A Study on Etiological Profile of Non-Compressive Myelopathies in a Tertiary Care Hospital in Central Tamilnadu

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ABSTRACT

Introduction: Non compressive myelopathy is defined as "spinal cord dysfunction in the absence of clinico-radiological evidence of spinal cord compression." It can result from demyelinating, infectious, autoimmune, vascular, degenerative and metabolic disorders in the absence of demonstrable compression by imaging techniques. We aimed in analyzing the etiological profile of non-compressive myelopathies in a tertiary care hospital of Central Tamil nadu.

Material and Methods: In the Neurology department, we conducted an observational study at Thanjavur Medical College, Thanjavur, from September 2017 to September 2018. Patients of non-compressive myelopathies who underwent magnetic resonance imaging (MRI) of the spine were segregated into two categories: Degenerative and non-Degenerative, as well as into acute, subacute and chronic myelopathies.

Results: The study had 75 patients with a median age of 34.5 years and male: female ratio of 1.35:1. Presentation was acute in 10 patients (13%), subacute in 5 (6.5%), chronic in 54 (72.5%) and history of relapse and remission in 6(8%) patients. Degenerative etiology was found for 42 (56%) others were non degenerative (demyelinating, autoimmune, vascular, nutritional, or physical agent). MRI study carried out in all cases showed signal changes in 51 cases (68%) which included myelomalacia, demyelination, atrophy of cord, infarction of cord. Etiological diagnosis could be established in 74 (97.3%) cases.

Conclusion: Underlying etiology (degenerative, demyelinating, autoimmune, infectious, vascular, metabolic disorder, or physical agent) was found in 91.3% patients of noncompressive myelopathy. Clinical features combined with MRI findings are helpful in defining the cause of non-compressive myelopathies. A follow-up of long term may reveal some of the diagnosis especially degenerative myelopathies in early stage.

Keywords: Non Compressive Myelopathy Central Tamilnadu Degenerative Paraparesis

INTRODUCTION

Term "compressive and noncompressive myelopathy (NCM)" was coined by Siccard and Frostier in 1921 based on myelographic clue. NCM was defined as "spinal cord dysfunction in the absence of clinico-radiological evidence of spinal cord compression." Since then there was significant advancement on the study of spinal cord diseases. It is now possible to rule out even small compressive lesions subsequent to the discovery of computerized tomogram (CT) and magnetic resonance imaging (MRI). Spinal cord disorders deliver an ample disability to the patient, and hardship to the family members and to society. Myelopathies may result from infectious, demyelinating, vascular, autoimmune, and metabolic disorders. A compressive lesion should be ruled out by MR imaging. A comprehensive data on Indian patients with non-compressive myelopathy in the light of newer diagnostic criteria and serological tests are lacking. No study from central Tamilnadu has been conducted to study the clinico-radiological profile of non-compressive myelopathy so far. In this study we are putting an effort to convey our experience on etiological pattern of NCM with special reference to its radiological features.[1-3]

The aims and objectives of the study were to determine the causes of noncompressive myelopathies and to study the clinical and radiological features of noncompressive myelopathies.

MATERIAL AND METHODS

An observational study was conducted at Neurology department Thanjavur Medical College, Thanjavur, from September 2017 to September 2018, and the data were collected prospectively.

The Institutional Ethical Committee of Thanjavur Medical College approved the study protocol. Informed consent was obtained from the patients. Study was conducted on Patients with neurologic dysfunction indicative of myelopathy from the Internal Medicine units and Neurology department.

Exclusion criteria: (i) Patients who didn't underwent magnetic resonance imaging (MRI) of the spinal cord, (ii) MRI of spinal cord indicative of compressive myelopathy.

Patients were enquired about the details of illness including other neurological symptoms, constitutional symptoms like fever, weight loss etc. Skin rash, photosensitivity, joint pain, jaundice, edema, bone pain, anemia, cough, chest pain, gastrointestinal bleeding, travel history, exposure to radiation and other toxic substances, and high-risk sexual behavior were also asked for. Detailed general examinations were

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done to get evidence for systemic illness and other clinical clues. The neurological examination included evaluation of higher functions, cranial nerves, motor and sensory systems, reflexes and cerebellar sings. The level of spinal cord dysfunction was determined using American Spinal Injury Association Impairment Scale. Modified Rankin Scale (mRS) was used to assess disability.

MRI of spinal cord was done in all patients, and other relevant blood tests. Cerebrospinal fluid (CSF) analysis included cell counts, protein, and sugar. Electrophysiological studies were done in selected patients using using Recorders and Medicare Systems (RMS)–EMG/ NCV/ EP system (RMS Private Limited, India).

Case definitions

Acute-to-subacute myelopathy (ASM) was defined as myelopathy with progression over at least 48 h to 21 days of onset of symptoms.[1] Chronic myelopathy (CM) was defined as myelopathy progressing over months to years. Acute transverse myelitis (ATM) was diagnosed as per TMCWG criteria.[2] Post infectious myelitis (PIM) was considered if there was infective illness preceding onset of neurological symptoms within 30 days. Acute disseminated encephalomyelitis (ADEM) was diagnosed as per criteria proposed by International Pediatric Multiple Sclerosis Study Group.[3] 2015 International Panel for NMO Diagnosis criteria was used to diagnose NMO spectrum disorder. The diagnosis of multiple sclerosis (MS) was made according to 2010 Revised McDonald Criteria.[4] Paraneoplastic myelopathy was defined in case of malignancy with or without paraneoplastic antibody after excluding other etiologies. Infectious myelopathy (IM) was diagnosed on serology or detection of pathogen in CSF or blood and correlating clinically.

RESULTS

During the study period, 75 patients were clinically diagnosed with non-compressive myelopathy with a mean age of 34.5 (table 1) and male to female ratio of 1.35 : 1(table 2). Most of the patients belonged to lower socio economic status (table 3). The clinical features of the patients were summarized in table 4. Patients who had an alternative diagnosis after investigations and patients who did not undergo MRI of the spine were excluded from the study.

Motor neuron diseases, degenerative ataxias, hereditary spastic paraparesis etc were classified separately as degenerative myelopathies. Degenerative etiology was found for 42 (56%) others were non degenerative (demyelinating, autoimmune, vascular, nutritional, or physical agent).

Among the non - degenerative causes (33 patients), Acute Myelopathy (AM) (table 5) was diagnosed in 10 (30.30%) patients. The median age was 35 years (+/- 14.25). Male: female ratio was 1.3:1. The common causes of AM were post infectious myelitis (4; 40%), NMOSD (2; 20%) clinically definite MS (1; 10%), infectious myelitis (1; 10%), spinal cord infarct (1; 10%), electrocution (1; 10%).

Chronic Myelopathy (CM) was diagnosed in 13 (43.3%) patients. The median age was 35 years (+/- 13.5). Male: female ratio was 2.5:1. The causes of CM were vitamin B12 deficiency (5; 38%), electrocution (2; 15.3%), infections like HIV, Syphilis, HTLV (1;7%) and radiation (1; 7%).

Cord signal changes (hyperintensity on T2-weighed images) on MRI were found in 22 (66.6%) patients. Cord swelling was seen in 2 (15.5%) patients. T1 hypointense lesions were found in 2 patients of NMOSD, one patient of radiation

Age group	No.	Percentage
0 - <10	1	1.33%
10 - <20	7	9.33%
20 - <30	17	22.67%
30 - <40	13	17.33%
40 - <50	24	32.00%
50 - <60	8	10.67%
60 - <70	5	6.67%

Table-1: Age distribution

Sex	No.	Percent
Female	29	38.67%
Male	46	61.33%

Table-2: Sex distribution

Socioeconomic status	No.	Percent
Lower	45	60.00%
Middle	28	37.33%
Upper	2	2.67%

Table-3: Socio economic status

Clinical features	No.	Percent
Bibrachial weakness	5	6.85%
Hemiparesis	4	5.48%
Monoparesis	5	6.85%
Paraparesis	38	52.05%
Quadriparesis	21	28.77%
Posterior column sensory loss	20	26.66%
Spinothalamic sensory loss	18	24.44%
Pyramidal signs	61	81.33%
Spasticity	58	77.33%
Wasting	28	37.33%
Sphincter involvement	26	34.66%
Peripheral neuropathy	18	24.00%
Cerebellar signs	11	14.66%
Brainstem involvement	6	08.00%
Flexor spasms	6	08.00%
Paresthesia	4	05.33%
Encephalopathy	3	04.00%
Pigmentary retinopathy	2	02.66%
Dystonia	1	01.33%
Radiculopathy	1	01.33%

Table-4: Clinical features

Duration	Frequency	Percent
Acute	10	13.33%
Chronic	54	72.00%
Relapsing	6	8.00%
Subacute	5	6.67%

Table-5: Duration of myelopathy

myelopathy and one patient of unknown AM. Vertebral signal changes were seen in the patient with radiation myelopathy and the case of SCI. Diffusion restriction was seen in the patient with SCI and one patient of Post Infectious Myelitis. Syrinx was found in one patient with NMOSD.

Among 42 degenerative myelopathy cases, 2 cases were subacute and 40 cases were chronic. Subacute cases were due to syrinx. Among the chronic degenerative myelopathies which included hereditary non-compressive myelopathies, 13 (30.95%) cases were ALS, 12 cases were SCA (28.57%), 7 (16.6%) Hereditary septic Paraparesis, 3 Hirayama disease (7.14%), monomyelic amyotrophy 1 (2.38%), others were spino muscular atrophy, primary lateral sclerosis.

MRI of spinal cord was normal for 13 (30.95%) cases of degenerative myelopathy. 11 out of 12 cases (26.2%) of SCA showed Spino cerebellar degeneration. 6 other (14.29%) cases showed spinal cord degeneration. Changes were more in the cervical region (9.53%). 3 (7.14%) cases showed T2 hyperintensity in middle cerebellar peduncle.

DISCUSSION

There are several studies in India showing the incidence of non-compressive myelopathy in the range 14%-62% among different series reporting myelopathy. In our study it was 46%. In the present study, a probable aetiology of noncompressive myelopathy in 74 (97.3%) cases has been noted which was comparable to other series from India.

The present study revealed that the non-compressive myelopathy affected patients in the prime of their lives with 71% of them in the age group 20-50 years. There was a slight male preponderance which was consistent with studies from other parts of India.[5]

Degenerative myelopathies and non- degenerative myelopathies need to be discussed separately.

Genetic tests carried out for spino cerebellar ataxias were found to be conclusive in one patient with SCA-3. Rest of the cases (2, 4.35%), genetic tests with selected mutation panels were negative. Those cases required further testing for specifying the variety of SCA.

Prevalence of MND shows variation from 4 per 100,000 in a Bangalore study (Gourie Devi et al.)[6] and 2.86 per 100,000 in rural Bengal study (Das et al.)[7] The high incidence of motor neuron disease (MND) (13, 28.2%) in the study is due to referral from local hospitals from nearby districts. The present study showed predominant male affection for MND (1.9: 1) which is similar to the figure in western literature where a male predominance with a range between 1.2: 1 to 20: 1.

ALS was the most common form which is consistent with other studies from India and abroad

The striking features of this study were two of patients with wasting and weakness restricted to either a single upper limb, slow course and lack of involvement of other part of CNS and PNS.

This type which Gourie Devi coined the term "Monomelic Amyotrophy" constituted 22.7% of this series. This was Reported first from Japan by Hirayama et al.

Many of the non-degenerative causes were treatable. In western countries non-compressive myelopathies are mostly either due to demyelinating diseases or hereditary degenerative diseases[8], whereas para infectious mediated damage is more in the developing countries.[9] However more primary demyelinating disease like multiple sclerosis and NMOSD are increasingly been recognised in India.[10]

In the present study, a probable aetiology of noncompressive myelopathy in (91.3%) cases has been noted which was comparable to other series from India.

Among the non- degenerative causes inflammatory lesions remained the major bulk of non compressive myelopathy. 3 cases of Post infectious myelitis had clear history of febrile illness within previous 30 days were negative for evidence of MS and NMOSD. One case with sore throat as well as respiratory tract infection and one case with gastrointestinal problem were soon followed by acute onset of paraparesis.

The western pattern of multiple sclerosis where brainstem and cerebellar involvement is common, but the opticospinal form is commoner in India. Our study revealed a similar pattern of involvement. Previous studies reporting infectious myelitis from India revealed viral etiology as the commoner cause.[12]

The patient with spinal cord ischemia developed paraplegia within 2 hours. Clinical examination was consistent with anterior spinal artery infarct. MRI spinal cord revelaed Owl's eye appearance along with evidence of vertebral body infarct indicating vascular etiology of the disease.

Atrophy of cord and long segment involvement was associated with poorer prognosis. 3 of the 4 cases of NMOSD had long segment myelitis.

Delayed sequel of electrical injury has been described in the medical literature.[13] We observed 2 cases of such illness where there was insidious onset of progressive LMN type of weakness in upperlimbs. Radiological and eletrophysiological studies concluded them as anterior horn cell disorders. One case of post electrocution myelopathy presented as acute paraplegia with sphincter and sensory involvement.

Syphilis is now a rare disease. A routine test of CSF VRDL may be one of the methods to scout a diagnosis of syphilitic myelitis. Patient who was diagnosed with syphilitic myelitis had subacute presentation. Studies from Africa showed high incidence of HTLV-1 infection in progressive Paraparesis patients.[14] In our study one case of chronic paraperesis with bladder and sensory symptoms was diagnosed with tropical spastic Paraparesis.

In the nutritional myelopathy arm nerve conduction studies revealed neuropathy in 4 out of 6 cases. In 5 cases vitamin B12 levels were found to be low. 2 of them had Hypersegmented neutrophils. 4 cases had Macrocytosis.

MRI showed signal changes in 7-50% of cases in previous studies of demyelinating myelopathies which includes ATM, spinal cord MS and NMOSD.[15,16] MRI will be likely normal in early part of the disease. In our study 66% of such cases showed signal changes. According to literature early abnormal MRI findings were associated with bad prognosis.[16]

Many interesting findings noted in MRI were T2 hyperintensities in a case of Radiation Myelitis which is consistent with older studies on radiation myelitis.[17] One patient with vitamin B12 myelopathy showed T2 hyperintensity in the postero-lateral columns. All cases which showed evidence of necrotizing myelopathy had bad prognosis and poor outcome. Cranial MRI was abnormal in 4 cases (66%) myelitis due to multiple sclerosis.

From a study from 80s age above 40 years, rapid progression of the disease, prolonged spinal shock stage and absence of findings in CSF to be associated with poorer prognosis.

CONCLUSION

In our study non compressive myelopathy affected people across all age groups but most of them especially non degenerative causes belonged to productive members of the family. The causes of myelopathy included motor neuron diseases, degenerative ataxias, hereditary spastic paraparesis, immune-mediated, vascular and metabolic disorders, infectious, and physical agents. In spite of all possible investigations 8.7% of cases with non-compressive myelopathy are of "undetermined" origin from aetiological point of view. An underlying disease may be revealed in long-term follow-up of the patients as observed in other long duration studies.

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