

THE SPECTRUM OF INFLAMMATORY DEMYELINATING DISEASES OF THE CENTRAL NERVOUS SYSTEM

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ABSTRACT

INTRODUCTION

Idiopathic inflammatory demyelinating diseases (IIDDs) are rare neurological diseases. Their features differ from region to region. We characterize features of these diseases in Chittoor.

METHODS

We describe 100 patients of IDD from Sri Venkateswara Institute of Medical Sciences, Tirupathi from May 2012 – December 2013.

RESULTS

10 patients with multiple sclerosis, 14 with ADEM, 6 NMO, 9 with ATM and 9 ON presented with the mean of 32 years with slight female predominance.

CONCLUSION

The clinical and imaging profile of IDD in Chittoor, is comparable to other Indian and international studies.

KEYWORDS

Demyelinating diseases, NMO spectrum, Multiple sclerosis.

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INTRODUCTION: Idiopathic inflammatory demyelinating diseases (IIDDs) include a broad spectrum of disorders characterized by inflammation and selective destruction of the myelin. The common inflammatory demyelinating diseases of the central nervous system include multiple sclerosis, acute disseminated encephalomyelitis, disorders of neuro myelitis optica (NMO) spectrum and related disorders.¹ There are very few studies characterizing the disorders from South India. We describe the profile of IDD in our centre.

AIM OF STUDY: To study the clinical spectrum, radiological and laboratory profile of various subtypes of idiopathic inflammatory demyelinating disorders of the central nervous system.

OBJECTIVE: To study the clinical spectrum of IDD in patients presenting to SVIMS, Tirupathi.

MATERIAL AND METHOD: From May 2012 to December 2013 all patients attending outpatient department or

admitted in Neurology / Medicine wards of Sri Venkateswara Institute of Medical sciences, Tirupathi meeting the criteria of idiopathic inflammatory demyelinating diseases of the CNS²⁻⁵ were included. A detailed medical history and thorough clinical examination was performed in all cases. Patients were classified into MS, NMO, ADEM, ATM and CIS based on the standard criteria.²⁻⁵ Relevant investigations were performed at the discretion of the neurologist. All patients were managed using IV steroids, disease modifying therapy in appropriate situations and were followed up for response to treatment. The study was approved by the Institutional Ethical Committee. (ROC.No.A&E/08/IEC/SVIMS/04; 13-06-2012). Written informed consent was obtained from all patients or from guardian in case the patient was unconscious for participation in study.

Inclusion Criteria: Patients diagnosed by the criteria of idiopathic inflammatory demyelinating diseases of the CNS.²⁻⁵

Exclusion Criteria: Patients who met the following exclusion criteria were excluded from the study.

Patients suffering from neurologic conditions that could be better attributed to other white matter diseases of the CNS.

i. **Specific Infections:** HIV, Tuberculosis, Syphilis, Viral encephalitis, Neurocysticercosis.

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- ii. **Metabolic/Nutritional:** Subacute combined degeneration, Wernicke’s encephalopathy, Osmotic demyelination syndrome.
- iii. **Vascular:** Vasculitis, Cerebral venous thrombosis.
- iv. **Neoplastic:** Gliomatosis cerebri, Primary CNS lymphoma, Paraneoplastic syndrome.

Other: Degenerative disorders (spinocerebellar ataxia, Leukodystrophies), Radiation, Toxic (Carbon monoxide, chemotherapy with methotrexate and 5-Fluorouracil).

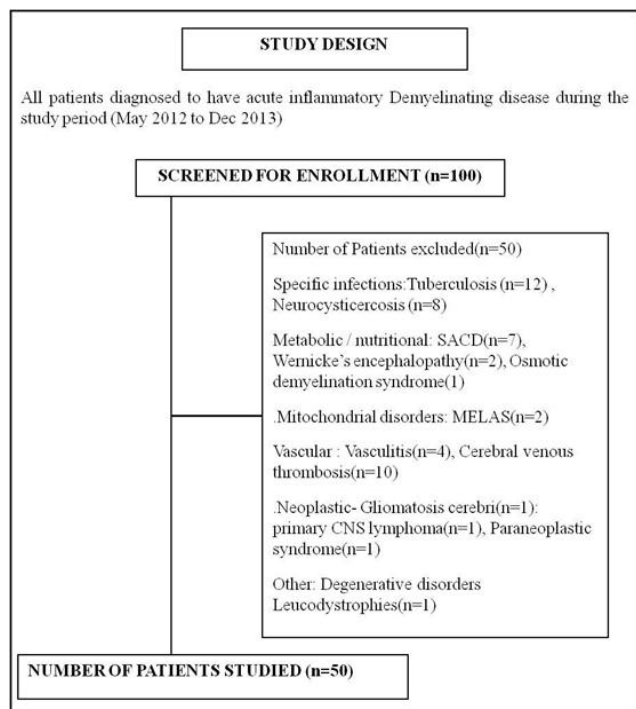


Figure 1

STATISTICAL ANALYSIS: Data was recorded on a predesigned proforma and managed using Microsoft Excel 2007 (Microsoft Corp, Redmond, WA). Descriptive statistics for the categorical variables performed by computing the frequencies (percentages) in each category. For the continuous variables, approximate normality of the distribution was assessed. Statistical analysis was done using Chi-square test for discrete variables and student’s T test for continuous variables. Variables following normal distribution were summarized by mean and standard deviation. Variables with non-parametric distribution were summarized as median [interquartile range (IQR)]. The statistical software IBM SPSS statistics version 20 (IBM Corp Somers NY, USA) used for statistical analysis.

RESULTS:

Age and Gender Distribution: In the present study mean age of patients was 31.72+15.32 years with median (IQR) 32(17-42) years. Majority were in their 2nd to 4th decade.

Characteristic	MS (10)	ADEM (14)	NMO (6)	ATM (9)	ON (9)
Demographic					
Age	37		38	36	32
M:F	2:8	4:3	2:4	4:5	1:2
Clinical Features					
Weakness	7	10		9	
Visual Disturbance	4	4			9
Ataxia	3	3		1	
Sensory Disturbance	5			3	
Bladder Disturbance	4			5	
Bowel Disturbance				6	
Seizures	1	4			
Abnormal Behaviour		4			
Tremor		2			
Speech Disturbance		7			
Altered Sensorium		5			
Fever		12		3	
Relapsing Course	4		3		

Table 1: Clinical features

Characteristic	MS (10)	ADEM (14)	NMO (6)	ATM (9)	ON (9)
High CSF Protein		2		1	
CSF Pleocytosis		4		3	
CSF Oligoclonal bands	1				1
Aquaporin Antibodies			5		
BAEP		1			
VEP	2	1			
SSEP		2			
MRI Brain Abnormality	7		2		3
MRI Spine Abnormality	3	7	6		

Table 2: Laboratory findings

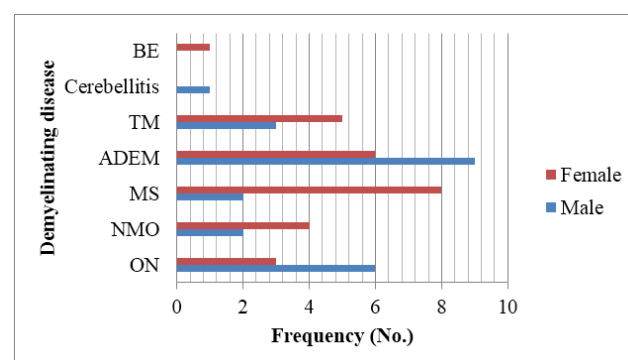


Figure 1: Disease frequency (n) of males and females in 50 patients

Treatment: All patients received pulse dose of steroids. Only two patients received plasmapheresis. One female patient with relapsing NMO who had recurrent admissions developed steroid resistance and one middle aged woman diagnosed with ADEM received 5 cycles of plasma exchange. She recovered fully after two months of intensive physiotherapy exercises.

DISCUSSION: The inflammatory demyelinating diseases of the CNS in Sri Venkateswara Institute of Medical sciences, Tirupathi was studied to know the spectrum. The clinical features and imaging findings of patients with multiple sclerosis, ATM and optic neuritis were comparable to similar studies from India (Table 3).

Variable	Western India, J Mani et al ⁽⁶⁾ (n = 31) Frequency (%)	South India, Sarma et al ⁽⁷⁾ (n = 68) Frequency (%)	Present Study (n = 10) Frequency (%)
MRI findings			
Subcortical	96	33.33	33.33
Periventricular	-	33.33	33.33
Infratentorial	-	25	66.67
Spinal cord	15/16	41.7	85.71
Balos MS	-	-	-
Tumefactive MS	-	-	11.11

Table 3: Comparative Imaging profile of patients with MS

Variable	Prabhakar s. et al ⁽⁸⁾ n=32 Frequency (%)	Murthy.et al ⁽⁹⁾ n=13 Frequency (%)	Present Study (n =9) Frequency (%)
MRI spinal cord			
Cervical	26	69.33	77.78
Dorsal	32	23	55.56
Lumbosacral	3	-	-

Table 4: Comparative Imaging profile of patients with ATM

Variable Frequency (%)	ONTT ⁽¹⁰⁾ et. al ⁽¹¹⁾ Frequency (%)	Rohit Saxena (n =9)	Present Study
Clinical features			
Loss of vision	90	100	100
Color desaturation	88	100	100
Painful eye movements	92.2	66	100
Optic disc swelling	35.3	61.76	77.78

Table 5: Comparative clinical profile of patients with optic neuritis

Loss of vision, Colour desaturation of present study were coinciding with Rohit Saxena et al study (44). Apart from all NMO patients one ADEM patient had bilateral optic neuritis.

Variable	Das k. et al ⁽¹²⁾ (n=62) Frequency (%)	JayaKrishna.et al ⁽¹³⁾ (n=14) Frequency (%)	Present Study (n =14) Frequency (%)
MRI			
Supratentorial	74.19	90	64.29
Thalamus	12.9	7	21.49
Basal ganglia	6.45	21	21.49
Medulla	12.9	7	42.85
Cerebellum	19.35	7	-
Spinal cord	-	64	50

Table 6: Comparative Imaging profile of patients with ADEM

Supratentorial lesions were predominant in all studies. When compared to other studies number of patients with ADEM were more in our study (Table 7 and Table 8).

Diagnosis	Butchi Raju G et al. ⁽¹⁴⁾ Study (n=56)	Present Study (n=50)
Multiple sclerosis	8	10
Neuro myelitis optica Encephalomyelitis	12	6
Acute disseminated Encephalomyelitis	9	14
Clinically isolated Syndrome (including transverse myelitis)	27	20

Table 7: Comparison of different types of Central Nervous system inflammatory demyelinating disorders to Indian study

Diagnosis	ChanK. H et al. ⁽¹⁵⁾ (n=210)	Present Study (n=50)
Classical MS	88(41.9%)	10(20%)
Neuromeningitis Optica	47 (22.4%)	6(12%)
Transverse Myelitis	20(5.6%)	9(18%)
Acute Disseminated Encephalomyelitis	9(4.3%)	14(28%)
Idiopathic Relapsing optic Neuritis	6(2.9%)	4 (8%)
Single attack of optic Neuritis	21(10%)	5(10%)
Single attack of Brain stem Encephalitis	3(1.4%)	1(2%)
Cerebellitis	NR	1(2%)
Single attack of myelitis	23(11.0%)	NR
Relapsing myelitis	10(4.8%)	NR
Optico spinal multiple sclerosis	3(1.4%)	NR

Table 8: Comparison of different types of Central Nervous system inflammatory demyelinating disorders to Hong Kong study

CONCLUSION: It reflects the spectrum of IDD in Chittoor region. Our study shows that the clinical spectrum of IDD is comparable to other Indian and International reports.

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