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LONGITUDINAL EXTENSIVE TRANSVERSE MYELITIS: A PROSPECTIVE, OBSERVATIONAL STUDY

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ABSTRACT Along with neuromyelitis optica spectrum disorder (NMOSD), there are various other causes of longitudinally extensive transverse myelitis (LETM). This study is planned to evaluate etiological, clinical and radiological profile of LETM. This was a prospective, observational, descriptive study. Total 46 (male 25, female 21) subjects were enrolled. Maximum patients were from age group of 20-40 years (47.82%). Common clinical presentations were paraparesis (30, 65.21%) and bowel/bladder disturbance (34, 73.91%). Most frequent location of LETM in MRI was dorsal cord (17, 36.95%). We found NMOSD (22, 47.82%) as most common etiology of LETM. We also found LETM of undetermined etiology in 7 (15.21%) subjects. LETM has broad spectrum of etiologies. Involvement of dorsal spinal segment is common whereas in NMOSD it is cervical. NMOSD is the most common cause of LETM and in about half of the patients other etiologies were found. Infectious causes like tuberculous myelitis needs specific treatment.

KEYWORDS : etiology, clinical, longitudinally extensive transverse myelitis, neuromyelitis optica spectrum disorder

INTRODUCTION:

Longitudinally extensive transverse myelitis (LETM) is characterized by contiguous inflammatory lesion of spinal cord extending to three or more vertebral segments on Magnetic resonance imaging (MRI).[1] If less than 3 vertebral segments are involved, it is called as short segment myelitis. Clinically LETM patient presents with bilateral (not necessary symmetric) motor, sensory, autonomic and bowel/bladder disturbances and usually progress for less than 4 weeks if left untreated. Although, neuromyelitis optica spectrum disorder (NMOSD) is the most common cause of LETM, other causes are autoimmune, inflammatory, infectious, neoplastic, paraneoplastic, vascular and metabolic disorders. [2] Some of these are readily treatable and some need long term immunosuppression. Recent study from India reported idiopathic cause is the most common. [3] Evaluation of patient is necessary depending on differential diagnosis for early diagnosis and prompt treatment to reduce disability. There are few studies on LETM from India. Further studies are required to describe phenotype and etiology of LETM for advances in treatment. This will also improve the approach and limit the number of tests depending upon distinct clinical and radiological features.

Hence, this study is planned to evaluate etiological, clinical and radiological profile of longitudinal extensive transverse myelitis.

MATERIALS AND METHODS:

This was a prospective, observational, descriptive study conducted in the Department of Neurology at a tertiary teaching hospital, between January 2018 and March 2022. This study was approved by the Institutional Ethics Committee. A written informed consent was obtained from patients or their legal guardians.

All patients presenting with clinical features of acute myelitis were subjected to MRI. The MRI protocol (1.5 or 3 Tesla) for brain included T1-weighted images (T1WI), T2-weighted images (T2WI), fluid-attenuated inversion recovery (FLAIR) images, diffusion-weighted images (DWI) with apparent diffusion coefficient (ADC) maps and contrast-enhanced T1-weighted images. In spinal cord MRI, T2-weighted sagittal and axial section, and contrast enhanced T1-weighted images.

The inclusion criteria were longitudinal extensive involvement of three or more vertebral segments of spinal cord on T2WI of MRI with acute (1-7 days) or sub acute (8-28 days) presentation.

Total 46 (male 25, female 21) subjects were enrolled. Their demographic profile, clinical features, laboratory investigations, MRI (brain, spinal cord, optic nerve), diagnosis, treatment, response to treatment and duration of stay in hospital were noted. Investigations noted were cerebrospinal fluid analysis (CSF), Serum NMO and MOG antibodies, markers of connective tissue disorders (ESR, ANA), CSF oligoclonal bands, Serum ACE level, serum vitamin B12 level and serum HIV (ELISA) test. Other tests were done depending upon etiology.

Diagnosis of NMOSD was done as per Wingerchuk et al (1) and of multiple sclerosis was done as per revised McDonald criteria. [4] Mean, percentage and ratio were used for statistical analysis.

RESULTS:

In our study, 46 (male 25, female 21) subjects were enrolled. Maximum were from the age group of 20-40 years (47.82%). Prevalence was slightly more in male with male to female ratio was 1.19:1. In 40 (86.95%) subjects, presentation was acute.

Common clinical presentations were paraparesis (30, 65.21%) and bowel/bladder disturbance (34, 73.91%) with visual impairment in 15 (32.60%). (Figure 1)

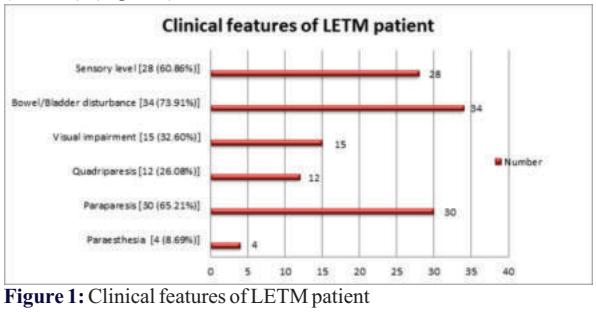

Figure 1: Clinical features of LETM patient

Most frequent location of LETM in MRI was dorsal cord (17, 36.95%) and least frequent was dorso-lumbar (4, 8.69%). More than 6 vertebral segments were involved in 21 (45.65%) subjects. Central enhancement on T2WI was seen in 39 (84.78%) and contrast enhancement on T1WI was seen in 37 (80.43%). In MRI, brain was involved in 22 (47.82%) and optic nerve was involved in 15 (32.60%). (Table 1)

Table 1: MRI Features Of LETM Patients

MRI Feature	Number (Percentage)
Location of LETM	
Cervical	11 (23.91)
Cervico-dorsal	14 (30.43)
Dorsal	17 (36.95)
Dorso-lumbar	4 (8.69)

Number of vertebral segments involved in LETM	
3 to 6	25 (54.34)
More than 6	21 (45.65)
Pattern of T2 enhancement- central/complete	39 (84.78)
Contrast enhancement	37 (80.43)
Brain	22 (47.82)
Optic nerve (ON)	15 (32.60)
Unilateral ON	7 (15.21)
Bilateral ON	8 (17.39)

In our study, we found NMO spectrum disorder (NMOSD) (22, 47.82%) as most common etiology of LETM with Myelin oligodendrocyte glycoprotein (MOG) antibody associated demyelination, multiple sclerosis and tuberculous myelitis in 3 (6.52%) subjects each. We also found LETM of undetermined etiology in 7 (15.21%) subjects and post infectious myelitis in 2 subjects. (Table 2) One female patient diagnosed with Sjogren's syndrome.

Table 2: Etiology Of LETM

Serial number	Disease	Number (Percentage)
1	NMOSD NMO Positive NMO negative	22 (47.82) 20 2
2	MOG	3 (6.52)
3	Multiple Sclerosis	3 (6.52)
4	ADEM	1
5	Infectious/post infectious Tuberculous myelitis Neurosyphillis Post- infectious myelitis	 3 (6.52) 1 2
6	Vascular Spinal dural AV fistula Anterior spinal artery infarction syndrome	 1 1
7	Spinal cord tumour- astrocytoma	1
8	Sjogren's syndrome	1
9	Undetermined etiology (Idiopathic)	7 (15.21)

In NMOSD patients, most common age group affected was 15 to 40 years (14, 63.63%) and male to female ratio of 1:2.6. Most frequent location of LETM was cervical and cervicodorsal (9, 40.90% each)(Figure 2). Contrast enhancement on MRI was seen in 19 (86.36%) subjects. Optic nerve involvement was seen in 10 (45.44%) subjects. We found high protein in CSF in 18 (81.81%) subjects. Out of 22, 11 (50.00%) patients treated with injection methyl prednisolone and plasmapheresis. Relapse was observed in 14 (63.63%) patients.

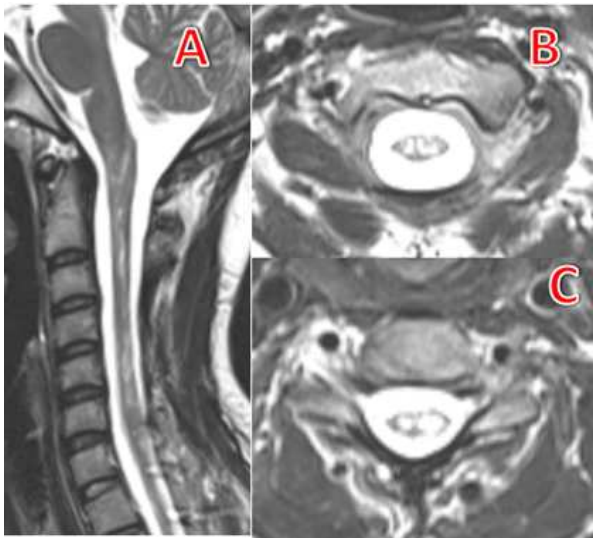

Figure: 2 MRI of 21 year female diagnosed with NMOSD with second attack of LETM. A- sagittal section of T2WI showing LETM with cord atrophy, B, C- axial section of T2WI at different level showing central enhancement

Average duration of hospital stay of LETM patient was 25 days.

DISCUSSION:

We enrolled total 46 patients and found that it was more common in less than 40 years age group. This can be explained by immunological etiology being the most common and was similar to recent studies published from India. [3, 5 and 6] Incidence was slightly more in male with male to female ratio of 1.19:1 and Devi A et al, also found male preponderance. However, studies from other countries [7] reported higher incidence in females. This can be explained by racial difference, access to medical services and etiological difference. Most of the patients (40, 86.95%) present with acute onset and percentage was higher than other study [6]. This can be described by etiological differences.

We found weakness (paraparesis and quadriparesis) was most common presentation followed by bowel/bladder disturbance. Similar to recent study [6], clinically sensory level was found in 28 (60.86%) patients.

In our study, most common location of LETM was dorsocervical (17, 36.95%) followed by cervical with central enhancement in T2WI and post contrast enhancement in most of the subjects. A study by Pekcevik et al [8] on MRI features in LETM also found similar observations. More than 6 segment involvement is common in LETM and also reported by recent Indian study. [3] Brain involvement is also found in patients other than NMOSD. Pekcevik et al [8] described brain lesions in LETM as specific and non-specific for diseases.

Almost half patients were diagnosed with NMOSD and 3 patients each with MOG antibody associated demyelination and multiple sclerosis (MS). With one patient diagnosed with acute demyelinating encephalomyelitis (ADEM), total demyelination patients contribute to 63% of LETM patients. Similar data was reported by Jain et al [5]. Although LETM is rare in MS as described in literature, we found in 3 patients of LETM with MS and recent studies [5] have also reported LETM in multiple sclerosis. Three patients were diagnosed with tuberculous myelitis. A study from India has reported 4 cases of tuberculous myelitis with LETM. We found 2 patients of post infectious myelitis, 1 patient each of neurosyphillis, Spinal dural AV fistula, anterior spinal artery infarction syndrome, Spinal cord astrocytoma and Sjogren's syndrome. Laboratory investigations and distinct radiological features required for diagnosis of these diseases is described in literature. Sjogren's syndrome (SS) presenting with LETM was already reported and NMOSD can be a neurological complication of SS as both of these diseases share common pathophysiological mechanism.

Similar to previous studies [5, 6], we could not find etiology in 15.21% of patients in spite of necessary investigations. Other studies also found Vitamin B 12 deficiency [6] and spinal cord compressive lesion [9] as etiology of LETM, however we didn't get.

Out of 22 patients of NMOSD, 20 were serum NMO antibody positive. Similar to recent study [6] females are more affected with male to female ratio of 1:2.6. As compared to other causes of LETM, optic nerve involvement was more common in NMOSD. Similar to study by Kim et al [10], we found central spinal cord enhancement on T2WI in all patients and frequent post contrast enhancement on T1WI. Contrary to Kitley et al [10], we found CSF pleocytosis in only 31.8% of patients. This could be explained by effect of treatment with steroid. Similar to previous study [10] we found high protein in CSF in NMOSD patients. All patients were treated with injection methyl prednisolone and 50% were treated with additional plasmapheresis. All patients except for one improved with treatment. As per the recent studies, early plasmapheresis is highly effective in acute attack of NMOSD. Like other studies, relapse in NMOSD was observed in 63.63% patients. Limitation of the study was being a single centre study and long term follow up was not done. Future studies should be multi-centre with long term follow up especially of NMOSD patients.

CONCLUSION:

LETM has broad spectrum of etiologies. Motor weakness and bowel/bladder disturbance are the most common clinical presentation. Involvement of dorsal spinal segment in LETM is common whereas in NMOSD it is cervical. NMOSD is the most common cause of LETM with female preponderance. In about half of the patients other etiologies were found. Distinct clinical, laboratory and radiological features will help in arriving at diagnosis and starting early treatment. Infectious causes like tuberculous myelitis is also the cause of LETM and need specific treatment, different from immunosuppression.

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