An interesting case of tumefactive demyelination
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Clinical History:
A 44-year-old gentleman, a known case of pulmonary sarcoidosis since 10 years, came to the ER department following an episode of seizure along with sudden onset weakness on the right side of the face and right lower limb. Clinically, vasculitis-induced stroke or a glioma were suspected.

Imaging Findings:
CECT showed an ill-defined hypoattenuating lesion in left corona radiata and centrum semiovale (Fig. 1). This was hyperintense on T2/FLAIR and hypointense on T1WI (Fig. 2); showed peripheral DWI hyperintensity and signal drop on ADC (Fig. 3) with subtle open ring type of peripheral enhancement (Fig. 4). Spectroscopy showed elevated choline, creatine, reduced NAA and a lactate doublet peak on long TE; elevation of glutamate-glutamine peaks on short TE (Fig. 5). Arterial Spin Labelling maps displayed peripheral arc of hyperperfusion (Fig. 6). Oligoclonal bands were not present on CSF examination and FDG PET-CT revealed reduced uptake in the lesion (Fig. 7). Since the diagnosis was indeterminate, a surgical biopsy was done and histopathology showed tumefactive demyelination with loss of myelin, preserved axons, presence of macrophages, lymphocytosis and astrocytosis (Fig. 8). Post-treatment MRI after 6 months demonstrated resolution of both diffusion restriction and enhancement. (Fig. 9)

Discussion:
Tumefactive demyelinating lesion (TDL), is a rare type of unifocal aggressive demyelination, usually larger than 2cm, commonly located in the frontal and parietal lobes and less commonly in the brain stem, temporal lobes, cerebellum and deep grey matter [1, 2]. It can mimic many malignant (lymphoma, glioma) and benign (abscess, acute disseminated encephalomyelitis, granulomas or stroke) lesions [2].

In our case, the left frontoparietal lesion was hypodense on CECT and the diagnostic challenge was to differentiate
it from a tumour or sarcoid-induced vasculitic infarct. The lesion displayed ill-defined T2/FLAIR white matter hyperintensity and T1WI hypointensity with no definite mass effect. It showed subtle open ring type of enhancement with discontinuity towards cortex and discontinuous peripheral restriction of diffusion which is common in TDL [3, 4]. Additional documented imaging features of TDL are necrosis, cystic changes, mass effect and variable amount of grey matter involvement [3]. They can also show uniform enhancement, ring-like or central venular, variable/complex enhancement [1, 2].

Studies have shown that mean relative cerebral blood volume and FDG uptakes in TDL is less than in tumours [5, 6]. Variable FDG uptake can be seen depending on the activity of lesion [5]. In our case, FDG uptake was low. The arterial spin labelling map showed hyperperfusion at the inner margins of the lesion, suggesting active disease.

Long TE MR spectroscopy showed increased choline; reduced NAA; elevated lactate peak. Short TE spectroscopy showed elevated glutamine-glutamate peak which is specific to TDL [7, 8]. There may be no significant differences in Cho/Cr ratios in TDL and gliomas. However, the mean NAA/Cr ratio may show marked reduction in central parts of gliomas compared to TDL and this can be used as a differentiating feature [7, 8]. In our case NAA/Cr ratio was mildly reduced (0.86). In the absence of oligoclonal bands in CSF and larger lesional size, a definite diagnosis could not be made and diagnosis of TDL was confirmed on surgical biopsy and systemic steroids were started [2]. Corticosteroid therapy usually improves the clinical symptoms and may reduce the size of the lesion as was in our case. TDL may progress to multiple sclerosis, remain isolated or show recurrence [1, 2]

It is crucial to differentiate TDL from its mimics and avoid an unnecessary brain biopsy. In indeterminate cases, biopsy should preferably be targeted from the wall of the lesion, contrary to tumours where the central zone of the lesion is preferred [9, 10].

**Differential Diagnosis List:** Tumefactive demyelination., Glioma, Stroke, ADEM, Lymphoma, Metastasis, Abscess, Neurosarcoïdosis

**Final Diagnosis:** Tumefactive demyelination.

**References:**


Description: Diffusion-weighted image showing altered arc-like hyperintensity at the periphery of the lesion, representing acute aetiology. Origin: Department of radiology Fortis Hospital, Noida U.P. India
Description: The diffusion hyperintensity shows signal drop on ADC maps suggestive of restricted diffusion at the periphery of the lesion (arrows); The central region of the lesion shows normal diffusion.

Origin: Department of radiology, Fortis Hospital, Noida U.P. India
Figure 2

Description: Axial T2WI image showing an ill-defined hyperintensity in left centrum semiovale. No definite mass effect is noted. Origin: Department of Radiology Fortis Hospital Noida, U.P, India
Description: Axial T1WI image showing an ill-defined hypointensity in left centrum semiovale. No definite mass effect is noted. Origin: Department of Radiology Fortis Hospital, Noida, U.P., India
Description: Axial FLAIR image showing an ill-defined hyperintensity in left centrum semiovale. Origin: Department of Radiology Fortis Hospital, Noida, U.P, India
Description: Coronal T2WI image showing an ill-defined hyperintensity in left centrum semiovale, periventricular and subcortical white matter. No definite mass effect is noted. Origin: Department of Radiology Fortis Hospital, Noida, U.P., India
Figure 3

Description: Post-contrast coronal MRI image showing a faint rim of discontinuous enhancement at the medial aspect of the lesion with open ring towards cortical grey matter. **Origin:** Department of Radiology, Fortis Hospital, Noida, U.P
Description: Post-contrast axial MRI image showing a faint rim of discontinuous enhancement at the medial aspect of the lesion with open ring towards cortical grey matter. Origin: Department of Radiology, Fortis Hospital, Noida, U.P.
Description: MRI spectroscopy indicating the region of interest. Origin: Department of radiology. Fortis Hospital, Noida, U.P. India
Description: MRI spectroscopy graph at 144 TE showing elevated choline and creatine levels with reduction in NAA and inverted lactate peak. Origin: Department of radiology. Fortis Hospital, Noida, U.P. India
**Description:** MRI Spectroscopy graph at 35 TE showing elevation of glutamine/glutamate peak.

**Origin:** Department of radiology, Fortis Hospital, Noida, U.P. India
Description: CECT showing an ill-defined hypoattenuating lesion in left centrum semiovale. Origin: Department of radiology, Fortis Hospital, Noida, U.P. India
Description: Photomicrograph of the biopsied specimen of the lesion showing prominent macrophages and a bizarre gemistocyte (large blue-stained cell) which may lead to incorrect diagnosis of glial neoplasm. Origin: Department of radiology, Fortis Hospital, Noida, U.P., India
Description: Photomicrograph of the biopsied specimen shows prominent perivascular lymphocyte and histiocytic proliferation. Origin: Department of radiology, Fortis Hospital, Noida, U.P., India
**Description:** Photomicrograph of the biopsied specimen after myelin stain showing normal myelinated brain parenchyma on right side (in blue), compared with area of the demyelination (pink on left side).

**Origin:** Department of radiology, Fortis Hospital, Noida, U.P., India
Description: Diffusion-weighted image shows no definite rim of restriction, which was appreciated in the initial scans. Origin: Department of Radiology, Fortis hospital, Noida, U.P
Description: Contrast-enhanced T1WI axial MRI image showing no enhancement. Surgical changes are also seen in left fronto-parietal calvarium (related to previous biopsy). Origin: Department of Radiology, Fortis hospital, Noida, U.P.
**Description:** Axial T2/ FLAIR image shows reduction in the size and extent of the white matter signal alteration. **Origin:** Department of Radiology, Fortis hospital, Noida, U.P
Description: T1WI showing hypointense lesion in left centrum semiovale region.
(for reference) Origin: Department of Radiology, Fortis hospital Noida, U.P., India
Description: A colored arterial spin labelling map demonstrating rim of hyperperfusion giving a characteristic open ring appearance (yellow; ROI-1) in the active zone of demyelination in left centrum semiovale region. Origin: Department of Radiology, Fortis Hospital Noida, U.P, India
Figure 9

Description: FDG-PET CT image demonstrating photopaenia (reduced uptake of FDG) in the lesion. SUV max 2.5 Origin: Department of radiology, Fortis Hospital, Noida, U.P