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Clinicoetiological and Radiological Correlation in Patients Presenting with Noncompressive Myelopathy

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Abstract: ***Background: Non - compressive myelopathy encompasses a large range of disease entities ranging from demyelination, infection, nutritional, toxic, hereditary - familial to degenerative conditions. With the advent of MRI which is a very sensitive modality for the intramedullary spinal lesions it has become pertinent to have a relook at the profile of non - compressive myelopathies in India. In this paper we want to present our experience with various non - compressive myelopathies and its radiological correlation. Aim: To study the clinical spectrum of non - compressive myelopathy and clinicoradiological correlation Objective: To study the clinical spectrum of patients with non - compressive myelopathy. Duration: 1 year. Methodology: Prospective observational study of patients in outpatient department of neurology dept GMKMCH Salem Results: Out of 30 patients with non - compressive myelopathy acute transverse myelitis was the commonest (15) followed by vitamin B12 defy (4) followed by hereditary spastic paraplegia (3) paraneoplastic myelopathy (2) HIV myelopathy (1). No etiology can be found out in remaining 5 cases. The most common abnormality detected in MRI was hyperintense lesions that occupy the central core. CSF done in ATM done showed rise in proteins with minimal pleocytosis. Conclusion: Acute transverse myelitis is the leading cause of non - compressive myelopathy. Clinical features combined with MRI findings are helpful in defining the cause of ATM.***

Keywords: Non compressive myelopathy, Acute transverse myelitis, NMOSD (neuromyelitis Optica Spectrum Disorder), AQP4 antibody

1. Introduction

Non compressive myelopathy includes a large range of disease entities ranging from demyelination, infections, nutritional, toxic, hereditofamilial and degenerative conditions. They are clinically characterized by patterns of selective involvement of different anatomical structures of spinal cord and these patterns helps in clinical diagnosis.

The disease spectrum is somewhat different in India as compared to western countries where infectious and nutritional causes are less common and demyelinating and familial causes are the leading causes. Despite an extensive work up, etiological diagnosis cannot be established in many cases. Close follow up of these patients are required in some cases where actual diagnosis can be revealed in due course of time.

2. Aims and Objectives

To study the clinical, etiological and radiological spectrum of patients with Non compressive myelopathy in tertiary care hospital in south India.

3. Review of Literature

Non compressive myelopathies are clinically characterized by patterns of selective involvement of different anatomical structures of the spinal cord and these patterns help the etiological diagnosis. Some of the classical syndromes with their commonest causes are as follows. Complete spinal cord syndrome (e. g. transverse myelitis), Brown Sequard syndrome (e. g. multiple sclerosis), anterior spinal cord syndrome (eg. anterior spinal artery infarct), posterolateral cord syndrome (e. g. vitamin B12 deficiency), central cord syndrome (e. g. neuromyelitis optica), posterior syndrome (eg. posterior spinal artery infarct and tractopathies (e. g. primary lateral sclerosis). Causative factors of non - compressive myelopathies could be broadly grouped in inflammatory and non - inflammatory causes. In the inflammatory group, transverse myelitis is an important and a common cause. The other causes are infectious, autoimmune, nutritional and vasculitic diseases. The non - inflammatory groups are of vascular, toxins and physical agents, degenerative, paraneoplastic, metabolic and inherited myelopathies. In various studies on non - compressive myelopathies, the etiological spectra have varied according to populations studied and also in the time frame. In the more recent studies, the numbers of idiopathic cases are decreasing with discovery of new tests and better resolution of neuroimaging. In the western literature, demyelinations and

immune causes are encountered more often as compared to the Asian literature but this trend is changing with better diagnostic facilities being available for conditions like neuromyelitis optica (NMO).

4. Materials and Methods

This study was an observational prospective study conducted in the dept of Neurology in Govt Mohan Kumaramangalam Medical College Hospital, Salem over a duration of 1 year. Non - compressive myelopathy patients admitted in neurology department were studied. They were examined clinically and followed up by laboratory investigations and neuroimaging studies. Details of onset of illness, antecedent events, symmetry/asymmetry of symptoms, family history, type of treatment given and complications were particularly recorded. MRI spinal cord for all patients and MRI brain for required patients were done. Estimation of CSF Analysis including OCB bands, Rheumatoid factor, ANA level, AQP4 Antibodies and vit B12 level were assessed

Inclusion Criteria

Patients presenting with Non compressive myelopathy diagnosed by clinical and radiological evaluation were included in the study.

Exclusion Criteria:

- Degenerative disc diseases
- Abnormalities such as Tumour, cyst and hematomas
- Spinal injury, Potts spine
- Radiation injury, Trauma

5. Results

Out of 30 patients included in the study 18 were females and 12 were males. Mean age at presentation was 40 years (18 - 65). Demyelinating disorders form the major etiology attributing to 50 % of the study population (15 patients) followed by vitamin B12 deficiency (4), hereditary spastic paraparesis (3), paraneoplastic myelopathy associated with RCC (1), HIV myelopathy (1), and post infectious myelitis (1). In other 5 patients no etiology can be made out (Two patients had probable ASA infarction)

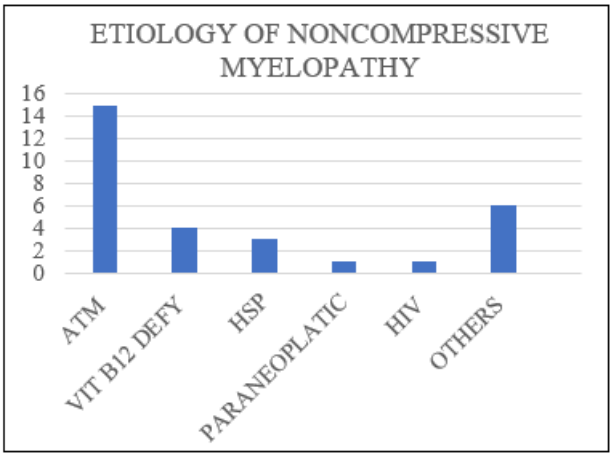

Chart 1

Among 15 patients with demyelinating lesion, 9 were females and 6 were males and mean age group was 35.4 (18 - 55yr).

For all patients with MRI Brain & Spinal cord, CSF analysis, VEP and required immunological profile has been studied. Among these 15 patients 9 patients had anti AQP4 AB positivity, 1 patient had CSF oliogoclonal band positivity, 2 patients fulfilled MS McDonald criteria, 1 patient satisfied with ADEM clinical criteria, and in 2 patients no etiology can be found out (Idiopathic Acute transverse myelitis). Among 9 patients with AQP4 positive, 6 patients had relapse out of which 3 patients had multiple relapses.4 patients had optic neuritis (2 unilateral +2 bilateral) with permanent vision loss in three of them.5 patients had involvement of complete transverse extent of spinal cord, 3 had central cord involvement and one patient had patchy involvement of spinal cord.

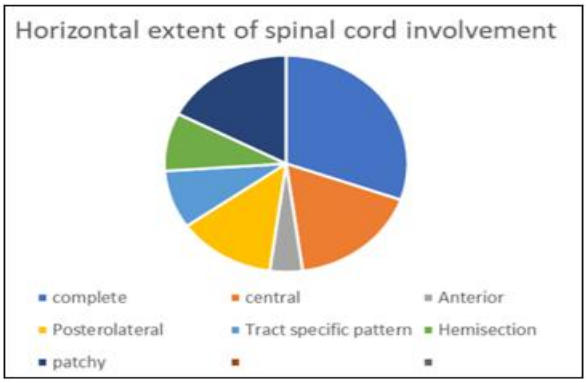

Chart 2

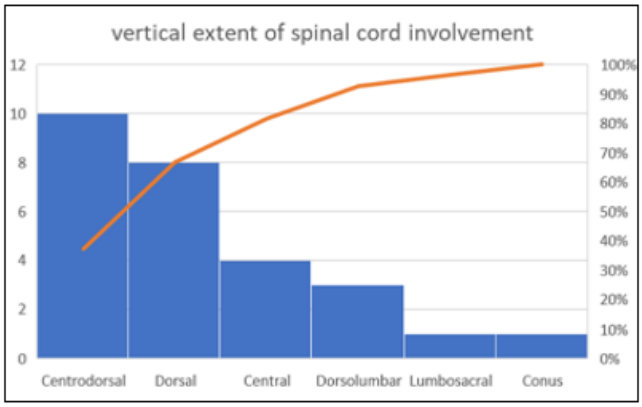

Chart 3

5 patients had extensive involvement of spinal cord from cervical to dorsal, 3 had exclusively dorsal involvement and one had cervical involvement. VEP was done in all patients, in this 6 out of 9 patients showed abnormality (prolonged P100 latency) 4 bilateral and 2 unilateral. Associated MRI brain lesions were found in 4 out of 9 patients. Most common site of lesion lower Brainstem. CSF analysis has been done in all patients, 4 had elevated protein with mild pleocytosis. one had normal protein and cell count. one patient with elevated protein with normal cell count. One antenatal patient (22 weeks) presented with quadriparesis with altered sensorium following an episode of fever. MRI brain and whole spine showed bilateral frontoparietal subcortical /deep white matter lesion with cervical spinal cord demyelination suggestive of ADEM. In patients with vit B12 deficiency (4), NCS showed Sensorimotor axonal peripheral neuropathy in 2 patients and pure sensory axonal neuropathy in 1 patient and UGI endoscopy showed atrophic gastritis in 2 out of 4 patient. In Patients with HSP (3) significant family history was not present and one patient belonged to complicated HSP

category. In patient with paraneoplastic myelopathy, associated with RCC, radical nephrectomy improved the clinical deficit.

6. Discussion

Non compressive myelopathies comprise a wide spectrum of disorders including infectious, demyelinating, nutritional, paraneoplastic and others. The prognosis and therapeutic outcome vary among these etiologies**.** Mean age at presentation was 40 years (18 - 65). Prabhakar et al[1] reported mean age at presentation to be 34.45 years ranging from 14 - 82 years. whereas Shabeer et al[2] reported cases ranged from 2 - 55 years with median age of 32.35 years.

In this study demyelinating disorders form the major etiology attributing to 50 % of the study population (15 patients) followed by vitamin B12 deficiency (4), hereditary spastic paraparesis (3), paraneoplastic myelopathy associated with RCC (1), HIV myelopathy (1), and post infectious myelitis (1). In other 5 patients no etiology can be made out. Two patients had probable ASA infarction. These results are consistent with similar studies carried out in India. Complete cord involvement were seen in majority of patients followed by central cord, anterior cord involvement in 2 patients, tract specific pattern in 3 patients. Among 15 patients with demyelinating lesions, 9 were females and 6 were males and the mean age group was 35.4 (18 - 55yr). For all patients with MRI Brain & Spinal cord, CSF analysis, VEP and required immunological profile has been studied. Among this 15 patients 9 patients had anti AQP4 AB positivity, 1 patient had CSF oliogoclonal band positivity, 2 patients fulfilled MS McDonald criteria, 1 patient satisfied with ADEM clinical criteria, and in 2 patients no etiology can be found out (Idiopathic Acute transverse myelitis). In Shabeer et al[2] NMOSD was the most common diagnosis seen in 35% followed by ITM - Idiopathic transverse myelitis in 20% and multiple sclerosis in 15%. Infectious myelitis seen in 10% 1 zoster myelitis and 1 case of scrub typhus associated myelitis. other diagnosis of ADEM in 7.5%, HSP Vit B12 deficiency, Copper deficiency, SLE and undefined etiology in 2.5 %. A study by Li et al[3] in Chinese population revealed NMOSD to be the cause in 40.3%, MS in 16.4% and ITM in 43.3%. Studies from African countries like Ethiopia and Niger have reported low prevalence of demyelinating disorder as a cause of noncompressive myelopathy. Indian studies, however reported higher incidence of NMOSD compared to MS and other etiologies. Mishra et al[4] reported an incidence of 55% of NMOSD, 10% for MOG associated disease and 5% for MS while studying demyelinating disorders of spinal cord at tertiary care centre. In our study among demyelinating group , 5 patients had involvement of complete transverse extent of spinal cord, 3 had central cord involvement and one patient had patchy involvement of spinal cord.5 patient had extensive involvement of spinal cord from cervical to dorsal, 3 had exclusively dorsal involvement and one had cervical involvement. This is not consistent with study done by Shabeer et al[2] showed that dorsal cord involvement was most common site of involvement in demyelinating group with an incidence of 38.7% followed by cervical 32.2% and cervicodorsal 19.35%. VEP done in all patients, in this 6 out of 9 patients showed abnormality (prolonged P100 latency) 4 bilateral and 2 unilateral. Associated MRI brain lesions was found in 4 out of 9 patients most common site of lesion lower Brainstem. CSF analysis has been done in all patients, 4 had elevated protein with mild pleocytosis. one had normal protein and cell count. one patient with elevated protein with normal cell count. One antenatal patient (22 weeks) presented with quadriparesis with altered sensorium following an episode of fever. MRI brain and whole spine showed bilateral frontoparietal subcortical /deep white matter lesion with cervical spinal cord demyelination suggestive of ADEM. In patients with vit B12 deficiency (4), NCS showed Sensorimotor axonal peripheral neuropathy in 2 patients and pure sensory axonal neuropathy in 1 patient and UGI endoscopy showed atrophic gastritis in 2 out of 4 patient. In Patients with HSP (3) significant family history was not present and one patient belonged to complicated HSP category. In patient with paraneoplastic myelopathy, associated with RCC, radical nephrectomy improved the clinical deficit.

7. Summary and Conclusion

- Significant proportion of patients (50%) have demyelination of spinal cord among Noncompressive myelopathy.
- NMOSD forms majority in demyelinating disease of spinal cord in comparison to MS and ADEM.
- Relapses are common in NMOSD, so timely diagnosis and appropriate management should be initiated at the earliest
- MOG ab positive myelitis were uncommon in adults.
- Also we have to suspect other etiologies like Nutritional, vascular, HIV, Infectious, autoimmune and paraneoplastic etiologies along with demyelination
- Definitive etiology could not be ascertained in one sixth of cases

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