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Spectrum of Demyelinating Diseases - A Etiological, Clinical Profile with Short Term Follow Up

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Abstract: ***Demyelinating diseases form a class of disorders characterized by loss or attenuation of the myelin sheath which include, Autoimmune, Infectious, Toxic / Metabolic, Vasular and Hereditary Disorder. Our Study aims to study the spectrum of patients with demyelinating illness and to study the antecedent events, clinical profile and etiological factors among them and to analyse the short term outcome. The spectrum of patients with demyelinating illness admitted comprised of 88% GBS, followed by 9.3% ADEM and 2.7% multiple sclerosis. Of the 23 cases that needed ventilator assistance, 8 weaned in less than 4 days, 7 between four and eight days, 6 between eight and twelve days and 2 above twelve days.***

Keywords: MS - Multiple Sclerosis; MRI - Magnetic Resonance Imaging; ADEM - Acute Disseminated Encephalo Myelitis; GBS - Guillian Barr'e Syndrome

1. Introduction

Demyelinating diseases form a class of disorders characterized by loss or attenuation of the myelin sheath in the relative absence of neuronal loss and it can occur in both in peripheral and central nervous systems. The myelin sheath provides a high reisistance, low capacitance insulation between the sites of action potential electrogenesis, the node of Raniver. It conducts impulses from one node to another in saltatory manner and therefore its loss is accompanied by significant conduction abnormalities. The demyelinating diseases include, Autoimmune (Acute disseminated encephalomyelitis, Acute hemorrhagic leukoencephalopathy, Multiple sclerosis), Infectious (Progressive multifocal leukoencephalopahty), Toxic / Metabolic, Vasular and Hereditary Disorder (Dysmyelinating) .

The term Guillian-Barr'e Syndrome defines a recognizable clinical entity that is characterized by rapidly evolving symmetrical limb weakness, loss of tendon reflexes, mild or absent sensory signs and variable autonomic dysfunctions. It has become the leading cause of acute flaccide paralysis after the eradication of poliomyelitis from many countries. Incidence was 4 per lakh population as per Kaplan et al. In1992, Ropper et al[3] reported the mean age of occurrence of GBS was 39 years withbimodal peak in 16-25 years and 45-60 years, Dowling et al in their study found no seasonal cluster. Rabies, BCG, T.T, influenza, measles, mumps rubella hepatitis B and polio was found to eb antecedent illness by Applebaum et al 1953, 1994. In typical GBS, Ropper et al 1992 found weakness in 60%, paresthesia in 80%, both in 30%, areflexia in 70%, hyporeflexis in 30% of patients. The pattern of weakness noted by Ropper et al 1992 was ascending in 54%, descending in 14% and equal in 32%. Anderson et al in 1982 noted ascending pattern in 43%, descending pattern in 5% and equal in 28% and in Ravn et al 1967 study it was ascending in 37% and descending in 4%.

Acute Disseminated Encephalo Myelitis (ADEM) is a demyelinating syndrome that occurs in association with an immunization or vaccination. The clinical presentation more common in children include, simultaneous bilateral optic neuritis, loss of consciousness, loss of Deep Tendon Reflexes and retained abdominal reflexes in presence of babinski's sign, central body temperature of greater than 100^0F and severe shooting limb pains. Multiple sclerosis has been the subject of study by workers all over the world. Low risk areas of less than 5 per 100,000 include most of Asia and all of Africa. Review of previous Indian literature of MS indicated that MS comprises 0.05 to 1.5% of the total neurologic inpatients. On an average, 1.6 to 5.4 new cases of MS[1] are seen at various centres per year. However two epidemiological studies among Parsis living in Bombay & Pune found a crude prevalance of 21 cases per 100,000 parsis in Bombay and 58 per 100,000 in Pune[2].

2. Aims and Objectives

- To study the spectrum of patients with demyelinating illness admitted in Govt. Rajaji Hospital, Madurai during the year 2016.
- To study the antecedent events, clinical profile and etiological factors among them.
- To analyse the short term outcome of the demyelinating illness.

3. Subjects and Methods

The study design was prospective one involving patients admitted in Medicine and all other wards for demyelinating illness.

Inclusion Criteria: Patients admitted for neurological symptoms and signs suggestive of demyelinating illness and investigation compatible with them were included in study group.

Exclusion Criteria: Patients with equivocal diagnosis or inadequate clinical details or investigations not compatible with demyelinating illness were excluded from study group.

4. Methodology

Patients general information antecedent events, symptoms were recorded in a predesigned proforma one allotted for each patient. Detailed clinical examination finings, pattern of weakness respiratory muscle weakness, requirement of ventilatory assistance were documented in that proforma. Statistical analysis was done on these data and Chi-Square Test, Student 't' test and p values were applied wherever possible.

S.No.	Demyelinating illnessess	No. of cases	Percentage
1	GBS	66	88
2	ADEM	7	9
3	MS	2	3

5. Results

Total cases with demylinating illness included in our study - 75Table 1: Showing the spectrum of distribution of demyelinating illnesses.

Table 2: Age and Sex distribution of patients with GBS

S.No.	*Age interval*	*Male*	*Female*	*Total*	*Percentage*
1	10-19	10	5	15	23
2	20-29	10	10	20	30
3	30-39	5	2	7	11
4	40-49	10	2	12	18
5	50-59	3	3	6	9
6	> 60	3	3	6	9

Table 3: Antecedent events preceeding GBS

S.No.	Antecedent event interval	No. of patients	%	't' test	'p' value
1	Respiratory illness	16	24	1.49	0.1413
2	Gastrointestinal illness	6	9	2.43	0.0204
3	Malignancy	1	2	2.58	0.0153
4	Pregnancy	3	5	2.58	0.0149
5	Post viral	2	3	2.78	0.0093
6	Non specific	8	12	2.25	0.0343

Table 4: Internal between onset and peak symptoms of patients with GBS

S.No.	Onset in days	No. of patients	%
1	< 7	7	10
2	8 - 15	44	67
3	> 16	15	23

Table 5: Signs and symptoms profile of patients with GBS

S.No.	Parameter	No. of patients	%
1	UL weakness	60	91
2	LL weakness	62	94
3	Ophthalmoparesis	10	15
4	Facial palsy	34	51
5	Bulbar palsy	26	39
6	Sensory symptoms	48	72
7	Dysautonomia	36	54
8	Respiratory failure	26	39
9	Ventilatory assistance	23	34

Number of patients died: 3 (5%)

Table 6: Ventilator days and outcome of GBS cases at the end of one month

S. No.	Ventilator days	Total	Good recovery	%	Mild-Moderate	%	Severe deficit	%
1	0 - 4	8	6	75	2	25	0	0
2	4 - 8	7	4	57	2	29	1	14
3	8 - 12	6	1	17	3	50	2	33
4	> 12	2	0	0	1	50	1	50

Chi-Square value - 4.3105: 'p' value - 0.635

Table 7: Age & Sex Distribution of patients with ADEM

S.No.	Age Interval	Male	Female	Total	%
1	10 - 19	2	2	4	57
2	20 - 29	1	1	52	29
3	30 - 40	1	0	1	14
4	Total	4	3	7	100

Males with ADEM - 4 (57%) Females with ADEM - 3 (43%)

Table 8: Antecedent events of patients with ADEM

S.No.	Antecedent event	No. of patients	%
1	Exanthematous fever	2	29
2	Post vaccinial	2	28
3	Non specific infections	3	43

Clinical and imaging profile of patients with ADEM are as follow:
Head ache - 4 (57%), Fever - 3 (43%), Meningismus - 4 (57%), Altered sensorium - 2 (29%),
Optic neuritis - 1 (14%), Cranial nerve palsies - 2 (29%), Pyramidal signs - 6 (86%),
Sensory deficits - 2 (29%), Ataxia - 2 (29%), Aphasia - 1 (14%), Seizures - 2 (29%),
MRI, periventricular lesions - 1 (14%), Thalamic lesions - 2 (29%),
Basal ganglia lesions - 2 (29%), Spinal cord lesions - 3 (43%),
Contrast enhancement - 6 (86%), CSF pleocytosis - 4 (57%)

Clinical and MRI profile of two cases of M.S.
20F; 1st attack - Rt hemiparesis; 2nd attack - Lt hemiparesis;
Course - Relaping remitting; Estimate site - Cerebral; MRI brain - 1Gd enhancing br lesion & 3 periventricular lesions; MRI spinal cord - Normal

31F; 1st attack - Paraparesis; 2nd attack - Quadriparesis;
Course - Relaping remitting; Estimate site - Spinal cord; MRI brain - Normal; MRI spinal cord - One lumbar healed other cervical contrast enhancing demyelinating plaque.

6. Discussion

Gullian Barre Syndrome, Antecedent illness - Various studies from India showed that 30 percent to 80 percent of GBS has antecedent illness. Our study showed 55 percent of patients had antecedent illnesses and the 'p' values for Gastro intestinal illness, malignancy, pregnancy, post viral and non specific infections were significant (Table 3)

Of the 23 cases that needed ventilator assistance, 8 weaned in less than 4 days, 7 between four and eight days, 6 between eight and twelve days and 2 above twelve days.

Overall mortality was attributed to the type of presentation of quality of intensive care and comobrid medical conditions and it varied from 2 percent[4] to 20 percent[5] . In our study the mortality was 5% and all those cases were died within 24 hours of admission due to respiratory failure.

7. Summary and Conclusion

The spectrum of patients with demyelinating illness admitted comprised of 88% GBS, followed by 9.3% ADEM and 2.7% multiple sclerosis.

The age group of patients with GBS varied from 10 to above 60 and 30% fell into third decade. Males: Females ratio is 1.63 : 1.

Antecedent events occurred in 55% of patients with GBS of which gastro intestinal illnesses constituted 9%, non specific fever 12%, pregnancy 5% and post viral 3% which were statistically significant.

All cases of ADEM belonged to 10 to 40 year age group and 57% fell into adolescent age group.

Non specific infections antedated 43% of ADEM and post viral and post vaccinal shared 29% and 28% respectively.

43% of ADEM were of meningitic form and remining encephalitic form and CSF pleocytosis was noted in 57% of cases.

References

[1] Jain and Maheswari et al 1985;
[2] Bharucha et al 1988, Wadia et al 1990;
[3] Massachusether General Hospital study;
[4] Lotter et al 1977; 5. Gibbel et al 1992