Section: Neurology

Original Research Article

ASSESSMENT OF CASES OF LONGITUDINALLY EXTENSIVE TRANSVERSE MYELITIS

Zubair Sarkar[1], Sushmita[2]

[1]Assistant Professor, Department of Neurology, GSVM Medical College, Kanpur, Uttar Pradesh, India, India.
[2]Senior Resident, Department of Neurology, GSVM Medical College, Kanpur, Uttar Pradesh, India, India.

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Corresponding Author:
Dr. Zubair Sarkar,
Email: zubair.sarkar89@gmail.com

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Abstract

Background: To assess cases of longitudinally extensive transverse myelitis. **Materials and Methods:** Fifty- six cases of longitudinally extensive transverse myelitis of either gender was recruited in this observational, cross-sectional study. Clinical characteristics, European Database for Multiple Sclerosis (EDMUS) grading scale and Magnetic resonance imaging (MRI) findings were recorded. **Results:** Out of 56 patients, males comprised 32 (57.1%) and females 24 (42.9%). Diagnosis of cases was systemic in 7, neuromyelitis optica spectrum disorder (NMOSD) in 23, infective in 11, other demyelinating myelopathy (ODM) in 10 and idiopathic in 2 cases. The difference was significant (P< 0.05). Onset was acute in 25 and subacute in 31 patients. Clinical features were paresthesia seen in 34, urinary retention in 25, optic neuritis in 12 and weakness in 7 cases. Pattern was paraparesis in 16, quadriparesis in 23, bladder involvement in 50 and respiratory muscle involvement in 37 cases. A non- significant difference was seen (P> 0.05). MRI findings were normal in 26 cases, OBEX lesion was seen in 14 cases, cortical and subcortical lesions in 10 cases and optic nerve lesion in 6 cases. A significant difference was seen (P< 0.05). EDMUS at nadir (median) score was 8 which improved to 2.4 at 6 months. A significant difference was seen (P< 0.05). **Conclusion:** In patients with longitudinally extensive transverse myelitis, most common cause was neuromyelitis optica spectrum disorder (NMOSD) followed by infective.

INTRODUCTION

The neurological condition known as "longitudinally extensive transverse myelitis" (LETM) affects a significant amount of the spinal cord's length and is characterized by inflammation and damage to the spinal cord.[1] Transverse myelitis is the term used to describe inflammation that spans the entire cross section of the spinal cord, whereas "longitudinally extensive" refers to inflammation extending more than 3 vertebral segments.[1] Aside from other underlying medical illnesses like multiple sclerosis (MS), neuromyelitis optica spectrum disorder (NMOSD), or other autoimmune disorders, LETM can also arise alone. Infections, such as bacterial or viral infections, vaccine reactions, or side effects from specific drugs are other possible causes.[3] Though the precise origin of LETM is frequently not fully understood, it is thought to involve an abnormal immune response that leads to inflammation and damage to the spinal cord. The inflammation can disrupt the normal functioning of the spinal cord, causing a range of symptoms.[4]

Common symptoms of LETM include sensory disturbances, motor impairments, bowel and bladder dysfunction, and pain etc. Patients may experience numbness, tingling, or a sensation of "pins and needles" in the affected areas of the body.[5] These sensations typically occur below the level of the inflammation. Weakness or paralysis may occur in the muscles below the level of the inflammation, leading to difficulty in walking, coordination problems, or even complete loss of movement.[6] LETM can affect the nerves that control bowel and bladder function, resulting in urinary or fecal incontinence or difficulty with urination or bowel movements. Some individuals may experience severe pain, which can be sharp, stabbing, or burning in nature. The pain may be localized to the affected area of the spinal cord or radiate to other parts of the body.[7] We performed this study to assess cases of longitudinally extensive transverse myelitis.

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MATERIALS AND METHODS

A sum total of fifty- six cases of longitudinally extensive transverse myelitis of either gender was recruited in this observational, cross- sectional study after considering the utility of the study and obtaining approval from ethical review committee. Patients' consent was obtained before starting the study.

Data such as name, age, gender etc. was recorded. Clinical characteristics such the pattern of weakness, visual symptoms, bulbar symptoms, bladder involvement, need for assisted ventilation, and acute episode treatment were all taken into consideration. The number of relapses and the response to treatment using the European Database for Multiple Sclerosis (EDMUS) grading scale was evaluated. A score of zero on the EDMUS grading system indicates no neurological impairments, One and two are able to run with few symptoms, three and four are able to walk for an unlimited distance but cannot run, five can walk for less than 500 metres without assistance, six requires assistance to walk for only 100 metres, seven requires assistance to walk for only 20 metres, eight is chair-restricted, nine is bed-ridden, and ten means that the person has died from LETM or its complications. Laboratory investigations involved estimation of viral markers, ESR, thyroid profile and vitamin B- 12 level. Cerebrospinal fluid (CSF) analysis was done for glucose, protein, leucocyte count, oligoclonal bands and tuberculosis polymerase chain reaction. Magnetic resonance imaging (MRI) at 1.5 Tesla was used to image the brain and spinal cord. The results were compiled and subjected for statistical analysis using Mann Whitney U test. P value less than 0.05 was set significant.

RESULTS

Table 1: Patients distribution

Total- 56		
Gender	Males	Females
Number (%)	32 (57.1%)	24 (42.9%)

Out of 56 patients, males comprised 32 (57.1%) and females 24 (42.9%) [Table 1].

Table 2: Diagnosis of cases

Diagnosis	Number	P value
Systemic	7	0.02
NMOSD	23	
Infective	11	
ODM	10	
Idioptahic	2	

Diagnosis of cases was systemic in 7, neuromyelitis optica spectrum disorder (NMOSD) in 23, infective in 11, other demyelinating myelopathy (ODM) in 10 and idiopathic in 2 cases. The difference was significant (P< 0.05) [Table 2].

Table 3: Assessment of parameters

Parameters	Variables	Number	P value
Onset	Acute	25	0.72
	Subacute	31	
Clinical features	Paresthesia	34	0.81
	Urinary retention	25	
	Optic neuritis	12	
	Weakness	7	
Pattern	Paraparesis	16	0.54
	Quadriparesis	23	
	Bladder involvement	50	
	Respiratory muscle involvement	37	

Onset was acute in 25 and subacute in 31 patients. Clinical features were paresthesia seen in 34, urinary retention in 25, optic neuritis in 12 and weakness in 7 cases. Pattern was paraparesis in 16, quadriparesis in 23, bladder involvement in 50 and respiratory muscle involvement in 37 cases. A non- significant difference was seen (P> 0.05) [Table 3].

Table 4: MRI findings

MRI findings	Number	P value
Normal	26	0.05
OBEX lesion	14	
Cortical and subcortical lesions	10	
Optic nerve lesion	6	

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MRI findings were normal in 26 cases, OBEX lesion was seen in 14 cases, cortical and subcortical lesions in 10 cases and optic nerve lesion in 6 cases. A significant difference was seen (P< 0.05) [Table 4].

Table 5: Neurological outcome

Neurological outcome	Median	P value
EDMUS at nadir (median)	8	0.001
EDMUS at 6 months	2.4	

EDMUS at nadir (median) score was 8 which improved to 2.4 at 6 months. A significant difference was seen (P< 0.05) [Table 5].

DISCUSSION

Diagnosis of LETM typically involves a comprehensive evaluation by a neurologist, including a thorough medical history, neurological examination, and various imaging studies such as MRI (magnetic resonance imaging) of the spinal cord.[8] Blood tests and cerebrospinal fluid analysis may also be conducted to rule out infectious or inflammatory causes. Treatment for LETM focuses on reducing inflammation, managing symptoms, and promoting recovery.[9] This often involves high-dose corticosteroids to suppress the immune response, intravenous immunoglobulin (IVIG), plasma exchange (plasmapheresis), and sometimes immunosuppressive medications. Physical therapy and rehabilitation may be recommended to improve muscle strength, mobility, and overall function.[10]

The long-term outlook for individuals with LETM can vary depending on the underlying cause and the severity of the inflammation. Some people may experience partial or complete recovery, while others may have ongoing neurological deficits. Close monitoring and ongoing medical care are important to manage symptoms, prevent complications, and optimize quality of life.[11]

In our study, out of 56 patients, males comprised 32 (57.1%) and females 24 (42.9%). Kannan et al.[12] examined the various aetiologies of LETM and compare the differences in their clinico-radiological characteristics and therapeutic outcomes in 42 patients. 80% of the patients in this study were female, with NMOSD having the highest female preponderance (87%). 16 patients had NMOSD, 7 had idiopathic disease, 5 had connective tissue disease, and 4 had an infectious aetiology, among other aetiologies. Patients with infective and systemic causes were more likely to have bladder involvement than patients with infective causes to have respiratory muscle involvement. The majority of instances with more than six segments involved were idiopathic cases. All patients who tested positive for Ro-52 experienced relapses. Compared to other aetiologies, NMOSD had a superior neurological result.

We found that diagnosis of cases was systemic in 7, neuromyelitis optica spectrum disorder (NMOSD) in 23, infective in 11, other demyelinating myelopathy (ODM) in 10 and idiopathic in 2 cases. Houzen H et al.[13] found that the crude prevalence of NMOSD was 4.1/100,000 with substantially more female patients than male patients (12:2). The mean age at onset for anti-aquaporin-4 antibody was 45.2 years, and the positive rate was 78.6%. All patients received preventive therapy, which included steroid or immunosuppressive drug treatment.

In this study, onset in patients was acute in 25 and subacute in 31 patients. Clinical features were paresthesia seen in 34, urinary retention in 25, optic neuritis in 12 and weakness in 7 cases. Pattern was paraparesis in 16, quadriparesis in 23, bladder involvement in 50 and respiratory muscle involvement in 37 cases. Jain et al.[14] observed that the majority of patients had severe bladder dysfunction and paraparesis when they first arrived. Thirteen of the twenty-one patients (32.81%) who had a clinical diagnosis of NMO also had serum NMO antibody results that were positive. Among the other aetiologies of LETM in our series, there were nine cases of multiple sclerosis, six cases of acute disseminated encephalomyelitis (ADEM), five cases of post-infection, four cases of subacute combined degeneration (SCD), four cases of tuberculous myelitis, three cases of spinal arteriovenous malformation (AVM), and three cases of systemic lupus erythematosus (SLE). Despite extensive studies, the aetiology in nine patients could not be determined.

MRI findings were normal in 26 cases, OBEX lesion was seen in 14 cases, cortical and subcortical lesions in 10 cases and optic nerve lesion in 6 cases. EDMUS at nadir (median) score was 8 which improved to 2.4 at 6 months. Angamuthu et al.[15] compared the specific characteristics of 71 LETM patients, it was determined that 56% of the total participants tested positive for AQP4-Ab. The AQP4-Ab positive group was found to have a greater female to male ratio. Holocord involvement was more frequently seen on magnetic resonance imaging in the AQP4-Ab negative group compared to the positive group. Hypointense lesions were not associated with more severe lesions. Older onset age, a higher proportion of female patients, a low incidence of conus involvement, and a higher prevalence of concurrent autoimmune illnesses are the key differences between AQP4-Ab positive and negative cases. There were no differences in the two groups' spasms, ocular neuritis onset, or attack severity.

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Sahoo et al.[16] studied 37 patients with paraplegia/paresis or quadriplegia/paresis. The average age was 35.97 + 13.2 years, with a female: male ratio of 1.05:1. Segments of the thoracic spinal cord were most frequently damaged (56.76%). It is more typical (40.54%) to involve three to five components. 26 LETM patients underwent a serum AQP4 antibody (Ab) test. Nine of them (34.62%) were found to have positive AQP4 Ab results. Clinical diagnoses for a total of 22 patients (59.46%) included NMO, postinfectious myelitis in 4, tuberculous myelitis in 1, subacute combined degeneration in 9, and idiopathic LETM in 9. Prognosis of LETM in the form of expanded disability status scale at 1 year was poor in patients with NMO particularly those with positive AQP4 Ab.

CONCLUSION

In patients with longitudinally extensive transverse myelitis, most common cause was neuromyelitis optica spectrum disorder (NMOSD) followed by infective.

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