ORIGINAL RESEARCH PAPER

Neurology

A STUDY OF ETIOLOGY AND CLINICO-RADIOLOGIC PROFILE OF LONGITUDINALLY EXTENSIVE TRANSVERSE MYELITIS IN A TERTIARY CARE CENTRE IN SOUTH INDIA

KEY WORDS: LETM, NMO

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ABSTRACT

Longitudinally extensive transverse myelitis [LETM] is defined as contiguous inflammatory lesions of spinal cord extending to three or more vertebral segments. The etiology being variable, Neuromyelitis optica (NMO) is the most common cause of LETM. The aim of this study is to study etiological, clinical and radiological profile and treatment outcomes in patients of LETM. Early diagnosis and management plays a major role in prognostication and improving longterm outcome.

INTRODUCTION

Longitudinally extensive transverse myelitis (LETM) also known as Longitudinally extensive spinal cord lesions (LESCL), represent extensive involvement of the spinal cord, with abnormal T2 signal traversing at least three vertebral body segments in length. 1 The clinical course is characterised by single or multiple attacks of paraparesis or quadriparesis, sensory deficits, bowel and bladder disturbances. In severe cases can lead to respiratory failure. 2 Neuromyelitis optica spectrum disorder (NMOSD) is the most common cause of LETM but it can also occur due to various etiological agents. 3

This study highlights the different etiology, clinico-radiological spectrum and treatment outcomes of LETM, and also emphasises the importance of early diagnosis and prompt initiation of therapeutic measures in the prevention of relapse and recurrences with improved long term outcome.

AIMS AND OBJECTIVES

To study etiology, clinical spectrum, radiological profile and treatment outcomes in patients of LETM.

MATERIALS AND METHODS

Study Design:
Hospital based cross sectional prospective study

Place of Research:
Department of Neurology, Madurai medical college, Madurai, Tamilnadu, India.

Study Period:
March 2021 to February 2022 [1 year]

Sample size:
Fifty [50]

Inclusion Criteria:
1. Age 5yrs to 80yrs.
2. Patients who presented with acute onset paraplegia/ paresis or quadriplegia/paresis with MRI spine showing LETM.

Exclusion Criteria:
1. Age < 5yrs and > 80yrs
2. Patients who presented with acute onset paraplegia/ paresis or quadriplegia/paresis with MRI spine showing Short segment Transverse Myelitis.
3. Patients with abnormal metabolic parameters.

Methodology:
After obtaining clearance from institutional ethical committee and taking consent from the subjects, a detailed analysis of their demographic data, clinical features, general physical and neurologic features has been done.

Those who met the inclusion criteria are subjected to detailed evaluation which included metabolic and haematologic parameters, neuroimaging, CSF studies, serum aquaporin 4 (AQP4), MOG antibodies, etiological evaluation including infective, inflammatory and connective tissue disorders workup.

All patients have been followed up for one year for prognosis.

RESULTS

1. The mean age in our series was 35.05 ±15.69 years.

2. Out of fifty [50] patients, thirty six [36] patients are female and fourteen [14] are males with a female predominance in our series with a female to male ratio of 2.6:1.

3. Thoracic and cervical spinal cord segments are the most commonly affected i.e in thirty eight [38] patients. [76%]

4. Optic nerve involvement is seen in twenty three [23] patients (46%).

5. The tendency to involve three to five segments is more common {seen in twenty eight [28] patients}. [56%]

6. Serum AQP4 antibody test is found positive in fifteen [15] patients (30%).

7. A total of twenty five [25] patients (50%) were clinically diagnosed as NMOSD according to Wingerchuks criteria 4, of these fifteen [15] are AQP4 positive, seven [7] are MOG antibody positive, three [3] cases are double seronegative NMOSD.

8. Of the other twenty five [25] cases twelve [12] patients are postinfectious myelitis(three [3] cases had tuberculous etiology, three [3] are secondary to HIV, six [6] with preceding viral prodromal febrile illness), three [3] cases are LETM secondary to connective tissue disorders, two [2] with underlying malignancies, three [3] with negative results on standard evaluation, three [3] patients expired and two [2] cases are lost for follow up, etiology for these cases is not known.

9. The patients with anti AQP4 antibody associated NMOSD and extensive involvement of cord with gray matter necrosis showed residual deficits and poor recovery on follow up assessment at one year.

10. Relapses are more common with AQP4 antibodies positive NMOSD.

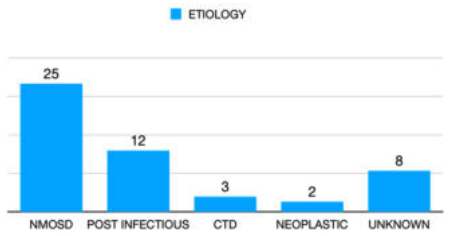


Chart 1 showing etiology of LETM in our study

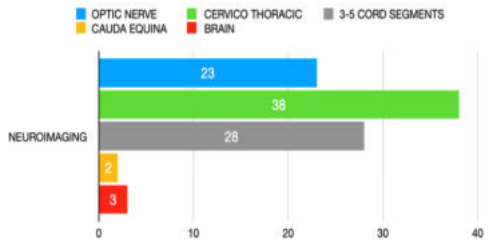


Chart 2 showing Radiological profile of LETM in our study

DISCUSSION

Longitudinally extensive transverse myelitis is a presentation of Acute Transverse Myelitis in all age groups. In our study mean age group is 35.05 ±15.69 years with female preponderance concordant with other studies. [5,6] AQP4 antibody positivity seen in adult age groups and MOG antibody in paediatric age groups.

The clinical profile depends on the type either partial or complete myelitis, the area of the cord involved whether cervical, cervicothoracic, dorsal, dorsolumbar or the entire length. 7 In our series cervicothoracic cord is most commonly involved.Those patients under MOGAD group showed involvement of caudal equina.

Longitudinal myelitis has multiple etiologies, infection and parainfectious causes have to be kept in mind to differentiate it from NMO as both have a different course of treatment and prognosis.

Central cord involvement and > 50% cross-section area involvement can be seen both in NMO and the infectious group, but cervico-dorsal location is more common in NMO.

Neuroimaging noted that a simultaneous bilateral posterior involvement of the optic nerve and chiasm showing nonspecific sheet thickening and hyperintensity with contrast enhancement is more classical of NMOSD. [8]

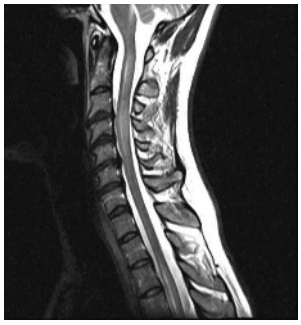
In our case study optic nerve involvement is associated with both AQP4 associated NMO and MOGAD; relapses and residual visual deficits are more common with AQP4 related NMOSD.

In our study those patients with extensive involvement of spinal cord and brain involvement showed rapid worsening of symptoms with impending respiratory failure requiring more aggressive management.

Those who recovered the acute event later presented with post myelitis sequelae and tonic spams on follow up. The patients under antibody positive NMOSD required long term immunosuppression.

Early diagnosis and management with steroids and immunosuppressants play major role in improving longterm outcome and preventing relapses. [9.10]

Figure 1 depicts T2W MRI spine saggital view showing abnormal T2 signal with cord swelling the cervicothoracic cord.



CONCLUSION

LETM is a heterogenous clinico-radiologic spectrum with varied etiologies, most commonly associated with NMOSD. Early Identification of the etiology plays a major role in best therapeutic management, prognostication and prediction of risk of recurrence.

Relapse prevention is crucial and achieved with long-term immunosuppression.

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