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Spectrum of spinal cord demyelination: A retrospective study in a tertiary care hospital of eastern India

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ABSTRACT

Background: The Idiopathic inflammatory demyelinating diseases of Central nervous system (IIDD) are common neurological disorders in neurology indoor. CNS demyelinations which affect brain, spinal cord and optic nerve are important neurological disorder in view of multi axial involvement and frequent relapse and causing significant morbidity.

Materials and Methods : It is a retrospective study done in a tertiary care hospital in eastern India from Aug2018-jul2019 (one year). Patients presenting with paraplegia or paraparesis, quadriplegia or quadriparesis with MRI showing features of demyelination were included in the study. Different criteria were taken for diagnosis to know the etiology of disease (mentioned in the introduction). Statistical Analysis Done Using SPSS Software Version-21.

Result: Among all spinal cord demyelination longitudinally extended transverse myelitis (LETM) was more common than short segment demyelination. In our study >40yrs group was most commonly affected with more female preponderance. Serum AQP4-Ab was positive in 28% of LETM patients. EDSS was higher in LETM patients than non LETM patients.

Conclusion: Among all spinal cord demyelination LETM was more common than short segment demyelination. Optic neuritis and multiphasic course was more associated in LETM with AQP4 positive NMOSD. Tuberculosis is a cause of LETM. Therefore, patients presenting with LETM require a thorough investigative workup to exclude treatable causes like infectious, inflammatory or metabolic.

Key message: Spinal cord demyelination possess a wide range with multiple etiologies. Disease course, treatment and prognosis varies in different conditions. All spinal cord demyelination should be evaluated in detail to prevent frequent relapse and morbidity As spinal cord biopsy is an invasive procedure with significant morbidity MRI lesion location, length and enhancement pattern will help to narrow the possibilities.

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1. Introduction

Demyelinating diseases are common neurological disorders that affect the central and peripheral nervous system. CNS demyelination which affects brain, spinal cord and optic nerve is an important neurological condition in view of multi axial involvement and frequent relapse which causes significant morbidity. The three main categories in the differential diagnosis of acute myelitis are demyelination, including multiple sclerosis (MS), neuromyelitis optica (NMO), MOG antibody disease and idiopathic transverse myelitis; infections such as herpes zoster and herpes simplex virus; and other inflammatory disorders such as systemic lupus erythematosus (SLE) and neurosarcoidosis. However, whether the cause of the acute myelopathy is inflammatory or not is not self-evident; therefore, the clinical and diagnostic workup for myelitis is required.[1] Magnetic resonance imaging (MRI) has become a critically important tool in diagnosis and differentiation of different demyelinating disorders.[2]

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Disease monitoring and treatment changes are often based on the combination of clinical symptoms and neuroimaging findings. Since its initial incorporation in the 2001 McDonald's criteria and subsequent revisions, it has been an integral part for diagnosis and treatment monitoring MRI also has been incorporated as well into diagnostic criteria, including neuromyelitis optica (NMO), transverse myelitis, pediatric MS, MOG Ab disease and acute disseminated encephalomyelitis (ADEM).[3–8] The objectives of this study was to know various MRI spectrums of acute myelitis and correlate them clinically along with serological spectrum with prognostication.

2. Materials And Methods

Retrospective study of 1 calendar year from Aug2018-jul 2019 in a tertiary care hospital in eastern India. Patients presenting with paraplegia or paraparesis, quadriplegia or quadriparesis with MRI showing features of demyelination were included in the study. Those with space occupying lesions, compression or trauma were excluded. They were evaluated in detail in terms of gender, age of presentation, preceding history of infection, vaccination, and various clinical presentation. All data were tabulated in a prestructured format. MRI was done in 1.5 Tesla machine with consultations from expert radiologist. All cord demyelination cases had to undergo brain MRI and optic nerve study to look for other associated sites of demyelination. Different criteria were taken for diagnosis to know the etiology of disease (mentioned in the introduction). Statisticalanalysis was done using SPSS Software Version-21.

3. Results

Total numbers of cases included in the series were 20. The most common age group of presentation was >40years of age, which was 75%. There was definite female predominance (65%) with F: M ratio of 1.8:1. Bladder and bowel dysfunction, paraplegia/paresis, quadriplegia/paresis were the most common presenting symptoms. Optic neuritis was present in 20 % of patients. In our series cervical cord was affected mostly followed by thoracic and cervico-thoracic segment (Figure 1). Holocord involvement was seen only in 5% of cases (Figure 2). In our series among all 20 patients 70% were having longitudinally extended transverse myelitis (LETM) and 30% were non-LETM. 3 to 5 segment involvement was more common in 50% cases followed by non LETM (<3 segments) involvement in 25% of cases followed by 6-10 segments among all spinal cord demyelination patients. 60% patients had abnormal brain lesions.

55% patients of spinal cord demyelination were clinically diagnosed as NMOSD according to revised Wingerchuk criteria. 10% patients were diagnosed as post infectious myelitis followed by MOG Antibody disease (10%), multiple sclerosis (5%), seronegative acute demyelinating encephalomyelitis (5%) tuberculous myelitis(5%). No cause could be determined in 10% patients (table-1). Among all LETM patients of (14 out of 20) 28% cases were positive for serum AQP4 antibody. 10% were positive for MOG antibody. In our series most patients suffered a monophasic course and only 20 % cases were having relapse. These recurrent episodes were mostly seen in AQP4 positive LETM. The Expanded disability status scale (EDSS) at one year follow up showed severe disability in NMO patients. Poor prognosis in form of higher EDSS (6±0.2) was found in AQP4-Ab positive LETM patients as compared to AQP4-Ab negative LETM patients (5±0.9). MOG Antibody disease, multiple sclerosis, ADEM and idiopathic LETM patients had comparatively better prognosis at one year.

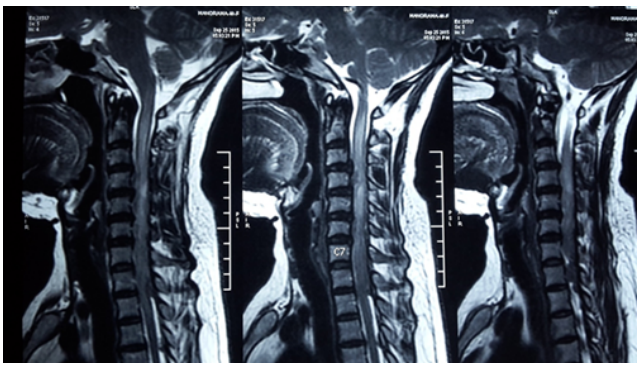

Fig. 1: (MRI Picture of a LETM)

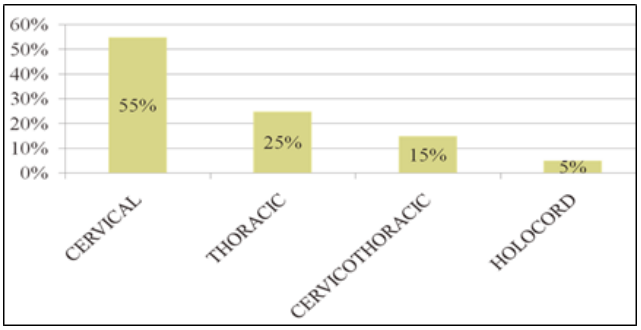

Fig. 2: (Segment in MRI)

4. Discussions

The most common age group of presentation was >40years of age, which was 75%.In a study by R.S. Jain et alfrom S.M.S. Jaipur, 59.37% of patients with LETM were young less than 30 years.[9] Slightly higher age of presentation may be due to small sample size and economically different layer of people admitted at corporate hospital. There was definite

Table 1: (Diagnosis)

Disease	Numbers of cases (%)
NMOSD	11 (55%)
Multiple sclerosis	1 (5%)
MOG Ab disease	2 (10%)
Postinfectious myelitis	2 (10%)
Tuberculous myelitis	1 (5%)
ADEM	1 (5%)
Idiopathic	2 (10%)

female predominance (65%) with F: M ratio of 1.8:1 which was concordance with many studies from different parts of world.[9] Bladder and bowel dysfunction, paraplegia/paresis, quadriplegia/paresis were the most common presenting symptoms in our series. Similar results were observed in several studies.[9,10] In our series among all 20 patients 70 % were LETM and 30% were non-LETM. In several studies and review literatures, the longitudinally extended transverse myelitis (LETM) is a predominant entity. It's a common feature in most of the demyelinating diseases.[6–9] We also coincide with the fact. As our cohort consists of NMOSD predominantly the involvement of cervical cord is more. In many studies it's apparent that the incidence of cervical cord involvement is definitely high in NMOSD in comparison to MOG Ab disease.[6] In our series, demyelinating disorders were the most common cause of LETM, in which NMOSD was the commonest etiology. Aquaporin-4 IgG antibody is very specific for neuromyelitis optica. Among all LETM patients of (14 out of 20) 28% cases were positive for serum AQP4 antibody. Other cases (7) were seronegative NMOSD. MOG Ab disease is a new spectrum of demyelinating disease. In our study 2 patients were included in the spectrum. It fitted into the criteria for MOG Ab disease.[11] In our series most cord demyelinations had a monophasic course and only 20% cases were having recurrent demyelination. These recurrent demyelinations mostly seen in AQP4 positive LETM. The Expanded disability status scale (EDSS) at one year follow up showed severe disability in NMO patients in comparison to other etiologies. In many studies we have found similar presentation. Prognostically NMOSD is a worse disease in comparison to ADEM and MOG Ab disease.[6]

5. Conclusion

Our study is one of the largest case series from single centre in a single year.Among all spinal cord demyelination LETM is more common than short segment demyelination.It's a disease of young people with female predominance.Cervical spinal cord segments were most commonly involved. NMOSD was the most common etiology**.** Serum AQP4-Ab was positive in 28% of LETM patients. They had more recurrences with poor prognosis in comparison to other groups. Optic neuritis and multiphasic course was more associated in LETM with AQP4 positive status. Therefore, patients presenting with LETM require a thorough investigative workup to exclude treatable causes like infectious, inflammatory or metabolic. Finding out exact diagnosis is very important in LETM as management depends on distinguishing inflammatory from noninflammatory causes.

6. Source of Funding

None.

7. Conflict of Interest

None.

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