

## Case report

### Tumefactive demyelination-A Rare Presentation of Multiple Sclerosis

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#### Abstract

**Background:** Multiple Sclerosis is an inflammatory demyelinating disease having varied manifestations in terms of clinical features and radiological features. Rarely it may present as a large demyelinating lesion with accompanying edema and mass effect, thereby simulating an intracranial tumor, known as Tumefactive Demyelination. Symptoms are usually related to the pressure of a focal mass lesion. When it manifests in a patient without pre-existing MS, it poses a diagnostic challenge as it may mimic a neoplasm, infarct or abscess. Thus, it is essential to recognize this rare clinical entity for proper patient management. **Case Presentation:** We report here a case of 40 years old female who presented with right sided hemiparesis. The initial brain imaging (CT) suggested left parietal lobe lesion suggestive of glioma. But MRI of the brain showed a cortical based lesion in left parietal lobe in parasagittal location which appeared hyperintense on both T2WI and FLAIR sequences. Further evaluation by MR spectroscopy suggested the lesion to be tumefactive demyelination. She was administered IV Methylprednisolone (1gm daily for 5 days) and responded well to treatment. The characteristic magnetic resonance findings of the patient, its acute onset, and its clinical improvement after corticosteroid therapy finally set the diagnosis of Tumefactive Demyelination. **Conclusion:** Tumefactive demyelinating lesions pose a unique diagnostic challenge in defining differential diagnosis and management and should always be considered in young patients presenting with tumour like lesions on imaging.

**Keywords:** Tumefactive demyelination; multiple sclerosis; MR Spectroscopy.

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#### Background

Multiple Sclerosis is an inflammatory demyelinating disease having myriad manifestations in terms of onset, clinical features, course of disease and radiological features. Rarely it may present as a large demyelinating lesion (>2cm), with accompanying

edema and mass effect, thereby simulating an intracranial tumor. This is known as Tumefactive Demyelination. The symptoms are generally atypical for multiple sclerosis (MS) and usually related to the pressure exerted by focal mass lesion. When it manifests in a patient without pre-existing MS, it

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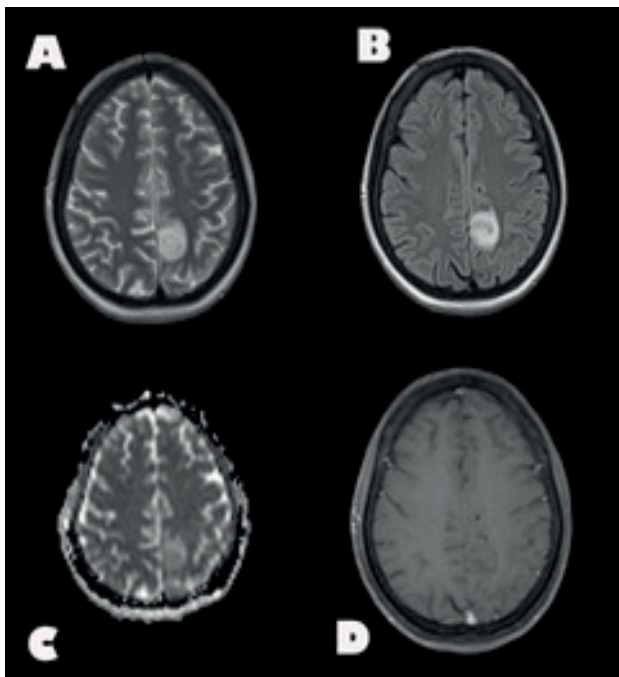
poses a diagnostic challenge both to the physician and the radiologist, as it may mimic a neoplasm, infarct or abscess. But it is essential to recognize this rare clinical entity for proper patient management and to avoid unnecessary medical therapy, harmful surgical resection, radiation therapy or drainage.

### Case presentation

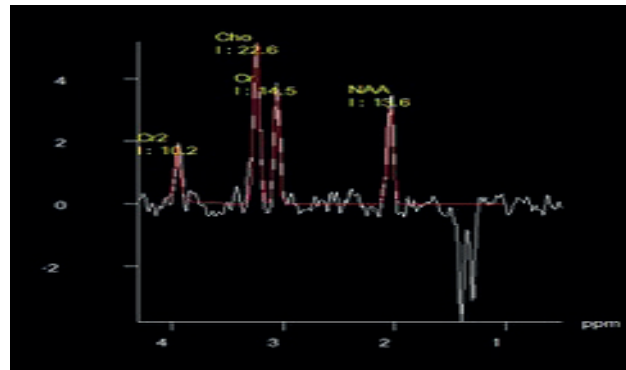
A 40 years old female, non-diabetic, non-hypertensive presented to our hospital with chief complaints of acute onset weakness of right side of body i.e., right leg and arm; slurring of speech and deviation of angle of mouth to left side. She was in usual state of her health in the night before she presented to our emergency department but woke up in morning with above mentioned neurological symptoms. She had no significant past medical history. There was no history of similar complains in the past and family history was unremarkable. The patient was a housewife and had no history of addiction to tobacco, alcohol, or smoking. On examination she was conscious and oriented, her pulse rate was 80/min regular, blood pressure 140/86 mmHg, respiratory rate 20/min and her peripheral oxygen saturation was 99% on room air. Her initial NIHSS (National Institute of Health Stroke Scale) score was 6. The

examination of other systems was normal. After the initial evaluation, an emergency non-contrast CT scan of head was done which revealed a small area of hypodensity located in left parietal lobe with provisional impression of acute infarct/glioma. She was not considered for thrombolytic therapy due to unknown time of symptom onset. She was planned for conservative management and admitted to CCW (Critical Care Ward). All the biochemical parameters were normal. Serology for HIV, HBV and HCV was negative. ANA (Anti-nuclear antibody) was negative, coagulation profile was normal and ESR was 12mm in first hour. Chest X-ray, USG abdomen and pelvis, Echocardiography (transthoracic), and bilateral carotid artery doppler study was normal. The patient was subjected to an MRI of the brain which showed a cortical based lesion in left parietal lobe in parasagittal location of about 2cmx3cm in size appearing hyperintense on both T2WI and FLAIR sequences (Fig-1).

There was no edema in the adjacent brain parenchyma. The lesion showed mild restricted diffusion; however no enhancement was noted on post contrast sequences. On MR spectroscopy there is elevation in the Choline levels with slight reduction in NAA with a prominent lactate doublet (Fig-2). These imaging findings were consistent with a diagnosis of tumefactive demyelination.



**Fig. 1:** A cortical based lesion in left parietal lobe in parasagittal location of about 2cmx3cm in size appearing hyperintense on both T2WI and FLAIR sequences



**Fig. 2:** elevation in the Choline levels with slight reduction in NAA with a prominent lactate doublet

During the initial period of her hospitalization, the patient deteriorated significantly. So on the basis of her radiological findings and clinical course; she was started on high dose intravenous steroid (Inj. Methylprednisolone 1gm/day) therapy. On the third day of treatment, the patient started improving and steroid was continued for 5 days. The patient responded to treatment and is in follow up.

## Discussion

Tumefactive demyelinating lesions (TDLs) of brain are common occurrences; nevertheless a major source of diagnostic dilemma for the physicians, radiologist and the pathologists. Clinical differential diagnosis includes demyelinating diseases, neoplasms, and infections such as abscesses. Such lesions with mass-like characteristics may be the presenting feature of multiple sclerosis (MS). The exact pathogenesis of Tumefactive demyelinating lesions is not clearly understood. These lesions are considered to be intermediate between the typical lesions of multiple sclerosis and acute disseminated encephalomyelitis<sup>1</sup>. These lesions may occur at any age groups but are most commonly observed in adult patients in the second and third decades of their life. Also, these are more prevalent in women similar to that of multiple sclerosis<sup>2</sup>.

Tumefactive demyelinating lesions are larger in size (>2cm) than those present in typical MS and this along with accompanying oedema results in clinical features which are generally atypical and do not conform to the diagnosis of multiple sclerosis. The common presentations are related to the presence of a focal mass lesion rather than demyelinating disorders, including headache and vomiting (features of increased ICP), local neurological deficits, hemiplegia, aphasia and seizures<sup>3</sup>.

For radiological analysis, MRI is the most sensitive imaging technique and method of choice for imaging demyelination lesions, tumefactive or not. Although the usual appearance of MS is multiple, small, demyelinating plaques, but it can simulate a mass lesion and hard to distinguish it from a brain tumor. TDLs present a diagnostic challenge, since they usually presents a subcortical mass in the hemisphere mimicking gliomas as a solitary lesion or metastasis as a few separate lesions. MRI characteristics, such as open-ring enhancement, peripheral restriction on diffusion-weighted imaging, or venular enhancement, may help in differentiating tumefactive MS lesions from neoplastic ones. The pathognomonic radiological features include large lesion showing little mass effect or edema, open ring like enhancement, central dilated veins within the lesions and reduced perfusion. Few non-specific findings related to the tumefactive demyelination include corpus callosum involvement and increased diffusion. On MR spectroscopy tumefactive demyelination may mimic primary glial tumor<sup>4,5</sup>.

Fluoro-deoxyglucose positron emission tomography (FDG-PET) scan may demonstrate hypermetabolism in the TD but not to the degree as seen in neoplastic lesion. It may be a useful adjunct to MRI or CT in evaluation of TD lesion<sup>6</sup>. Another useful adjunct in identifying TD is the demonstration of unmatched OCBs in cerebrospinal fluid (CSF), if there is no contraindication to lumbar puncture. In a study by Altintas A. et al,<sup>33</sup> patients of known MS who developed TD, 90% had positive unmatched oligoclonal bands (OCBs) in the CSF compared with 52% who presented with a tumefactive lesion as their first clinical event<sup>7</sup>.

In practice, biopsy should be restricted to patients who are not having established MS, with inconclusive or suspicious imaging including PET, negative OCBs and/or those in whom a diagnosis of MS would be unusual, like older or very young patients<sup>8</sup>. Histopathologically, the features of isolated TD lesion and typical multiple sclerosis are similar. It consists of areas of demyelination with hypercellularity and reactive astrocytes (may contain multiple nuclei (Creutzfeldt cells) closely intermingled with myelin-containing foamy macrophages. There is relative sparing of axons with perivascular and parenchymal lymphocytic infiltrates<sup>9</sup>.

Most tumefactive demyelinating lesions show an excellent response to corticosteroid therapy with significant decrease in size or disappearance of the lesions on follow-up imaging<sup>1,4</sup>. But there are no established guidelines for timing of further imaging. He J. et al observed that TD lesions may take more than 12 weeks to show significant resolution post corticosteroids and 2% will show persistent gadolinium enhancement after 6 months<sup>10</sup>. Hardy TA et al proposed that re-imaging should be done at 6-8 weeks (or earlier if patient is deteriorating) and further at 3 months unless new or recurrent symptoms develop. If any atypical feature is observed, patient should proceed to biopsy<sup>8</sup>.

TD is treated as an acute episode of MS with IVMP or plasma exchange. There have been no randomized controlled therapeutic trials but the largest follow up study, more than 80% patients had significant response to corticosteroids<sup>7</sup>. Till now, there is insufficient evidence to recommend MS disease modifying therapy (DMT) after an initial TD event in the absence of clinical or collateral radiological evidence of dissemination in space and time<sup>8</sup>.

In a study by Altintas A et al, out of 29 patients

whowere diagnosed with tumefactive lesions at the onset,19 eventually developed relapsing–remitting MS, while 10 remained as a clinically isolated syndrome (CIS) after median follow-up of 38.12 months<sup>7</sup>.

### Conclusion

Tumefactive demyelinating lesions poses a unique diagnostic challenge in defining differential diagnosis, management and long term outcome. It may be misdiagnosed both clinically as well as radiologically. This case report emphasise that tumefactive demyelination should be always considered in young patients presenting with tumour like lesions. Clinical and radiological follow up is very crucial in supporting the diagnosis. The clinical and radiological improvement without any long term treatment is typical of TDLs.

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- iv) DK-contributed significantly towards design of work, analysis and interpretation of data

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