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Original Article

Decade-Long Study on Phenotype and Prognosis of Lupus Myelitis (LM) in Systemic Lupus Erythematosus (SLE): Insights from a Single-Centre in India

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Abstract

Background: Systemic lupus erythematosus (SLE)-associated myelitis or lupus myelitis (LM), one of the twelve neuropsychiatric lupus (NPSLE) syndromes, is a rare but severe complication of lupus. In this study, we observed the clinical and imaging profiles of LM patients to assess long-term outcomes.

Methods: This was a retrospective study; data of LM with follow-up were extracted from the lupus registry in the last 15 years (2007-2022). Clinically, they were divided as grey matter myelitis (GMM) versus white matter myelitis (WMM). Disease activity was assessed by the SLE Disease Activity Index (SLEDAI-2K) & outcome by death, recurrence, and modified Rankin Score (MRS). Survival analysis was performed using the Kaplan-Meier (KM) and Weibull survival probability tests.

Results: 38 patients were included out of 1700 lupus patients over the last 15 years. Among them, 26 patients presented with GMM, and 12 presented with WMM. Patients with GMM had significantly higher SLEDAI and MRS at discharge compared to WMM patients. (*P*-value—.021 and .08, respectively). White matter myelitis patients had higher levels of anti-cardiolipin antibodies. (*P*—.005) MRI-positive myelitis was associated with higher dsDNA levels compared to MRI-negative myelitis (*P*-.03), but there was no significant difference in disease activity or outcome. The Weibull probability plot indicated poor survival status in GMM.

Conclusion: The prevalence of LM in our cohort is around 2%. Grey matter myelitis is associated with more active disease and significant disability. Survival analysis revealed a poor outcome for GMM in this study.

Keywords: GMM, lupus myelitis, MRI-negative LM, MRS score, WMM

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Introduction

Systemic lupus erythematosus (SLE)-associated myelopathy is classified as one of the twelve central neuropsychiatric lupus syndromes (NPSLE), characterised by neurological deficits caused by spinal cord pathology directly related to SLE. Among these syndromes, lupus myelitis (LM) is considered the prototype of lupus myelopathy. While LM is rare, it is a severe complication of lupus, with a prevalence estimated at approximately 1% based on data collected worldwide.[1,2] Clinically, myelitis can be categorised into two subsets: patients presenting with acute flaccid paralysis are presumed to have grey matter involvement, known as grey matter myelitis (GMM). In contrast, those presenting with spastic paraparesis are believed to have white matter involvement, known as white matter myelitis (WMM).[3] Although this clinical subtype has yet to be extensively studied in lupus, large cohort studies that differentiate these phenotypes in cases of idiopathic myelitis have been published.[4] Lupus myelitis diagnosis typically relies on clinical features supported by magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF) studies. However, there are cases where the imaging results may be negative despite evident clinical features. These cases are referred to as MRI-negative LM.[5] Identifying and characterising these instances is crucial for improving diagnostic accuracy and patient management. In this study, we aimed to retrospectively collect data on patients diagnosed with LM and analyse their clinical presentation, imaging features, and long-term outcomes.

Objectives

The objectives of the study were to: 1. Evaluate the clinical and investigative profile of LM. 2. Identify and compare two distinct clinical phenotypes of LM and analyse disease activity and outcomes between the

two groups. 3. Compare the characteristics of LM cases with positive and negative findings on MRI. 4. Analysis of survival status in LM cases.

Material and Methods

Patients

This study utilised a retrospective design in our hospital setting. The relevant clinical data about cases of LM occurring from 2009 to 2022 were gathered from the lupus registry of the Rheumatology and Clinical Immunology Department at NIMS University (Approval No: 1189, Date: 2009). To be classified as LM, patients had to exhibit neurological deficits characterised by motor impairment such as paraplegia or quadriplegia or sensory weakness accompanied by abnormal bladder or bowel function, all in the context of a confirmed lupus diagnosis while ruling out other potential causes.

Inclusion criteria-Patients were required to fulfil the classification criteria for SLE, which could be either the revised American College of Rheumatology (ACR) criteria from 1997 or the SLE international collaboration classification criteria (SLICC) from 2012.[6,7] The diagnosis of acute transverse myelitis was established using the diagnostic criteria proposed by the Transverse Myelitis Consortium Working Group.[8] Patients who presented with concomitant autoimmune diseases and demonstrated radiological evidence of mass lesions or abscesses were excluded from the study. Before enrollment, written informed consent was obtained from all eligible patients in their native language. The study adhered to the principles of the Declaration of Helsinki.

Clinical and Investigation Profile

Basic demographic and clinical information were obtained from the Lupus Registry. Additional information was obtained through telecommunications to supplement this data. The Systemic Lupus Erythematosus Disease Activity Index 2000 (SLEDAI-2K) was utilised to assess the disease activity during the myelitis episode. The levels of dsDNA were measured using the ELISA method, and complement levels (C3 and C4) were determined through nephelometry. Anti-cardiolipin (ACL) antibodies (IgG and IgM) were measured using ELISA. However, Lupus anticoagulant (LAC) and anti-beta-2-Glycoprotein 1 (B2GP1) antibodies were unavailable for most patients in the record and hence not included in the study. Anti-neuromyelitis optica (anti-NMO) or anti-myelin oligodendrocyte glycoprotein (anti-MOG) antibody tests were conducted for selected patients.

The outcomes of the illness were categorised as death, recurrence, or disability. Death could occur either during the patient's hospital stay or during the follow-up period. Disability was assessed using the modified Rankin scale (MRS)—a well-known validated scale for neurological deficits in CNS events. The MRS scale ranges from 0 to 6, where 0 indicates no disability, and 6 indicates the patient's death. The MRS was documented for the patients at discharge and on the last follow-up.

Imaging and CSF Analysis

We documented the MRI (1.5 T or 3 T, GE or Siemens) of the whole spine with or without contrast and the CSF examination (cell count and protein level) findings of the patients whenever the information was accessible.

Statistical Analysis

Demographic factors, clinical characteristics, and the outcomes of patients with LM were analysed using percentages and mean values. To compare the differences between patients with GMM and WMM and patients with positive and negative MRI findings of LM, the Chi-square test or Student's t-test was utilised. A *P*-value less than .05 was considered statistically significant. Survival analysis was conducted using both Kaplan-Meier and Weibull statistics based on the length of survival measured in months from the last myelitis event.

Main Points

- Lupus myelitis is a severe complication of SLE, necessitating early diagnosis and aggressive immunosuppression.
- Grey matter myelitis (GMM) is associated with higher disease activity and worse outcomes compared to white matter myelitis (WMM).
- MRI-negative lupus myelitis exists, highlighting the importance of clinical and cerebrospinal fluid (CSF)-based diagnosis.
- Patients with GMM have poorer survival rates, underscoring the need for intensive management and long-term follow-up.

Result

Demographic and Baseline Characteristics

Among 1700 patients diagnosed with SLE, 38 individuals were clinically diagnosed with LM, indicating a prevalence rate of approximately 2%. The medical records of all LM patients were scrutinised, and it was observed that these individuals were admitted to either the rheumatology or neurology ward for treatment. The mean age at presentation was 25 ± 8.5 years with a female-male ratio of 35:3. Twenty-six patients presented with GMM, while twelve had WMM-like presentations. The disease duration was less than five years for 30 patients (78.9%). Other NPSLE events were present in 64% of cases, with seizure and peripheral neuropathy being the most common. Extra neurological major organ involvement included nephritis and myocarditis in 14 and 6 cases, respectively. The mean SLEDAI score was 14.8 ± 8.9, and the mean MRS score at discharge was 3.47 ± 1.53. Infections during hospitalisation or follow-up were seen in 25 patients, with five severe infections leading to sepsis. Most of the infections were either due to long-standing immunosuppressive therapy or neurogenic bladder.

Lupus myelitis was observed as an initial feature in 19 (54.2%) cases. The mortality rate is high in this group at 37% (7/19), with a mean SLEDAI of 15.4. Clinical presentations, such as GMM, were seen in 12 cases and WMM in 7 cases. Apart from methylprednisolone and cyclophosphamide, IVIG and plasma exchange were used for one patient, respectively.

Antibody Association

Elevated dsDNA and hypocomplementemia at the time of LM attack were present in 25 (68%) and 31 (86%) cases, respectively. ACL antibody positivity was seen in 13 out of 33 patients (39.3%). For five patients, anti-NMO antibodies were sent, with one being positive.

Imaging and Fluid Analysis

Magnetic resonance imaging imaging was conducted for 32 patients, revealing that 13 had MRI results suggestive of LM. The remaining patients did not show any signs of LM on their MRI scans, but all of them had definite myelitis as per clinical diagnosis. Cerebrospinal fluid was done for 19 patients, with 12 (63%) showing elevated protein and 7 (37%) showing pleocytosis. (Table 1 - Demographic and Clinical Characteristics).

Grey matter myelitis vs White matter myelitis

The GMM-like presentation was observed in 26 patients, while the WMM-like presentation was seen in 12 patients. In the GMM group, the median SLEDAI (interquartile range) was 12 (20) compared to a median SLEDAI of 9 (14.5) in the WMM group **(*P*=.021).** Interestingly, anti-cardiolipin antibodies were

Table 1. Demographic and Clinical Characteristics

	Variable (N = 38)	Frequency	Percentage (Where Applicable)
Demography	Mean age at presentation (SD) (years)	25 ± 8.5	-
	F:M	35:3	-
	Diagnosis with myelitis at the same time	19	54.2
	Myelitis evolving in less than one month	33	86.8
Clinical characters	Fever at the onset of LM	35	92.1
	Flaccid/spastic paralysis	26/12	68.4/31.5
	Sensory loss	30	78.9
	Bladder involvement	31	81.5
Investigation	CSF pleocytosis	7/19	36.8
	CSF elevated protein	12/19	63.1
	MRI done	32//38	84.2
	Positive MRI findings	13/32	40
	Mean MRS score (SD)	3.57 ± 1.5	
	Mean SLEDAI (SD)	14.8 ± 8.9	
	Anticardiolipin antibody positive	13/33	39.3
	Anti-dsDNA antibody positive	25/37	67.5
	Low complements	31/36	86.1
Treatment	Pulse corticosteroid (methylprednisolone)	32	84.2
	Cyclophosphamide	26/37	70.2
	Rituximab for LM	2	5.2
	Intravenous immunoglobulin	1	2.6
	Oral anticoagulant	6	15.7
Complications	Total death	13	34.2
	Relapse	7	18.4
	Total no of infection	25	65.8
	Urinary tract infection	12	31.6
	Herpes Zoster	2	5.3
	Sepsis	5	13.2

more frequently positive in the WMM group **(*P*=.035)**. However, the two groups had no significant differences regarding clinical parameters, dsDNA and complement levels. Six patients with GMM-like presentation had succumbed during their hospital admission. In contrast, no patients with WMM-like presentation died during admission, indicating a potentially better prognosis in this subgroup. The Mean MRS was 3.8 ± 1.7 in GMM and 3 ± 1.18 in WMM, predicting a poor outcome in GMM (*P*=.08). Anticoagulants were more commonly prescribed in WMM (*P*=.04). Although the mean survival time was higher in GMM than in WMM, it was not statistically significant. Urinary tract infections were more commonly seen in WMM (Table 2 shows the comparison of GMM versus WMM).

MRI-Negative LM

Magnetic resonance imaging was performed on 32 patients, with 19 without imaging changes. MRI-positive lesions included longitudinally extensive transverse myelitis (LETM) in four patients or localised to the cervical, thoracic, or conus segment in nine patients. Upon comparing both groups, there were no significant differences in the mean SLEDAI or MRS score. However, serum anti-dsDNA levels were significantly higher in the MRI-positive group (*P*=.02). (Table 3 - MRI positive versus MRI negative myelitis).

Outcome and Relapse

During the follow-up period, a total of 13 patients succumbed to their condition. Among them, six patients died during hospital admission (online supplementary Table 1 -In-hospital LM mortality data). Sepsis was the primary cause of death in 83% of cases. Additionally, seven patients had a history of a second attack of myelitis (online Supplementary Table 2 - Relapse LM data). Notably, two of them underwent a second episode within a year of their initial episode.

One patient with LETM and anti-NMO antibody positivity had a total of three episodes of LM to date, and her clinical phenotype always suggested a GMM lesion (flaccidity with hyporeflexia). Magnetic resonance imaging imaging was conducted on all patients with relapses, revealing that three patients exhibited evidence of long-segment myelitis (cervical to conus), two had localised involvement (cervical and thoracic), and two were MRI negative. Anti-NMO antibody testing was performed on five patients, and three of them had recurrent myelitis, with one patient testing positive for anti-NMO antibodies. Escherichia coli predominantly caused urinary tract infections (UTI), with two experiencing recurring UTIs, significantly impacting their quality of life. Bedsores were present in 16% of the cases.

Survival Analysis

We performed a Kaplan Meier (K-M) survival analysis comparing GMM and WMM, and the results indicated that the mean survival time for GMM was 110 months (95% CI-76-145), which was longer than the mean survival time for WMM at 103 months (95% CI-82-123 months) but without statistical significance, (Log Rank (Mantel-Cox),*P*=.207) (Figure 1 - K-M analysis). We also used machine-learning estimates to determine probability plots for life status. We used Weibull, lognormal, Exponential and Log logistic survival plots. It was found that Anderson Darling's test value with all of these statistics correlated with each other, indicating a normal distribution of the dataset with a minimum departure from the line of normality. (online Supplementary figure). Among them, Weibull was found to be the closest to the line of normality and was used to formulate a final model of survival status. It suggested that 25%, 50 %, and 75% of patients with GMM had a chance to die within 13 85 376 months from the first attack, while the same for WMM is 96,256, 556 months, respectively, implying GMM has a worse survival outcome (Figure 2 - Weibull probability plot for life status).

Discussion

A diagnosis of LM is established when myelitis occurs in the context of SLE, and other secondary causes are excluded. Though its

Table 2. Showing Comparison of GMM Versus WMM

	Variables	GMM	WMM	*P*-value
Demography	No of the patients (n)	26	12	-
	Age (Median) (IQR) (years)	24(10)	26.5(14)	-
	F:M	24/2	11/1	-
Clinical presentation	Myelitis as the initial presentation	13/25	6/12	.9
	Bladder involvement	24	10	.40
	Other NPSLE events	17	6	.36
Other organ involvement	Nephritis	11	3	.30
	Myocarditis	6	0	-
	GI (enteritis, pancreatitis)	6	2	.65
	Thrombocytopenia	13	3	.14
	Leukopenia	10	4	.76
Investigation	CSF pleocytosis	6/13	1/6	.22
	Elevated CSF protein	8/13	4/6	.83
	Low complement	22/24	10/11	.67
	dsDNA positivity	19/26	7/11	.57
	ACL (IgG/IgM) positivity	6/23	7/11	.035
	Imaging-MRI positive of the spine	9//21	4/11	.72
	long segment involvement	3/9	1/4	.76
	Median SLEDAI (IQR)	12(20)	9(14.5)	.021
Treatment	Methylprednisolone pulse	23/26	9/12	.28
	Cyclophosphamide	17/25	9/11	.32
	Rituximab	1	1	
	Plasma exchange	2	0	-
	Intravenous immunoglobulin (IVIG)	1	0	-
	Anticoagulant	2/26	4/11	.04
Outcome	Mean MRS at discharge	3.86 + 1.7	3 + 1.2	.08
	No of death	10/26	3/12	.42
	Death during admission	6	0	-
	Recurrence of myelitis	5	2	.85
	Mean survival time (months)	110.8	103.3	.20
	Chance of death of 25% of patients (months) from last attack	<13	<96	
	Chance of death of 50% of patients	<85	<256	
Infections	Infections on follow up due to immunosuppressive therapy and residual neurological deficit	13 (sepsis-5, infected ulcer -4, UTI-4, zoster 1)	7 (UTI,1 -sepsis)	.63
	Severe infection	5	1	
	Urinary tract infection (UTI)	4	7	.004

prevalence in SLE is less compared to other NPSLE manifestations, lupus myelitis has severe complications and a poor prognosis in most cases. This study reported 38 cases of LM out of 1700 lupus patients followed up in the Rheumatology ward and outpatient department in a tertiary care hospital over the last 15 years.

We conducted a comprehensive literature review on LM using MeSH terms in PubMed and other databases. Table 4 shows a detailed comparison of the demographic and clinical characteristics of our patients with other cohorts.

Prevalence and Demography

Different studies found a prevalence of 0.5-3% for LM across the world (Mehta et al.—0.56% in 1768 cases,[9] Hyrvb et al.—2.1% in 233 cases,[10] and Mok et al.—3.2% in 315 LM patients).[11] Our cohort's prevalence was 2%. Lupus myelitis may be the initial manifestation or occur many years after SLE diagnosis. In most case series, 60-70% of patients had LM as the initial lupus manifestation,[11,12] which is consistent with our study, where 54% of cases had LM as the initial presentation. Myelitis can also be a late manifestation, as Saison et al.[13] reported 8 out of 20 LM cases with myelitis onset after a median period of 8.6 years. In our case series, 17 patients had a later onset of myelitis. Lupus myelitis is more common in patients with high disease activity. In different studies, LM occurred in 60-70% of cases with increased disease activity (SLEDAI > 4).[14] In our research, it was seen in 23 (65%) patients with SLEDAI > 4, while 12 patients had SLEDAI < 4.

Antibody Levels

Mok et al.[11] found that 40% of LM cases had positive anti-dsDNA antibodies, and 30% had hypocomplementemia at diagnosis. Similarly, Hryb et al.[10] reported a high prevalence of 80% positive dsDNA and 80% low complement levels in LM cases. Our study observed that 46% of patients had positive dsDNA antibodies, and 57% had low complement levels during myelitis attacks. Furthermore, Lavalle et al.[19] discovered a significant association between myelitis in SLE and the presence of antiphospholipid (APL) antibodies. Similarly, Katsiari et al.[20] found that 54% of patients had APL antibodies detected at the onset of transverse myelitis. However, their positivity did not predict the involvement of the thoracic spine, which is often associated with thrombosis-related injury. Additionally, APL antibodies did not correlate with relapsing myelitis, other CNS manifestations of lupus or worse clinical outcomes. They opined that adding anticoagulants to the patients was no additional benefit. Cruz et al.[17] reported that eleven of the fifteen (73%) LM cases had APL positivity. Six of the eleven APL-positive patients were treated with aspirin, and five received warfarin. They found that those

Table 3. MRI Positive Versus MRI Negative Myelitis

Variables	MRI Positive	MRI Negative	*P* Value
n	13	19	
Mean age (years) and sex	27.07 ± 11.7, all females	24.05 ± 7.09, two males	.36
Myelitis as the initial presentation	6	12	.9
Initial bladder involvement	3	6	.27
Low complement	11	15	.16
dsDNA positivity	11	9	.03
Anti-NMO antibody	1/4	-	
Mean SLEDAI (SD)	12.5 ± 8.2	13.05 ± 9.5	.86
Mean MRS (SD)	3.8 ± 1.4	3.2+1.5	.26
Death	3	7	.41

treated with both cyclophosphamide and warfarin had a significant recovery of their symptoms. In our study, 26% of cases were positive for ACL antibodies, and six patients received anticoagulants as adjunctive therapy.

CSF Study

Cerebrospinal fluid analysis is commonly abnormal in most patients and exhibits significant variability. In hyperacute cases, there may only be a slight increase in proteins and polymorphonuclear pleocytosis with hypoglycorrhachia. Hyrb et al.[10] reported that CSF analysis was abnormal in all five LM cases with variable results. Mehta et al.[9] reported elevated CSF protein in 55.5% of cases of LM and lymphocytic pleocytosis in 22.2%. Our study observed elevated CSF protein levels in 63% of patients, with 37% displaying pleocytosis. Birnbaum et al. found that GMM had higher rates of elevated CSF protein and pleocytosis than WMM. However, our study did not observe a significant difference in CSF findings between GMM and WMM cases.

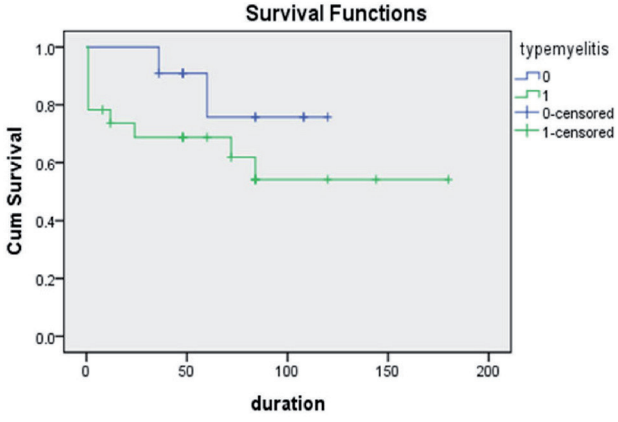

Figure 1. Kaplan-Meier analysis.

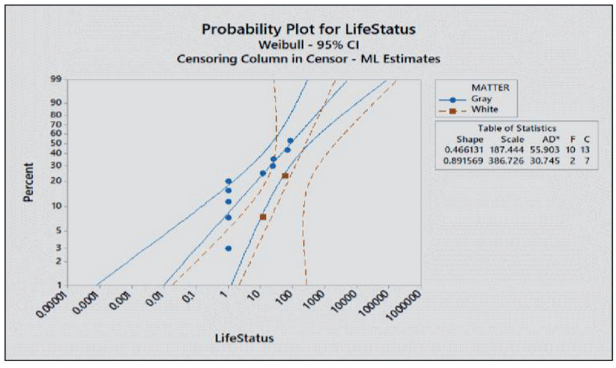

Figure 2. Weibull probability plot.

Grey matter myelitis vs White matter myelitis

The spinal cord has distinct neuroanatomic tracts. Lesions that preferentially affect the central grey matter can produce a pattern of weakness associated with decreased tone (i.e., flaccidity) and hyporeflexia. Lesions that preferentially affect the outer white matter are instead associated with increased tone (i.e., spasticity) and hyperreflexia. In 2009, Birnbaum et al.[3] reported two distinct cohorts of LM with clinical features suggesting grey and white matter myelitis. They observed that GMM is more common in active lupus with poor long-term outcomes. Early urinary retention and fever were two ominous signs of GMM involvement in lupus and urgent immunosuppression. In contrast, the WMM course is more indolent, associated with anti-Ro or lupus anticoagulant antibodies and could be seen in less severe lupus. In their study, Williams et al.[18] found that two out of 15 cases had GMM-like presentation with no difference in disease activity and disease outcome in both groups. Though there was a gross discrepancy in the findings of both these studies, our study favoured Birnbaum et al., which had high disease activity and poor outcomes in GMM compared to WMM. However, surprisingly, in this cohort, a patient with anti-NMO positivity and LETM had a GMM-like presentation in each relapse of LM.

MRI Negative Lupus Myelitis

Magnetic resonance imaging can be negative in 20% of cases of transverse myelitis, more frequently seen in idiopathic transverse myelitis, Myelin oligodendrocyte associated antibody disease (MOGAD), paraneoplastic myelopathy and GAD-65 receptor associated myelopathy.[21,22] Less sensitivity to detect inflammation and timing of imaging might contribute to this finding. Mok et al.[11] reported abnormal MRI signals in 56% of cases at the time of active myelitis, while Kovacs et al.[12] reported abnormalities in 70% of cases. Das et al.[23] reported 8 cases of MRI-negative lupus myelitis and proposed a new subtype of LM, "MRI-negative myelitis with selective tract involvement". All patients with MRI-negative lupus myelitis in our cohort showed high disease activity at the onset of myelopathy. On follow-up, improvement of myelopathy features with no or minimal deficit was observed in 5 of the eight patients (62.5%). In our study, 13 patients had MRI changes while 19 patients were MRI negative, and 7/11 WMM had MRI negative. In contrast, 12/21 GMM had MRI-negative myelitis, indicating that MRI-negative myelitis can be seen in many lupus cases characterised by selective tractopathy.

Table 4. Comparison of our Study with Other Studies

Variables	Zhang et al[15]	Hryb et al[10]	Kovacs et al[12]	Mehta et al[9]	Hiroshi Oiwa et al[16]	D Cruz et al[17]	Williams et al[18]	Present Study
Population	45	5/233	14/600	10/1768	3	15	7 had LM	38
Same time diagnosis	22	3	7	7	1	15		19
Mean age (years)	36.6	25.4	42	21	31, 25, 35 years	38.7	41	25
Neurological other than myelitis	9	-	8	2	1	2	0	24
Renal	18	1	5	1	0	2	0	14
Hematological	24	-	2	0	1	7	0	Tcpenia 16, Leukopenia 13
APL positivity	13	3	6	4	2	11	2	13
MRI abnormality	29	all	8	60%	67%	13/15	13/14	13/32
Death	-	1	no	no	no	1	1	13
Relapse	ASIA scale D	2	2 had partial recovery	4/10	0	no	1	7
CYC	36	3	9	6/10	2	11	2	26
Anticoagulant	No data	3	No data	No data	No data	5	No data	6

Treatment and Outcome

Recent reviews have recommended combining combined treatment with intravenous methylprednisolone and cyclophosphamide, which appears more effective than methylprednisolone alone.[24,25] Cruz et al.[17] study found that one patient died due to chronic myelomalacia who was only on steroids, while patients with both methylprednisolone and cyclophosphamide had significant recovery. Plasma exchange therapy, intravenous immunoglobulin and rituximab have been used in refractory cases.[10] Rituximab is often used in NMO-positive LETM in lupus and plasma exchange in refractory NMO-positive cases.[26] In our study, rituximab was given to two patients, one with NMO-positive, recurrent long-segment myelitis. The role of anticoagulants as an adjunctive therapy has variable results in the literature, mainly in APL-positive cases.[10] Cruz et al.[17] reported a good outcome with anticoagulants, while Kovacs et al.[12] found no satisfactory results. In our study, anticoagulants were used for 6 cases without any significant improvement.

The three primary outcomes observed in LM were death, relapse and recovery with variable prognosis. Mehta et al.[9] reported that of the eight patients on follow-up, 90% had no to minimal disability at their last follow-up. Williams et al.[18] showed that the outcomes of subjects were relatively favourable; 87% of subjects were treated with steroids at the onset of myelitis, and 87% had a single episode of myelitis. All subjects had either minor impairment in motor function (strength ≥ 4/5) or normal function at 1-year follow-up. Oiwa et al.[16] reported three cases of LM, all of them requiring wheelchair support and a catheter on follow-up. Birnbaum et al. 11 said that the median EDSS score was higher in GMM compared to WMM in follow-up visits, indicating GMM has more disability in the long term. Hyrb et al.[10] reported 5 cases of LM, with one dying due to sepsis and the rest of them having a significant disability in the long term. In our study, 83% of LM patients were treated with a pulse dose of methylprednisolone, with 83% having a single myelitis episode. 34% of patients had no to a slight disability during discharge, and 64% (16/25) patients were regular follow-up with mild to moderate disability (MRS 1 or 2); most common was residual weakness and urinary incontinence leading to long-term self-catheterisation. Flores Silva et al.[27] reported a five-year mortality of 31% in 35 LM cases because of sepsis (in 10 cases) or pulmonary embolism (in one case) and concluded that the short-term outcome of LM was generally good, but the long-term fatality rate was high. In our study, six patients had in-hospital mortality while seven patients died at follow-up, mainly due to sepsis, suggesting overall poor long-term outcomes in LM.

There are limitations in the study. As this was a retrospective study, some data were missing, potentially causing recall bias and limiting the ability to establish a strong causal association. Anti-NMO antibody, which has a strong association with myelitis, was done for a few patients due to financial constraints. Hence, a strong conclusion on the association of anti-NMO antibodies in LM could not be inferred. Not all patients underwent testing for all three types of anti-phospholipid antibodies. Magnetic resonance imaging with contrast was performed on fewer patients, which could lead to challenges in diagnosing LM through imaging. Modified Rankin Score was used as a clinical outcome measure, though there are new scoring systems of outcome measures in transverse myelitis. The difference between GMM and WMM was purely clinical, and imaging was not analysed to differentiate the site of involvement in the spinal cord.

Lupus myelitis is a rare but severe complication of SLE, with a prevalence of 2% in our patients. It can occur as the first symptom of SLE or develop years after the initial diagnosis. Lupus myelitis is more common in patients with high disease activity, and its prognosis can vary, often leading to severe complications such as bladder problems, urinary tract infections, and the need for assistance with daily activities. In some cases, MRI may not show abnormalities, termed MRI-negative LM. However, there is no significant difference in disease activity or outcomes between cases with MRI-positive LM and those with MRI-negative LM in our study. The clinical differences between GMM and WMM have not been deeply studied due to limited supporting literature.

Ethics Committee Approval: This study was approved by Ethics Committee of NIMS University, (Approval No: 1189, Date: 2009).

Informed Consent: Written informed consent was obtained from the patients/patient who agreed to take part in the study.

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