Original Research Article

Role of imaging in the evaluation of longitudinally extensive transverse myelitis

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Abstract

Longitudinally extensive transverse myelitis (LETM) is defined as a spinal cord lesion that extends over three or more vertebral segments, as seen on sagittal MRI of the spine. The clinical presentation of a patient with LETM is often dramatic and can consist of paraparesis or tetraparesis, sensory disturbances, gait, bladder, bowel and/or sexual dysfunction. LETM is a characteristic feature of neuromyelitis optica, but such spinal lesions can also occur in various other autoimmune and inflammatory diseases that involve the CNS such as multiple sclerosis, sarcoidosis or Sjogren syndrome or in infectious diseases with CNS involvement. Patients with a neoplastic disorder or traumatic spinal cord injury can also present with longitudinal spinal lesions. Various imaging findings in LETM are done in a prospective study from January 2012 to October 2013.

Key Words: LETM, NMO (Neuromyelitis optica), MRI (Magnetic Resonance Imaging).

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INTRODUCTION

By definition Longitudinally Extensive Transverse Myelitis (LETM) is T2 weighted intra medullary high signal intensity extending across three or more vertebral segments on Sagittal Spinal MRI[1,2]. Neuromyelitis optica is the most common cause of LETM[3,4] Neuromyelitis optica (NMO) is an inflammatory demyelinating disease of the central nervous system (CNS) that most commonly targets the optic nerves and spinal cord selectively, common around fourth decade of life with a female predominance, especially in AQP-4 antibody positive patients, but the reasons are not known. Optic neuritis in NMO can be bilateral or unilateral and tends to be severe, with frequent early permanent vision loss[5]. Advancing our understanding of NMO pathogenesis was the discovery of NMO-IgG autoantibodies against aquaporin 4 (AQP4), the most abundant water channel protein in mammalian central nervous systems[6,7,8] which were detected in 60%to90% of patients with NMO but not in patients with classic multiple sclerosis[9-15]

Neuromyelitis Optica Spectrum Disorders

1. Typical Neuromyelitis optica
2. Asian optic-spinal multiple sclerosis
   - Optic neuritis or longitudinally extensive myelitis associated with systemic autoimmune disease
   - Optic neuritis or myelitis associated with brain lesions typical of neuromyelitis optica (hypothalamic, corpus callosal, periventricular or brainstem)
3. Limited forms of neuromyelitis optica
   - Idiopathic single or recurrent events of longitudinally extensive myelitis (≥3 vertebral segments spinal cord lesion seen on MRI)
   - Optic neuritis: recurrent or simultaneous bilateral

Revised Diagnostic Criteria for Typical Neuromyelitis optica[16]

1. Required criteria: Optic neuritis, Acute myelitis

2. Atleast 2 of 3 supportive criteria:
    - Contiguous spinal cord MRI lesion extending over 3 vertebral segments
    - Brain MRI not meeting diagnostic criteria for multiple sclerosis
    - NMO-IgG seropositive status

Findings in NMO in Spinal cord MRI

- A central cord lesion extending over 3 or more vertebral segments (rather than in the periphery of the cord as generally occurs in patients with prototypic MS)
- In NMO, the long cord lesions are typically cigar-shaped areas of T2 hyperintensity with cord swelling, hypointense on T1, showing variable patterns of enhancement.
- On follow-up MRI studies, cord atrophy and a cavity resembling a syrinx may be present.

Brain neuro imaging in NMO

The NMO-distinctive lesions can be tentatively classified into the following categories

1. Periependymal lesions in area postrema, periaqueductal area, hypothalamus, around lateral ventricles, and undersurface of corpus callosum
2. Medullary lesions centered around gray matter and often contiguous with upper cervical lesions are highly characteristic of NMO and appear to have predictive value for the diagnosis of NMO.
3. Punctate or fusiform T2W hyperintensities typically located in sub cortical or sub callosal white matter is the most frequent finding

AIMS AND OBJECTIVES

To analyse the MR imaging findings in Longitudinally extensive transverse myelitis cases

MATERIALS AND METHODS

Inclusion Criteria: All patients admitted for non-traumatic paraplegia or quadriplegia with or without recurrent optic neuritis in Gandhi Hospital, Secunderabad, between January 2012 to October 2013 were evaluated. The diagnosis of longitudinally extensive transverse myelitis was based on clinical presentation and MRI spine

Exclusion Criteria: Patients with typical lesions of multiple sclerosis, surgical causes of optic neuritis and spinal cord involvement, normal MRI spine or lesions <3 vertebral segments.

Method of collection of data

Study Design: Prospective cross sectional study
Study Sample: 40 patients.
Study Period: January 2012 to October 2013.
Study Area: Department of Radiodiagnosis, Gandhi medical college.
Study Equipment: 1.5 Tesla Siemens MAGNATOM
Ethical clearance from institutional ethics committee has been obtained.

Investigation Protocol

- It included careful evaluation of the clinical features and routine biochemical investigations (complete haemogram, blood urea, serum creatinine, and random blood sugar and serum electrolytes)
- Imaging protocols included for Brain MRI - Non contrast Axial T2- and T1-WI and contrast TI-WI
- Imaging protocols included for spinal MRI
- Non contrast T1, T2-weighted sagittal and axial images
- T1-weighted sagittal and axial images with gadolinium contrast,
- Serum anti- aquaporin-4 (Aqp-4) antibodies was done in 23 patients using ELISA technique. Values below 5 IU/L were considered as negative.

OBSERVATIONS AND RESULTS

Age distribution of LETM cases studied

Table 1: The Mean age of presentation is 30. 18 years, Most of the patients were in 2nd and 3rd decades of their lives

Age	Number of patients	Percentage
1-10	00	0%
11-20	11	27.5%
21-30	11	27.5%
31-40	08	20%
41-50	08	20%
51-60	02	05%

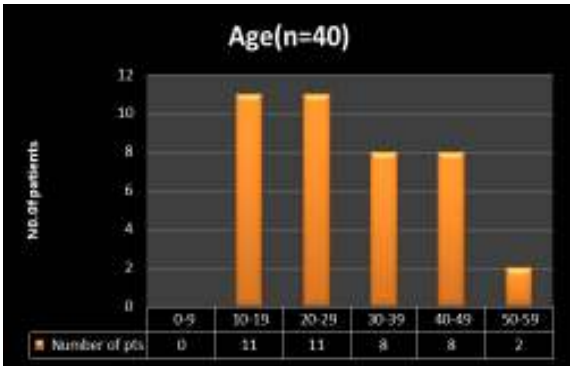

Figure 1:

Distribution of sex: There were 24 Female and 16 male patients, Female to male ratio is1.5

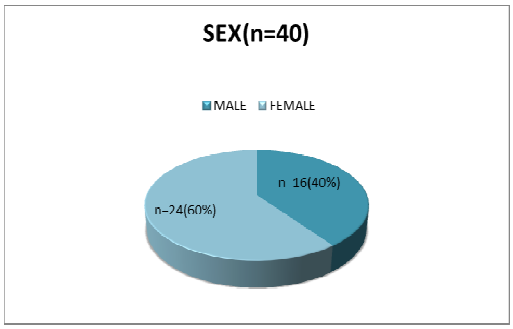

Figure 2:

Visual Symptoms: Visual impairment either at the time of presentation, or in the remote past with Visual acuity ranging from 6/18 to total loss of vision was seen in 11/40 (27.5%) of patients

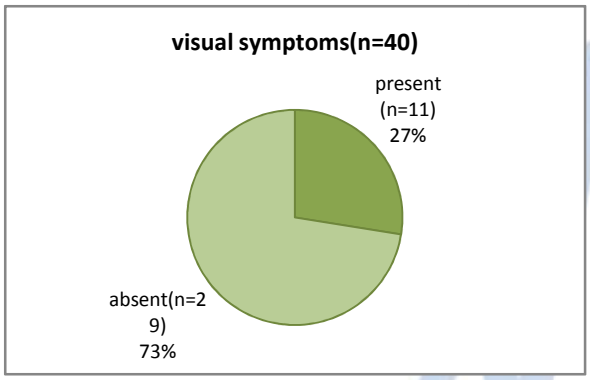

Figure 3:

Aquaporin-4 antibodies: We performed AQP-4 antibodies in 23 NMO spectrum disorder patients. AQP-4 was positive in 8/23 (34.79%) and negative in 14/23(65.21%) of patients.

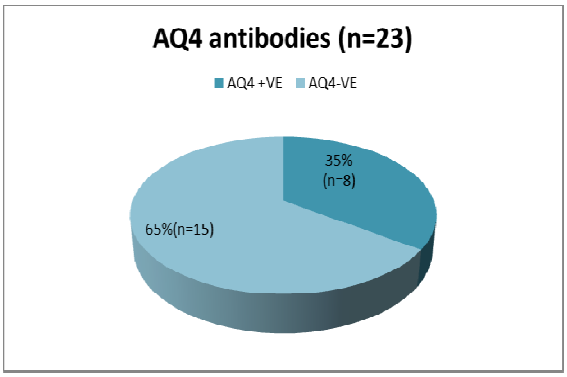

Figure 4:

Vertebral Segments: LETM was seen in 40/40 (100%) patients. Very long LETM (> 10 vertebral segments) was seen in 20/40 (50%). Mean number of vertebral segments was 9.90

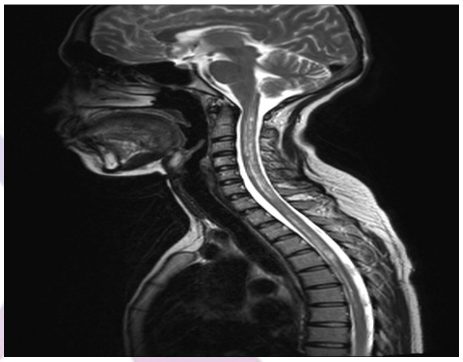

Figure 5:

Extent of lesion: Extent of lesion was in cervico dorsal in 26/40 (65%), cervical only in 9/40 (22.5%) and dorsal region only in 5/40 (12.5%) of patients.

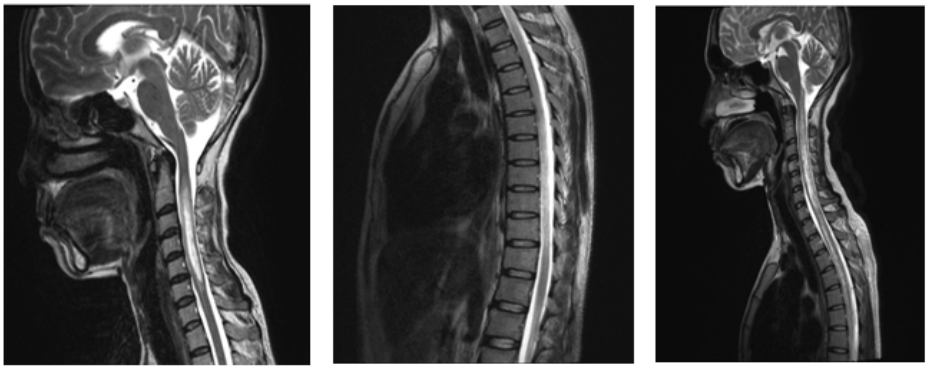

Figure 6:

Character of lesion: 38 patients had confluent spinal cord lesions; 2 patients had patchy lesions.

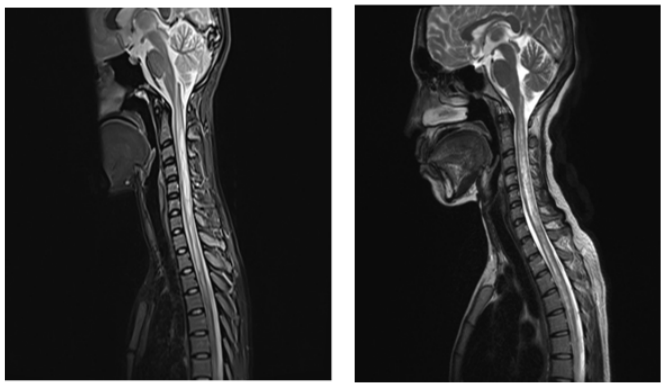

Figure 7:

Axial Cord Diameter: All patients had greater than 2/3rd axial spinal cord diameter involvement – Holocord involvement

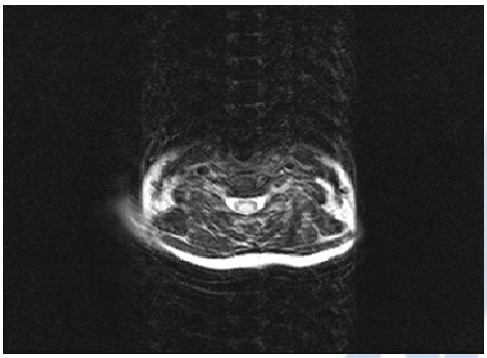

Figure 8:

Contrast Enhancement: Mild to moderate contrast enhancement of lesions was present in 38/40(95%).

Brain Involvement: None of the patients had typical brain lesions of MS, however typical brain abnormalities of NMO were found in 2/40 patients. One patient had abnormal T2W and FLAIR peri aqeductal hyper intensities on MRI brain.

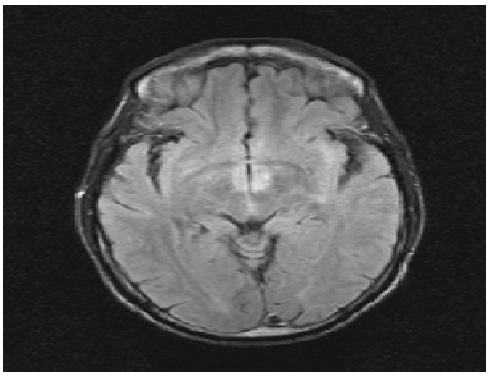

Figure 9:

Other patient had extensive T2W hyperintensity seen from medulla oblongata to D7 vertebral body level.

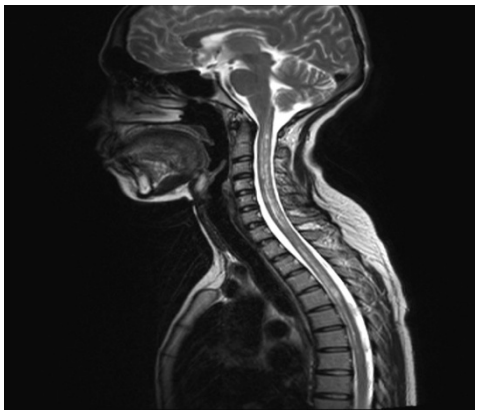

Figure 10:

DISCUSSION

In our study mean number of involved vertebral segments is 9.90. This finding is comparable to australian study by QIU[17] where the mean number of vertebral segments was 8.5. Most of the patients (65%, n=26/40) had cervico dorsal myelopathy. Isolated dorsal myelopathy is less common in our study. In most of the cases the lesion started from mid cervical cord and extending in to dorsal cord region. This neuro imaging finding is comparable with previous literature.

The lesions are confluent in 95% (n=38/40) patients. According to the literature the NMOSD spinal lesions are mainly confluent; patchy only when MRI done too early or after two months. Contrast enhancement was present in 95 %(n=38/40) patients. Enhancement was absent in two patients. On axial sections holocord involvement was present in all patients. This is comparable with those described in the literature. MRI brain was abnormal only in 2 patients. One patient had T2W and FLAIR bilateral peri aqueductal hyper intensity and other patient had extensive T2W hyperintensity seen from medulla oblongata to D7 vertebral body level

CONCLUSION

MRI is the most important and highly sensitive investigation in the diagnosis of longitudinally extensive transverse myelitis. MRI also helps in differentiating from other conditions like transverse myelitis and syrinx.

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