

Non Compressive Myelopathies

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Introduction

Medical myelopathies can lead to disabilities and reduction of quality of life rapidly and profoundly, often in the prime time of life. Hence the physician has to be up to date with the current knowledge of these disorders.

Various studies on non-compressive myelopathies are available from India. Those undertaken before the MRI era^{1,2} have discussed nutritional and infectious causes. Investigative facilities have now improved in India and newer studies are needed to redefine the profile of various aetiologies of

non-compressive myelopathies in the Indian setting. The manuscript entitled "Etiological spectrum of non-compressive myelopathies in a tertiary care centre" by Kamble et al³ in this issue of JAPI is timely in this regard.

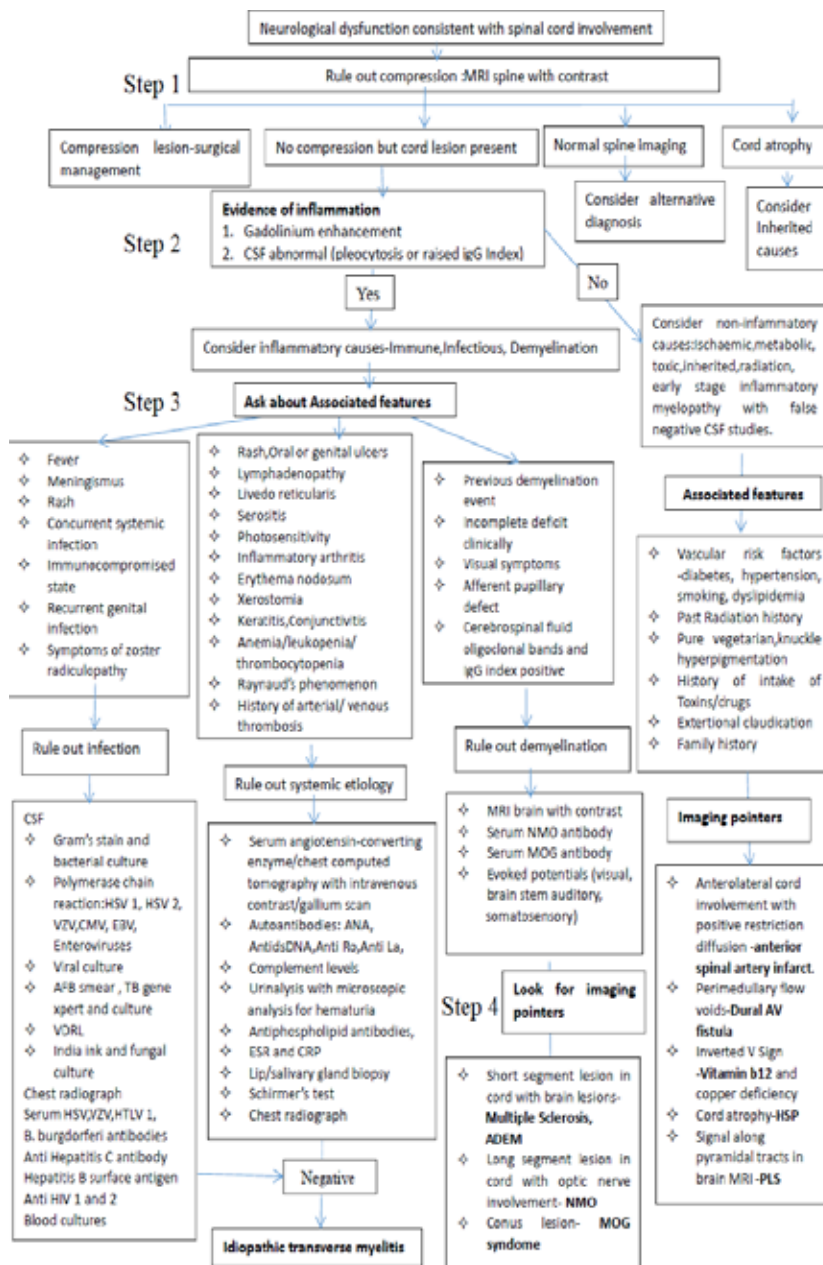
Clinical Syndromes

Non compressive myelopathies are clinically characterised 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 (eg. transverse myelitis), Brown Sequard syndrome (eg. multiple sclerosis), anterior spinal cord syndrome (eg. anterior spinal artery infarct), posterolateral cord syndrome (eg. vitamin B12 deficiency), central cord syndrome (eg. neuromyelitis optica), posterior syndrome (eg. posterior spinal artery infarct and tractopathies (eg. primary lateral sclerosis).

Aetiology

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.⁴ The other causes are infectious, demyelinating and vasculitic diseases. The non-inflammatory groups are of vascular, toxins and physical agents, degenerative, 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,



Algorithm 1: Approach to non-compressive myelopathies

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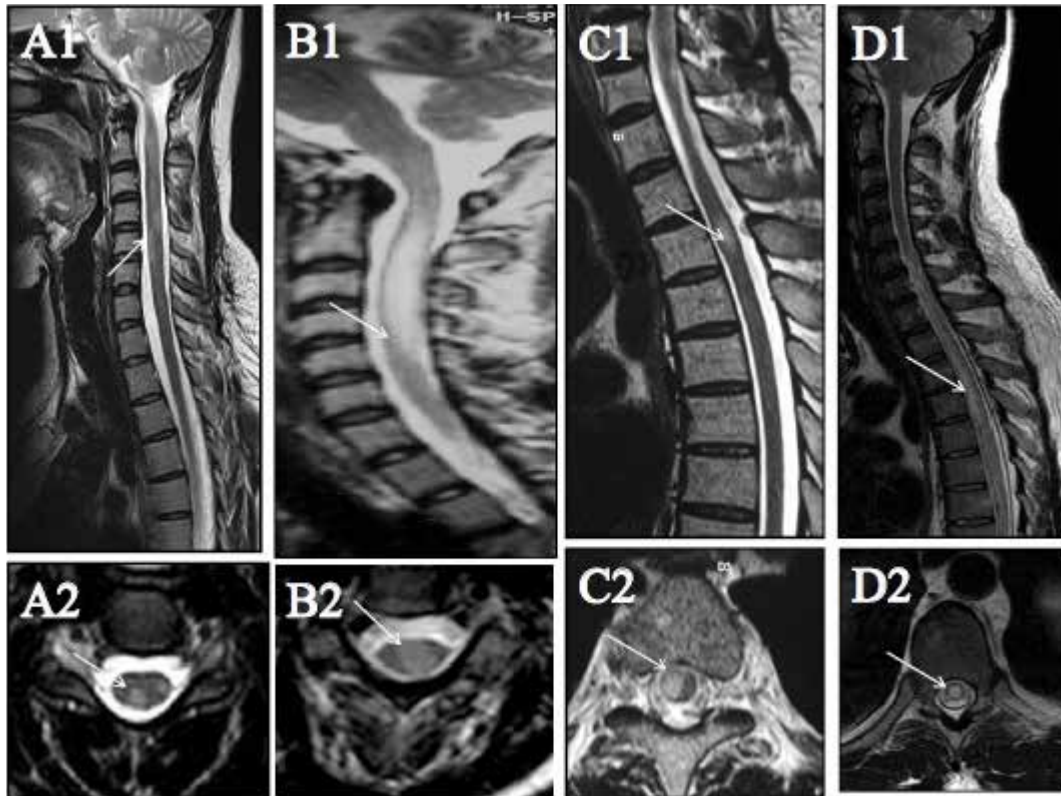


Fig. 1: Distinctive spinal MRI imaging findings: Inflammatory myelopathies. Multiple sclerosis (A1-A2)--Sagittal and axial T2 weighted image showing short segment lesion in the spinal cord located peripherally; Neuromyelitis optica (B1-B2)--Sagittal and axial T2 weighted image showing long segment lesion (>3 vertebral segments) in the spinal cord located centrally; Herpes zoster myelitis (C1-C2)--Sagittal and axial T2 weighted image showing short segment lesion in the spinal cord placed laterally; Tuberculous myelitis (D1-D2)--Sagittal and axial T2 weighted image showing long segment lesion in the spinal cord located centrally.

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).

In this context, the study by Kamble et al³ in this issue of the journal is interesting. This investigation included 80 patients of non-compressive myelopathies, out of which 44 had acute-subacute and 36 had chronic myelopathies. Etiological spectrum of acute to sub-acute myelopathies suggested post infectious myelitis to be the most common, followed by NMO spectrum disorders (NMOSD) and Multiple sclerosis (MS) where as in chronic myelopathies, vitamin B12 deficiency was most often seen. A noteworthy finding in this study was of the large number of cases of post-infectious myelitis due to dengue virus. Spinal cord involvement in dengue is uncommonly reported.⁵ Hence these cases of dengue myelitis are of interest and it will be useful to compare these with accumulating experience from regions of India where

dengue is frequent. In a recent study of 151 patients from Northeast India, commonest cause of acute myelopathy was NMOSD which suggest incidence of various etiologies varies in different regions of India.⁶ A multicentric study from France comprising of 288 patients of non-compressive myelopathy, the common categories were NMO, MS, and para infectious demyelinations. No aetiology was found in 15.6%.⁷ Compared to that, the series from Kota documented 48% idiopathic cases, a much larger percentage, which is most likely a reflection of the limited access to tests and financial constraints.

Diagnostic Evaluation

Evaluation of a patient with symptoms of myelopathy requires a comprehensive approach as the list of causes is huge. An algorithmic approach could be utilized (Algorithm 1). The first step is to perform an MRI of the spine to rule out a compressive aetiology which may need urgent intervention. Once compression is excluded, the next step is to confirm the evidence of inflammation by MRI spine contrast study and cerebrospinal

fluid (CSF) to differentiate between inflammatory and non-inflammatory aetiologies, as their treatment modalities are different. If there is evidence of inflammation, the next step is to search for the cause of inflammation as it can be due to demyelination, infectious and secondary to systemic immune conditions. Hence, further inquiry into history should be undertaken as mentioned in the algorithm before proceeding further and rest of the workup can be planned according to associated features pointing to demyelination, infections and systemic immune conditions. When no evidence of inflammation exists, consider non inflammatory aetiologies like vascular, radiation, metabolic, inherited etc. Imaging features should be carefully studied and these may point to some specific aetiologies and aid in planning further work up.

Role of Neuroimaging

Imaging has now become a vital and integral part of the work up of non-compressive myelopathies. The modalities include magnetic resonance imaging, digital subtraction

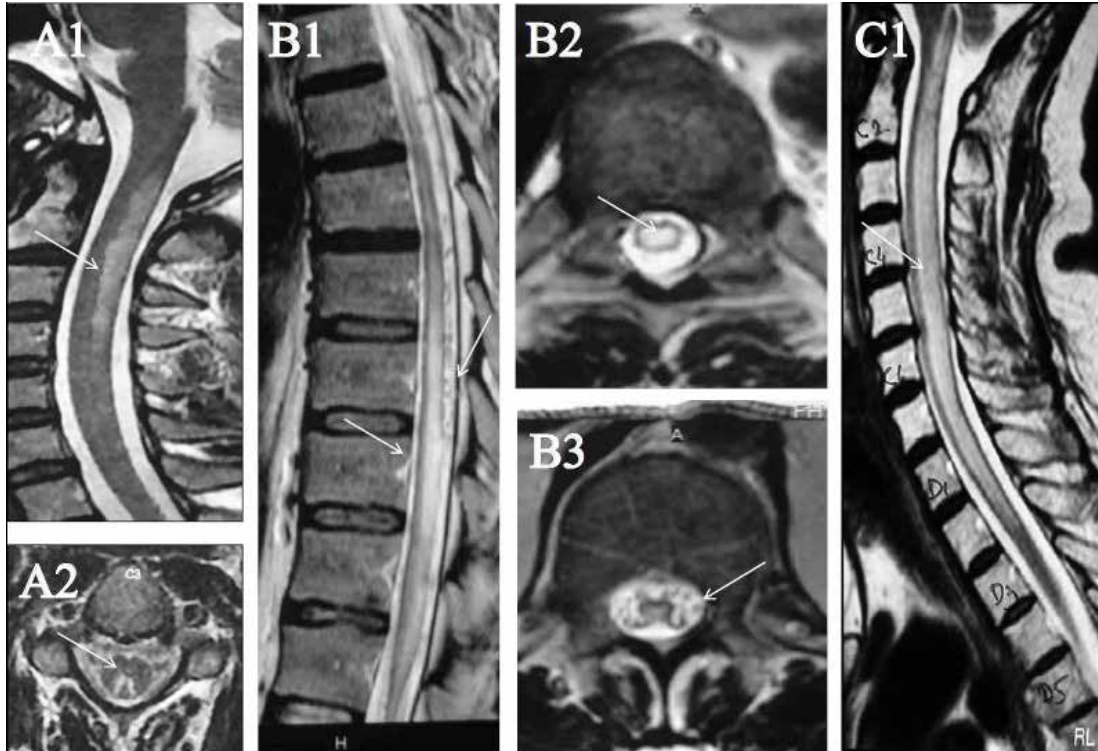


Fig. 2: Distinctive spinal MRI imaging findings: Non-inflammatory myelopathies. Vitamin B12 deficiency (A1-A2)-Sagittal and axial T2 weighted image showing long segment lesion in the spinal cord located in dorsal columns giving inverted V sign on axial image; Spinal Dural arterio-venous fistula (B1-B3)-Sagittal and axial T2 weighted image showing long segment lesion in the spinal cord located centrally with perimedullary flow voids; Post Radiation myelitis (C1)-Sagittal T2 weighted image showing long segment lesion in the spinal cord located centrally

angiography and sometimes the computerized myelography. Various specific imaging characteristics of cord lesions have been documented in different conditions which help in narrowing the diagnostic possibilities and at times give an accurate diagnosis as mentioned in Algorithm 1. In addition, persistent enhancement of cord lesion for more than 2 months with dorsal subpial and central canal enhancement (Trident sign) is seen in neurosarcoidosis.⁸ 'Bagel sign' a central hyper intense cord lesion on T2 axial with enhancement and a hypo intense centre suggests Behçet's disease.⁹ Brain MRI should be done when suspicion of MS exists and patients have short segment lesion in spinal cord. Figures 1 and 2 depict the characteristic MRI findings in non-compressive myelopathies.

In the study by Kamble et al³ in this issue, the MRI spine showed long segment lesions in 14 and short segment lesions in 18 patients. Most common aetiology for long segment lesions was NMOSD and for short segment lesions infectious/postinfectious myelitis. Previous Studies of NMOSD from India have shown similar results.^{10,11}

Surprisingly, Kamble and colleagues report that both their multiple sclerosis patients had long segment lesions. This is distinctly unusual and calls for more diagnostic scrutiny of such patients.

Conclusions

As can be seen from the above discussion, a wide variety of diseases present with non-compressive myelopathies. A detailed history and clinical examination coupled with the appropriate work up can lead to the final diagnosis in majority of cases. Neuroradiology and serological markers have increased our diagnostic accuracy. The study by Kamble et al is important as it provides the current spectrum of non-compressive myelopathies in Kota region. More studies are needed to define the pattern of existing and newly recognised conditions like the Myelin oligodendrocyte antibody (MOG) syndrome in India. Regional as well as multicentric national studies are clearly required to elucidate the prevalence of various aetiological subsets in India.

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