



A PROSPECTIVE STUDY ON DEMYELINATING DISEASES OF CENTRAL NERVOUS SYSTEM IN A TERTIARY CARE HOSPITAL

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ABSTRACT

Introduction: Demyelinating diseases characterised by inflammation and selective destruction of CNS myelin and are most common cause of non traumatic disability in young

Aim: To study clinical presentation, course, prevalence of primary and secondary demyelinating lesions and response to steroids and short term follow up

Materials And Methods: Prospective study in teaching hospital from May 2016 to September 2017 of fifty suspected male and female of age > 13 years subjected to serological and radiological tests

Results: Most common age group affected was 20-30 years, accounting for 30% of cases. 18% of cases were <20 years of age, 22% and 14% were 31-40 and 41-50 years respectively. None of the patients were > 60 years of age. 31 patients (62%) were females and 19 (38%) were males. 25 cases (50%) were transverse myelitis; 12 (24%) ADEM; 5 (10%) multiple sclerosis; 2 (4%) NMOSD and 6 cases (12%) were secondary demyelinating diseases. Paraparesis and paraplegia most common presentation (48%) followed by bladder involvement (32%). Quadriparesis and quadriplegia seen in 22% , Sensory involvement in 6%, INO 4%, motor and optic nerve involvement 4% , hemiparesis in 4% and ataxia was seen in one case.

Conclusions: Demyelinating diseases of CNS are common in younger age group, females are most affected. Primary are more common than secondary .NMO responds poorly to steroids

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INTRODUCTION

Demyelinating disorders of CNS are characterized by inflammation and selective destruction of central nervous system (CNS) myelin. The peripheral nervous system (PNS) is spared and most patients have no evidence of an associated systemic illness. Inflammatory demyelinating diseases of the central nervous system occur throughout the world and are the foremost cause. of the nontraumatic neurological disability in young adults Multiple sclerosis ^(1,2) is the most common of these disorders, others include acute transverse myelitis, Acute disseminated encephalomyelitis (ADEM) ^(3,4,5) and Neuromyelitis optica (Devics disease) ^(6,7) . Although these disorders are all similarly characterized by focal CNS demyelination they vary in their clinical course, prognosis, regional distribution, pathology and pathogenesis.

Aim

To study clinical presentation , course ,prevalence of primary and secondary demyelinating lesions and response to steroids and short term follow up .

MATERIALS AND METHODS

This is prospective observational study conducted in Andhra medical college , over the period of 18 months from May 2016 to September 2017. This study was approved by the ethics committee of our College. The study population involves 50 cases of demyelinating disease of central nervous system. The patients were selected from general medical ward and neuro medical ward.

Inclusion Criteria: Any male or female patients more than 13 years of age with long tract involvement (like pyramidal tract, MLF, posterior column, cerebellar pathways) with or without optic nerve and bladder involvement.

Exclusion Criteria: Patients without evidence for long tract Involvement were excluded from this study

After taking due consent, detailed history, physical examination, appropriate blood investigations, CSF analysis

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and imaging of patients done as deemed necessary. All the patients were treated with 1g IV methyl prednisolone for 5 days. Then put on oral prednisolone tapering dose and Azathioprine 2 mg/kg /d. All the patients were reassessed after 6 weeks.

RESULTS

Age Distribution

Table 1 age distribution of cases

Age group in years	Number of cases (n=50)
<20 yrs	9
20-30	15
31-40	11
41-50	7
51-60	8
>60 yrs	0

Gender Distribution

Out of 50 cases, 31 were females and 19 were males.

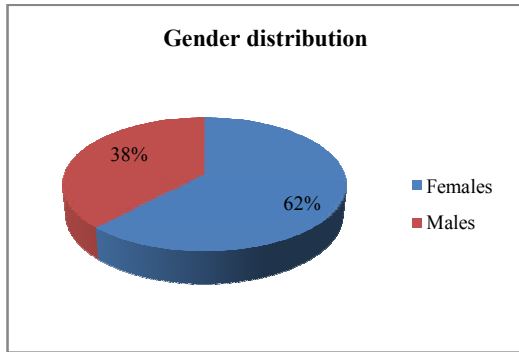


Figure 1 Gender distribution among cases

Clinical Presentation

Paraparesis and paraplegia most common presentation (48%) followed by bladder involvement (32%). Quadriparesis and quadriplegia seen in 22% , sensory involvement in 6%, INO 4%, motor and optic nerve involvement 4% , hemiparesis in 4% and ataxia was seen in one case

Table 2 Clinical presentation

Clinical presentation	Presentation in number
Paraparesis and paraplegia	24
Quadriparesis or quadriplegia	11
Bladder involvement	16
INO	2
Sensory involvement	3
Ataxia	1
Hemiparesis	2
Motor and optic nerve involvement	2

Primary demyelinating diseases were more common than secondary in this study

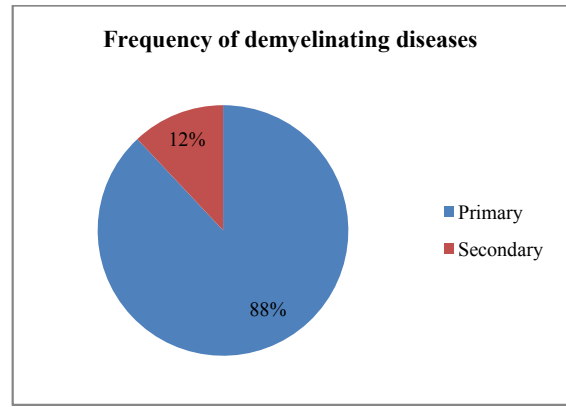


Figure 2 Frequency of primary & secondary demyelinating diseases 25 cases (50%) were transverse myelitis; 12 (24%) ADEM; 5 (10%) multiple sclerosis ; 2 (4%) NMOSD and 6 cases (12%) were secondary demyelinating diseases.

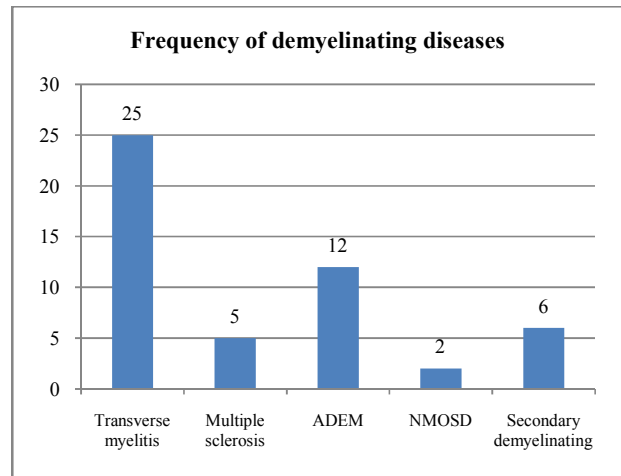


Figure 3 Frequency of demyelinating diseases

Response to Pulse Steroid Therapy

All the patients of this study were treated with pulse steroid regimen, INJ.METHYL PREDNISLONE 1G Daily for 5 days

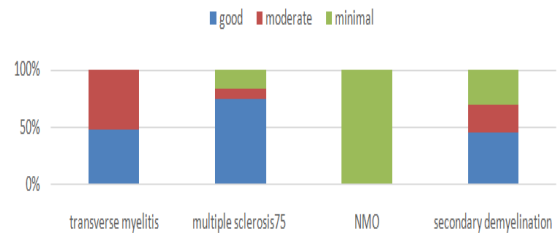


Figure 4 Response of demyelinating diseases to steroids

DISCUSSION

In this study, the age distribution showed that younger age groups were more affected , 70% of the patients belong to less than 40 years of age and no cases were seen in more than 60 years of age. This is in accordance with literature that demyelinating diseases are most common in young age. The median age of presentation is 23 years while the study by Sheffali Gulati *et al* (8) the median age at initial presentation was 7 years (range: 1-12 years) . The mean age of presentation was 10.5 years in a Canadian study of Banwel.B *et al* (9).

With regards to the sex predilection, females were more affected (62%), than males (38%). In prospective study of SHANIJA.P *et al*⁽¹⁰⁾, similar to our study females were more affected (58%), than males (42%). In prospective nation wide dutch study⁽¹¹⁾, the ratio between females and males in all demyelinating diseases patients did not differ significantly between the presenting phenotypes.

The clinical presentation varied from motor weakness in the form of paraplegia, paraparesis, quadriplegia, quadriparesis, monoperesis, or rarely hemiplegia, sensory involvement in the form of paraesthesia, brain stem involvement in form of bilateral INO, optic nerve involvement with optic atrophy, cerebellar ataxia, and bladder involvement. In our study, Paraparesis and paraplegia was most common presentation (48%) followed by bladder involvement (32%). Quadriparesis and quadriplegia seen in 22%, sensory involvement in 6%, INO 4%, motor and optic nerve involvement 4%, hemiparesis in 4% and ataxia was seen in one case. Review of literature showed similar presentation in most of the studies.

In our study, 50% of cases were transverse myelitis, 24% ADEM, 10% multiple sclerosis while in prospective Dutch study, multiple sclerosis was most common (33%), and transverse myelitis (9%); ADEM (29%) and NMOSD (5%) accounted for rest of cases. In prospective study of SHANIJA.P *et al*, transverse myelitis accounted for 42%, multiple sclerosis 24%, ADEM 22%. In Canadian study of Banwell B *et al*, the most common presentations were optic neuritis (23%), ADEM (22%), and transverse myelitis (22%). In our study, secondary demyelination accounted for 12% of cases while in study of SHANIJA.P *et al*, secondary demyelination was seen in 8%.

In our study, multiple sclerosis responded well to steroids while NMO showed very minimal response. This is similar to study of SHANIJA.P *et al*, 75% of multiple sclerosis and 50% transverse myelitis showed good response while NMO showed no response. In the retrospective study of Sheffali Gulati *et al*, Steroids caused significant improvement in acute episodes of demyelination. However, recurrent demyelinating disorders like multiple sclerosis and NMO required long-term immunomodulation. Azathioprine currently is the most favored long-term immunomodulator used in NMO. Interferon- β and glatiramer acetate are currently recommended for multiple sclerosis. However, azathioprine may be a suitable alternative in a resource-limited setting.

All the patients were followed up for six weeks after steroid therapy. The recovery from neurological impairment was good in 75% of MS patients, 50% of the idiopathic demyelinating transverse myelitis patients

CONCLUSIONS

Demyelinating disorders of the central nervous system are common among younger age group. Females are more affected than males. Primary demyelinating diseases are more common than secondary demyelinating diseases. Most of the patients presented with pyramidal tract involvement followed by bladder involvement. NMO responds poorly to steroids.

Limitations of This Study

Small sample size. Long term follow up required

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