



Neuroimaging features of tumefactive demyelinating lesions: A rare case report

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ABSTRACT

Tumefactive demyelinating lesion is a special type of neuro-inflammatory disease with unusual variants to be found in magnetic resonance imaging. It manifests as a solitary or multiple lesion greater than 2 cm in diameter and often clinically challenging because of its varieties of symptoms. We present a rare case of a 40-year-old male, presented with initial complaints of headache and unconsciousness. Yet, the physical examination findings showed normal results. Magnetic resonance imaging revealed a tumefactive demyelinating lesion suspected with brain neoplasm.

1. Introduction

Tumefactive demyelinating lesions (TDLs) are a special type of inflammatory disease of the central nervous system (CNS) that occur in patients with or without the presence of a definitive neurological disorders, most often found in multiple sclerosis (MS) [1,2]. On neuroimaging, it typically manifests as a tumor-like lesion or mass with a diameter greater than 2 cm, with or without the involvement of mass effect, edema, and ring enhancement after contrast medium administration [1]. Epidemiology studies show a rare prevalence of TDLs, with only 1–2 cases per 1000 cases of MS [3,4]. Decade of 2 and 3 years of life is the range that is commonly known to be affected by TDLs [3,5]. The establishment of diagnosis has become a challenge because of its nature of clinical and radiological manifestations, which are usually mimicking brain neoplasm [2]. Currently, magnetic resonance imaging (MRI) is the main modality used for the diagnosis of TDLs and also helpful to differentiate other demyelinating disorders [2,6]. In our study, we describe a case of brain TDLs in male patient presented with a new onset of impaired consciousness. The patient had unique features on MRI imaging.

2. Case presentation

A 40-year-old male presented to the emergency room with a first

episode of impaired consciousness for over 30 min. The patient was alert after the intravenous administration of normal saline. On history taking, patient complained of chronic recurrent headaches, but did not disrupt his quality of life. He also had no history of impaired consciousness before. The patient had neither trauma nor another past medical condition. Vital signs were stable. General physical examinations were within normal limits. Neurological examination revealed no cranial deficit, language disturbances, or motoric and sensory problems. Laboratory examination did not show abnormal findings. The patient was decided to undergo head MRI examination, and the result revealed two lesions in the right parietal lobe, precisely at the white matter area with an open ring enhancement on the grey matter side of the lesion, the largest size of the lesion was about $2.56 \times 2.59 \times 2.16$ cm (Fig. 1). This finding led to the differential diagnosis of TDLs and brain neoplasm. Subsequently, the patient was referred to the neurosurgery department and planned for further histopathology evaluation and management. The patient had given informed consent regarding the collection and publication of the data.

3. Discussion

Our study presents one of the rare cases that TDLs were found on the MRI as the first clinical and radiographical event without any pre-existing inflammatory demyelinating diseases of the CNS [5]. Its

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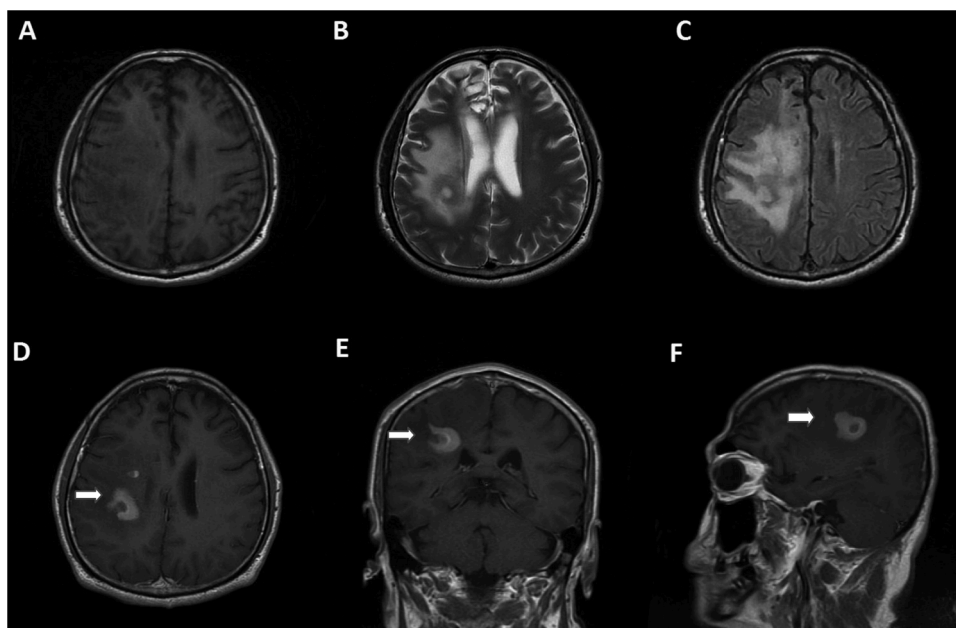


Fig. 1. (A) Axial T1-weighted image showing a hypointense lesion with edema, (B) Axial T2-weighted image, (C) Axial fluid-attenuated inversion recovery (FLAIR) shows a hyperintense lesion with edema, (D) Axial T1-weighted image post-contrast, (E) Coronal T1-weighted image post-contrast, and (F) Sagittal T1-weighted image post-contrast showing two lesions at white matter with an open ring enhancement towards the grey matter side of the lesion (arrow).

course is uncommon and often manifests variably that are resembling the brain neoplasms [2,3]; thus, it becomes a challenge for clinicians to be able to establish the diagnosis of TDs. Some studies report that TDs often occur at 20–30 years of life [3,5,8], although Kim et al. reported that the onset of symptoms occurs averagely at 42 years [9].

The patient was a 40-year-old man, which is at the typical onset age of TDs symptoms [9]. He had a history of chronic headaches, which can indeed be found in people with TDs, but this complaint is quite nonspecific and leads to many differential diagnoses. Impaired consciousness can also be the feature of TDs, but is rarely found as an initial attack episode. Ekmekci et al. had firstly demonstrated an atypical symptom and incidentally detected TDs during MRI, known as silent TDs [1]. This occurrence is uncommon, especially among patients with unknown history of inflammatory demyelinating diseases.

It has been stated in the literature that the TDs may alter the motoric, sensory, cognitive, and cerebellar function, which depends on the different sizes and location of the lesion [3,4]. The symptoms like headaches, slurred speech, limb weakness, and altered consciousness often occur due to the mass effect that will be pushing or displacing the surrounding tissue, leading to increased intracranial pressure and cerebral herniation [2–5,7].

Magnetic resonance imaging of the brain has a major role in establishing the diagnosis of TDs, including three plane scout, sagittal fast fluid-attenuated inversion recovery (FLAIR), axial spin-echo proton density/T2, axial fast FLAIR, and axial gadolinium-enhanced T1 [5,10]. Some conditions may be associated with TDs, including MS, Balo disease (Balo concentric sclerosis), Marburg's MS, acute disseminated encephalomyelitis, and Devic's disease [1,10–12]. Manifestation of TDs may also be mimicking glial tumors, CNS lymphoma, and brain abscess [1,9].

Certain imaging characteristics can be helpful in diagnosing TDs. The lesion may be single or multiple and appear simultaneously or sequentially [1]. Lucchinetti et al. reported that TDs mostly affect the white matter rather than grey matter [1,3–5]. It also associates with mass effect in about 45 % and edema in about 77 % of the patients [4,5]. The imaging findings may also include T1 hypointensity, T2 hyperintensity, contrast enhancement, and peripheral restriction around lesion based on diffusion-weighted images (DWI) [1,3,5].

The ring enhancement, whether with a complete or incomplete portion of the ring pointing towards the grey matter, is an important diagnostic clue to a tumefactive lesion [3]. This ring enhancement pattern represents the active inflammation process, whereas the non-enhancing central represent a more chronic phase in which the blood-brain barrier disruption has partially resolved [3]. Jeong et al. reported that the most common enhancement pattern found in TDs is the ring enhancement [8]. Kim et al. reported that either focal, complete, and incomplete ring enhancement were more frequent and significantly different in TDs patients compared to tumor patients [9]. Other studies revealed that the incomplete or open ring enhancement is highly indicative of TDs, with a sensitivity of 71.4 % and specificity of 98 % [3,4].

In addition to other modalities of neuroimaging, MR spectroscopic may be helpful as an invasive diagnosis of TDs with some classical metabolite abnormal elevation findings such as glutamate/glutamine ratio peak. Cianfoni et al. reported an elevation of N-acetyl aspartate (NAA), choline (Cho), and glutamate/glutamine ratio peak at 2.1–2.5 ppm in their four patients with TDs. They also concluded that the elevation of glutamate/glutamine ratio peak was marked; there is significant active demyelination disease and inflammatory processes of CNS [13]. However, the diagnosis of TDs is still challenging for both clinicians and radiologists. The gold standard modalities, such as brain biopsy and histologic examination of the lesion, are often needed as final diagnostic approaches.

4. Conclusion

Our report describes a rare case of TDs with atypical clinical manifestations and unique findings on MRI images, proposing the challenge of establishing the correct diagnosis of TDs for clinicians and radiologists. Magnetic resonance imaging is still the best modality tool for the initial detection of TDs and may be used to reduce the necessity of invasive diagnostic procedures for TDs.

Authors' contribution

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CRedit authorship contribution statement

Conceptualization, Methodology, Writing - original draft. **Sony Sutrisno**: Investigation, Resource, Funding acquisition. Investigation, Resource, Funding acquisition. **Krisnhaliani Wetarini**: Writing - review & editing, Visualization.

Declaration of Competing Interest

The authors declare no conflict of interest.

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