Case 15617

Eurorad

Tumefactive demyelinating lesion

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Section: Neuroradiology
Area of Interest: Neuroradiology brain
Procedure: Imaging sequences
Imaging Technique: MR
Imaging Technique: MR-Spectroscopy
Special Focus: Inflammation Case Type: Clinical Cases
Authors: Klemen Mihel?i?, Jernej Avsenik
Patient: 45 years, male

Clinical History:

45-year-old man with 7 year history of dysarthria presented in 2018 with clinical worsening. Past MRI showed multiple brain lesions, biopsy showed no tumour cells but was otherwise inconclusive. LP was negative for oligoclonal bands. The patient was diagnosed with status post ADEM. There was moderate improvement after corticosteroids.

Imaging Findings:

MRI from 2015 showed small T2/FLAIR hyperintense lesion in the left frontal white matter (Fig. 1). No post-contrast enhancement was seen. Small area of atrophy with surrounding gliosis after biopsy was present on the right.
MRI in 2018 (Fig. 2) showed enlargement of the previously described lesion in the left frontal lobe and newly developed round lesion in the right upper frontal gyrus. Both lesions were clearly hypointense on T1, hyperintense on T2 / FLAIR imaging and were surrounded by marked focal oedema. Intense post-contrast open ring enhancement was present as well, best seen on coronal and sagittal reformations (Fig. 3).
MR spectroscopy in the right frontal lobe lesion showed elevated choline, low N-acetyl aspartate and pathological lactate-lipid peak (Fig. 4).
DSC MRI showed low rCBV in the centre of the lesions (Fig. 5).
DWI showed low diffusion peripherally and high ADC values centrally as well as in the perilesional oedema (Fig. 6).

Discussion:

Background
Tumefactive demyelinating lesions (TDL) are a rare disease of the central nervous system (CNS). TDLs are most commonly observed in the context of multiple sclerosis (MS). Other primary demyelinating diseases such as acute disseminated encephalomyelitis (ADEM) and acute haemorrhagic leukoencephalitis can also manifest as TDL. It remains unclear whether TDLs represent a variant of MS or a demyelinating disease on its own [1, 2].
The prevalence of TDL is estimated to be 1–3/1000 cases of MS with an annual incidence of 0.3 per 100000. They may occur at any age but are most prevalent between 20-40 years of age [1].

Clinical Perspective
Clinical presentation is often atypical for a demyelinating disease and usually polysymptomatic. Motor, sensory, cognitive and cerebellar symptoms are predominant and they may develop over days or weeks. TDLs are often supratentorial with the most common symptoms being memory dysfunction (17%), aphasia (17%), apraxia (4%), Gerstmann syndrome (4%), seizures, impaired consciousness and visual field deficits [2, 3].

Imaging Perspective
Diagnosis of TDL is not always straightforward since the clinical and radiological findings can mimic other lesions

such as brain tumour. MRI is the gold standard imaging modality for TDL diagnosis, including gadolinium enhanced images. TDLs are mainly localised in the subcortical hemispheric white matter but may also be found in the corpus callosum. Most are focal and supratentorial with a tendency towards the frontal and parietal lobes [3-5]. MRI features consist of: size of more than 2 cm, incomplete ring enhancement (as compared to the complete ring of enhancement seen in abscesses and tumours) and large number of lesions [2-5]. High apparent diffusion coefficient (ADC) values are typical, however, low diffusion has been observed in active demyelinating lesions.
Perfusion images may aid in the differentiation of TDL from brain tumours by measuring the mean relative cerebral blood volume within the TDL which will be significantly less than in the brain tumours. However, perfusion imaging may not be conclusive in the cases of glioblastoma multiforme (GBM) and primary CNS lymphoma for example. To differentiate between primary CNS lymphoma and TDL, magnetic resonance spectroscopy (MRS) and CSF analysis may be useful. MRS of TDLs could show an increased choline and/or lactate peaks although these are not specific and may be observed in neoplasms as well [6].
Differential Diagnosis List: Tumefactive demyelinating lesion, CNS neoplasm, Brain metastasis, Brain abscess, Granulomatous disease, Vasculitis, ADEM

Final Diagnosis: Tumefactive demyelinating lesion

References:

Algahtani H, Shirah B, Alassiri A (2017) Tumefactive demyelinating lesions: A comprehensive review. Multiple Sclerosis and Related Disorders 14:72-9 (PMID: 28619436)
Frederick MC, Cameron MH (2016) Tumefactive Demyelinating Lesions in Multiple Sclerosis and Associated Disorders. Current Neurology and Neuroscience Reports 16(3):26 (PMID: 26847090)
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Cha S, Pierce S, Knopp EA, Johnson G, Yang C, Ton A, et al. (2001) Dynamic Contrast-enhanced T2*-weighted MR Imaging of Tumefactive Demyelinating Lesions. American Journal of Neuroradiology 22(6):1109 (PMID: 11415906)
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Sarbu N, Shih RY, Jones RV, Horkayne-Szakaly I, Oleaga L, Smirniotopoulos JG (2016) White Matter Diseases with Radiologic-Pathologic Correlation. RadioGraphics 36(5):1426-47 (PMID: 27618323)

Figure 1

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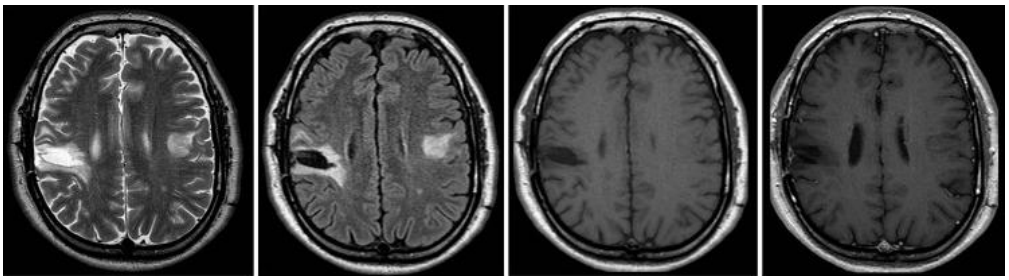

Description: MRI from 2015 (left to right: Axial T2, FLAIR, T1, T1 C+) shows small T2/FLAIR hyperintense lesion with no post-contrast enhancement in the left frontal lobe white matter. Postoperative changes are seen on the right. **Origin:** Avsenik J, Clinical Institute of Radiology, University Medical Centre, Ljubljana, Slovenia.

Figure 2

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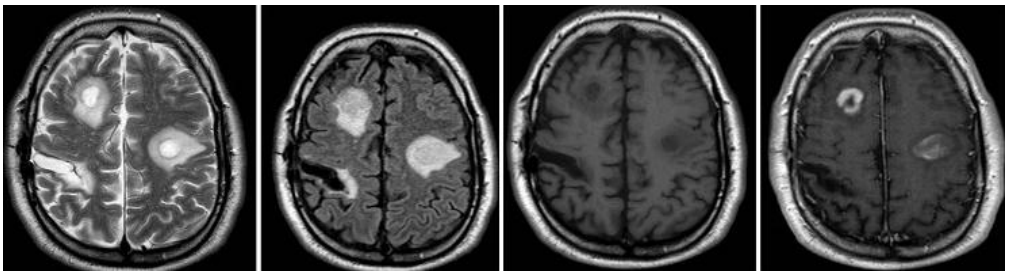

Description: MRI in 2018 showed enlargement of the left frontal lobe and newly developed lesion in the right upper frontal gyrus. Both lesions are hypointense on T1, hyperintense on T2/FLAIR and show marked post-contrast enhancement. **Origin:** Avsenik J, Clinical Institute of Radiology, University Medical Centre, Ljubljana, Slovenia.

Figure 3

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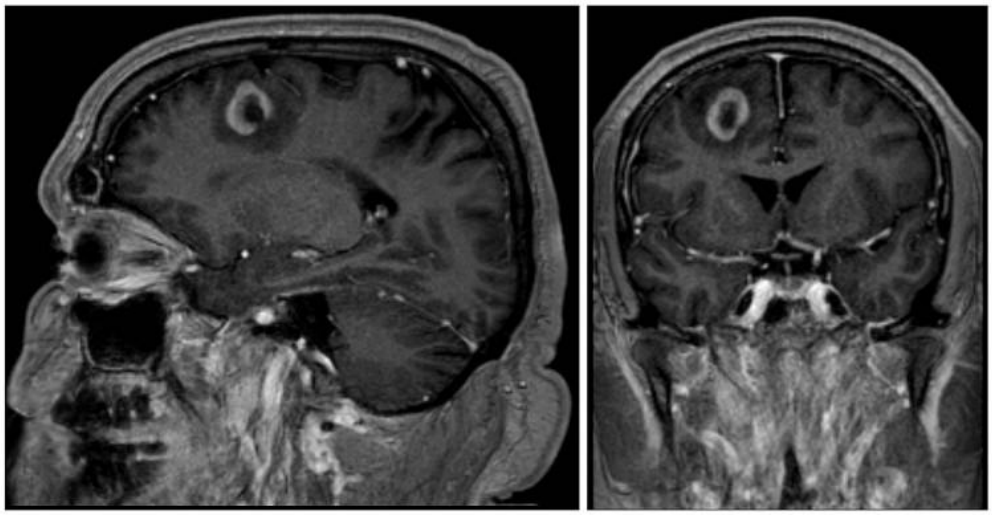

Description: Sagittal and coronal reformations show marked post-contrast enhancement in the characteristic open-ring pattern. **Origin:** Avsenik J, Clinical Institute of Radiology, University Medical Centre, Ljubljana, Slovenia.

Figure 4

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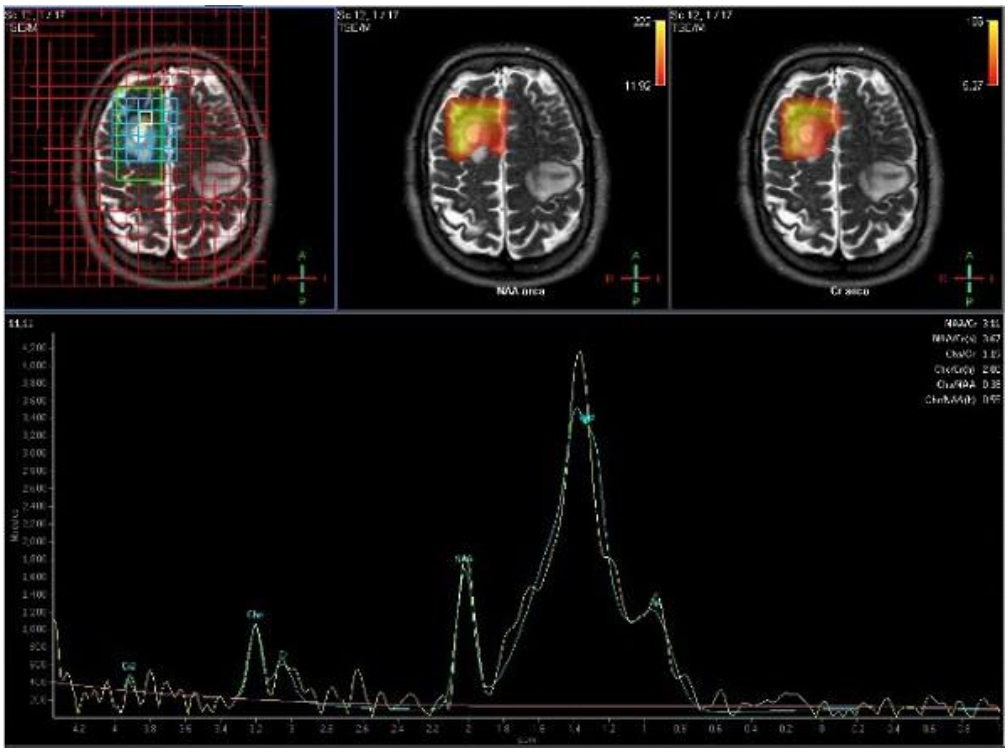

Description: MRS shows elevated choline, low N-acetyl aspartate (NAA) and pathologic lactate-lipid peak inside the right frontal lobe lesion. **Origin:** Avsenik J, Clinical Institute of Radiology, University Medical Centre, Ljubljana, Slovenia.

Figure 5

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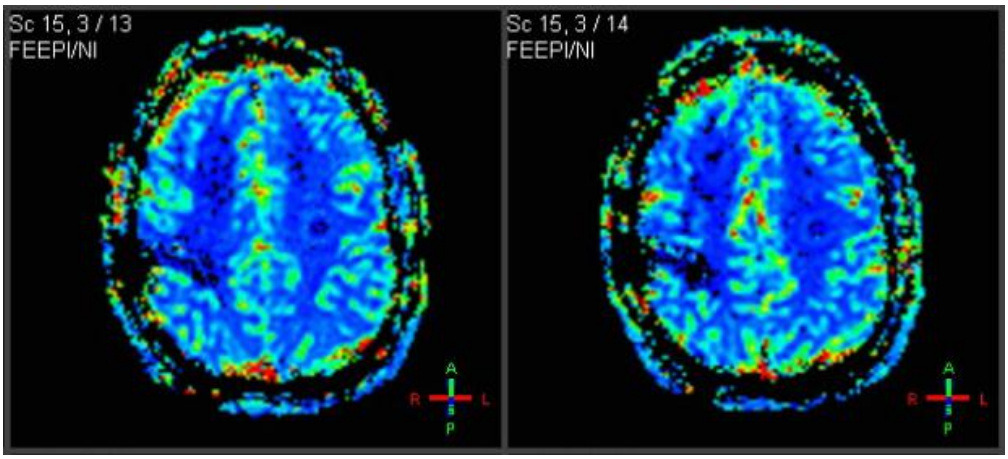

Description: Low rCBV values are seen in the center of the left frontal lobe lesion, as well as in the atrophic area after surgical biopsy on the right. **Origin:** Avsenik J, Clinical Institute of Radiology, University Medical Centre, Ljubljana, Slovenia.

Figure 6

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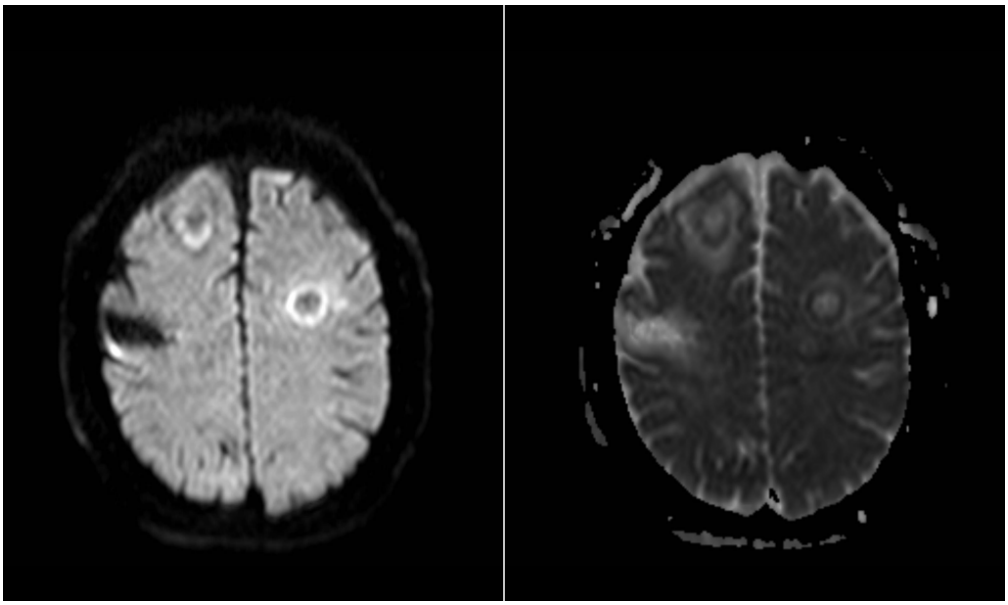

Description: DWI (left) and ADC map (right) show low diffusion on the periphery and facilitated diffusion in the center of the lesions as well as in the perilesional oedema. **Origin:** Avsenik J, Clinical Institute of Radiology, University Medical Centre, Ljubljana, Slovenia.