

Mini Review on Leptomeningeal Enhancement (LME) in Neuromyelitis Optica Spectrum Disorders (NMOSD)

Poh Kok Wei*, Ngan Cheng Huong and Wong Ji Yin

**Medical Department, Tuanku Ja'afar Hospital, Seremban, Ministry of Health Malaysia.*

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ABSTRACT

Leptomeningeal enhancement (LME) is an infrequent finding in Neuromyelitis Optica Spectrum Disorders (NMOSD). There is currently limited literature available on this topic. Nevertheless, it is present during an MRI may represent a significant radiological finding. This brief review summarized the current knowledge on LME and its role in NMOSD.

Keywords: Leptomeningeal enhancement, LME, Neuromyelitis Optica Spectrum Disorders, NMOSD

Abbreviations

AQP4: Aquaporin-4; IgG: Immunoglobulin G; LETM: Longitudinally extensive transverse myelitis; LME: Leptomeningeal enhancement; MRI: Magnetic resonance imaging; NMOSD: Neuromyelitis Optica Spectrum Disorders

LEPTOMENINGEAL ENHANCEMENT IN NMOSD

Neuromyelitis Optica Spectrum Disorders (NMOSD) is inflammatory disorders of the central nervous system characterized by episodes of immune-mediated demyelination and axonal damage predominantly affecting the optic nerves and spinal cord [1]. Current consensus on the diagnosis of NMOSD with Aquaporin-4 (AQP4)-IgG requires presence of at least one of the six core clinical characteristics of NMOSD which include optic neuritis, acute myelitis, area postrema syndrome, acute brainstem syndrome, symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions and symptomatic cerebral syndrome with NMOSD-typical brain lesions [2].

One of the most distinct features in NMOSD is the longitudinally extensive transverse myelitis (LETM), which was defined as a lesion that spans over 3 or more contiguous vertebral segments [3]. Leptomeningeal enhancement (LME), with its frequency unknown, has been reported only by limited literature [4-7]. However, its importance in diagnosing and managing NMOSD has yet to be confirmed.

FREQUENCY OF LME IN NMOSD

LME is not detected by the MRI in most cases of NMOSD. In a study by Asgari et al. [5], a retrospective case series of 11 AQP4-IgG-positive NMOSD patients; only 5 out of 11 had LME during initial clinical presentation of NMOSD. Similarly, as described by Long et al., where they identified only 12.8% of NMOSD to have LME [3]. Conversely,

LETM were found in a higher frequency, reported by Asgari et al. to be 83.3% [8]. LETM, although a distinctive feature in NMOSD, its characteristic may change depending on time of the MRI was done and the treatment given [8,9]. Also, LME is in spatial relationship with intra parenchymal contrast enhancement of the MRI brain and spine cord LETM [5]. The median interval between symptom onset and LME has been reported to be 12 days [5]. However, this number may not reflect the true timing as it was based on retrospective analysis of small sample size patients. Hence, it is possible that the presence of LME also depends on the timing of the MRI done and whether treatment was given.

LOCATION OF LME IN NMOSD

LME could occur anywhere from the cortex to the cauda equina. In the retrospective case series by Asgari et al. [5], LME was found in cerebral hemispheres, conus to cauda equina, multifocal segment along the spinal cord and along the thoracic segments. A reported case series by Tahara et al. [10], identified LME on cerebral cortex. The LME were found in occipital, parietal or posterior temporal region

Corresponding author: Poh Kok Wei, Tuanku Ja'afar Hospital, Seremban Jalan Rasah, Bukit Rasah, 70300 Seremban, Negeri Sembilan, Malaysia, Tel: +60123305811; E-mail: dr.teddypoh@gmail.com

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among these 3 cases. On the other hand, a literature review by Sun et al. [11], found that majority of LME occurred at frontal lobe [11]. Hence, there may not be a specific location of LME for NMOSD. Pertaining to the LME along the spinal cord, it has been described to involve from 5 to 17 vertebral segments length [5]. Interestingly, LME of the entire spinal cord has been reported as well [6]. Unless more data is available, there is currently no specific region of LME that is pathognomonic for NMOSD.

HOW LME OCCURRED IN NMOSD?

The reason for the leptomeningeal gadolinium enhancement during MRI was hypothesised to be a result of the disruption of the blood-brain barrier by the antibodies targeted towards AQP4 water channel [4,12]. These ventricular margins contain ependymal cells and astrocytes which are abundant in AQP4 water channel [13,14].

DOES LME ONLY OCCUR IN NMOSD?

It is important to know that LME has a wide range of causes including infection, tuberculosis, neoplasm, autoimmune and vasculitis [15,16]. One interesting note, a study by Long et al. [4], found that 12.8% of patients with NMOSD had LME around the brainstem compared to none in patients with multiple sclerosis. However, the sample size was small to make any definite conclusion. With the current limited literature available on LME, perhaps it is important to exclude the common causes of LME. On the other hand, the presence of LME should not dissuade a clinician from diagnosing NMOSD.

FUTURE RESEARCH

We believed that further studies are needed to explore the importance of LME in NMOSD. Specifically, looking into the temporal and spatial relationship with LETM, clinical presentation and its prognosis.

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