TWO case reports. *Neurol Neuroimmunol Neuroinflamm* 2017; 4(3): e335.
30. Höftberger R, Guo Y, Flanagan EP, et al. The pathology of central nervous system inflammatory demyelinating disease accompanying myelin oligodendrocyte glycoprotein autoantibody. *Acta Neuropathol* 2020; 139(5): 875–892.
31. Lassmann H, Brunner C, Bradl M, et al. Experimental allergic encephalomyelitis: the balance between encephalitogenic T lymphocytes and demyelinating antibodies determines size and structure of demyelinated lesions. *Acta Neuropathol* 1988; 75(6): 566–576.
32. Jung HN, Kim ST, Cha J, et al. Diffusion and perfusion MRI findings of the signal-intensity abnormalities of brain associated with developmental venous anomaly. *Am J Neuroradiol* 2014; 35(8): 1539–1542.
33. Jung G, Schröder R, Lanfermann H, et al. Evidence of acute demyelination around a developmental venous anomaly: magnetic resonance imaging findings. *Invest Radiol* 1997; 32(9): 575–577.
34. Iv M, Fischbein NJ and Zaharchuk G. Association of developmental venous anomalies with perfusion abnormalities on arterial spin labeling and bolus perfusion-weighted imaging. *J Neuroimaging* 2015; 25(2): 243–250.

Appendix

List of abbreviations

ADEM	Acute disseminated encephalomyelitis
DVA	Developmental venous anomaly
MS	Multiple sclerosis
CT	Computed tomography
MRI	Magnetic resonance imaging
FLAIR	Fluid attenuated inversion recovery
DSC	Dynamic susceptibility contrast
SWI	Susceptibility weighting imaging
ASL	Arterial spin labelling.