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Multiple Sclerosis - Clinical Analysis of Cases Seen During a Period of 22 Years at NIMHANS

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Abstract

Analysis of 65 cases of Multiple Sclerosis (MS) seen during a period of 22 years (1961 1982) at the National Institute of Mental Health & Neuro Sciences, Bangalore showed that optico-spinal form of MS was the commonest clinical type. The frequent occurrence of typical cases of neuromyelitis optica and tonic spasms in MS compared to the Western series are noteworthy differences in MS in Orientals and Caucasians. The low incidence of MS in India is reemphasised in the present study.

Key words -
Multiple Sclerosis,
Clinical features,
Orientals,
Caucasians

Multiple Sclerosis (MS) is known to have a low incidence in Asia including India [1], [2], [3], [4], [5]. Recently, Kuroiwa et al [6] collected data from 7 Asian countries and Hungary by using standardised coded international survey sheet. Analysis of this data showed that the prevalence rate of MS was 10 to 30 times less in Oriental population compared with Caucasians. The present authors participated in this programme and 50 cases (MS probable-42; Neuromyelitis optica-8) seen over a period of 20 years from 1961 to 1980 at the National Institute of Mental Health & Neuro Sciences, Bangalore were included in this study [7]. The present report is a further extension and includes 65 cases of MS (MS probable-56; Neuromyelitis optica -9) seen over a period of 22 years from 1961 to 1982. Physicians in India hesitate to make a diagnosis of MS in view of its rarity. Neverthless it is important to know the variation in the clinical picture of MS in Orientals and Caucasians. The present study also attempts to focus on these aspects.

Material and Methods

Of the 65 cases reported in the present study, 47 patients were personal1y examined by one or both authors and in the rest 18, case records were reviewed. The data was analysed as per the protocol of "Multiple Sclerosis and allied diseases (International survey)".

MS "Probable" (56 cases)

Criteria of Schumacher et al. , [8] were followed for the diagnosis of "MS Probable".

1. There must be objective abnormalities attributable to 'dysfunction of central nervous system.
2. Involvement of two or more separate areas in the central nervous system.
3. Central nervous system disease must reflect predominant white matter involvement.
4. Dissemination in time between two or more episodes of one month or more, each episode lasting at least 24 hours.
5. Age at onset should be between 10 and 50 years.
6. Past history of remission.
7. Decision by a competent physician that the symptoms and signs cannot be explained by some other disease process.

53 cases conformed to all the above criteria except 3, whose age of onset was below 10 years. However since they fulfilled all other conditions and the clinical features were typical of MS they were included in this category. Other authors have also included patients with age of onset below 10 years in this category [9].

Neuromyrlitis Optica (9 cases)

The criteria for diagnosis were

(a) Acute or subacute onset
(b) involvement of both optic nerves and spinal cord
(c) the two lesions should occur either concomitantly or within an interval not exceeding one month and
(d) improvement may or may not occur.

All the patients were admitted in the Neurology unit for detailed clinical assessment. Routine haematological tests and cerebrospinal fluid (CSF) examination, were done. Immunoelectrophoresis of CSF could not be carried out, due to lack of facilities. Appropriate neuroradiological procedures were done, whenever indicated, to exclude other causes. Neurological deficit and physical disability was recorded during the follow up period.

Resu1ts

Approximately 3 new cases of MS were seen in an year, forming less than 1 percent of total neurology admissions. Of the 65 patients, 39 (60%) belonged to urban areas while 26 (40%) were from rural areas. The distribution in various socio economic groups was as follows lower 47 (72.3%); middle 11 (16.9%) and upper-7 (10.8%). There was no correlation of economic status with the age of onset or severity or type of MS.

A.MS "Probable"

Age of onset is shown in Table I. The average age of onset was 27.4 years (range 4 ½ to 52 years). The maximum incidence was in the 3rd and 4th decades of life. Preceding nonspecific symptoms were seen in less than 20% of the patients; the commonest were fever (8 cases) and headache (5 cases). Vague pains, abdominal pain, diarrhoea, vomiting, giddiness were rare symptoms (8 cases).

Table I - Age of onset of 65 patients of Multiple Sclerosis

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The onset of illness was acute in 37 (66.1%), subacute in 10 (17.8%) and insidious in 9 (16.1%). The commonest initial neurological symptom (table II) was visual loss (46.4%), unilateral involvement being more common (30.3%) than bilateral loss. Other presenting neurological symptoms were motor weakness, sensory symptoms, unsteady gait, sphincter disturbances and double vision, in the decreasing order frequency.

Table II - Incidence of initial neurological symptoms in MS "Probable"

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During the course of illness (Table III), motor weakness was the commonest feature followed by visual loss. Visual loss was more often bilateral. Optic atrophy was noticed in nearly two thirds of patients. Once again bilateral involvement was more frequently seen than unilateral affection. Sphincter disturbance was the next common symptom. Ataxia of limbs/trunk was seen in more than half the number of patients. Mental symptoms were present in 23 per cent while tonic spasms were observed in 12.5 per cent. Lherrnitte's sign and convulsions were each present in 10.7 per cent. Other rare clinical features are listed in Table III. Cerebrospinal fluid examination was done in 55 patients. There was no pleocytosis in 44 cases (80%), mild increase of cells (5-20/cmm) in 7 patients (12.7%) and more than 20 cells/cmm in 4 cases (7.3%). Moderate elevation of protein (40-100 mg%) was observed in 19 patients (34.5%) and normal levels in 35 cases (63.6%). Only in one patient protein content was more than 100 mg per cent.

Table III - Symptoms during the course of illness in MS "Probable"

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The common sites of lesion were in the optic nerve (78.6% :44 cases) and spinal cord (78.6% :44 cases), followed by brain stem (57.1% :32 cases) cerebrum (19.6%: 11 cases) and cerebellum (12.5% :7 cases). In 42.9% (24 patients) more than two sites were affected and in the rest 57.1% (32 cases) 2 sites were involved.

B.Neuromyelitis Optica (9 cases)

The mean age of onset was 23.1 years with a range of 5 to 49 years. The symptoms were noticed for the first time before the age of 20 years in 6 patients (66.7%) (Table 1). The sex ratio M: F was 3.5:1. The onset of disease was acute (less than a week) in 7 (77.8%) and subacute (more than a week) in 2 patients (22.2%).

Motor weakness and bilateral visual loss occurred simultaneously in 5, motor weakness preceded visual loss in 2 and visual loss occurred before motor weakness in 2 patients. Cerebrospinal fluid cell count was normal (0-5/cmm) in 4 patients, slightly elevated (5-20/cmm) in 3 and moderately elevated (20-30/cmm) in 2 patients. The protein levels were normal in 6 patients and in the rest 3 were moderately elevated (40-100 mg%).

Forty four of the total group of 65 cases were treated with prednisone, prednisone and ACTH in 9 and ACTH alone in one case. In 11 no drugs were given as the patients were already in remission. Active and passive physiotherapy was initiated in all patients with motor disability.

C. Follow-up data

All the patients included in the study were initially admitted for detailed assessment. Two patients died in the hospital. Sixty three patients were seen on more than one occasion in the outpatient clinic. During the period of study (1980-1983), a fresh attempt was made to call all the patients by writing letters for reassessment. Information obtained in response to letters revealed that 5 more patients had expired during the follow up period bringing the total deaths to 7. Of the 58 alive patients, 41 reported to the outpatient clinic and were reassessed in a detailed manner. In the remaining 17 patients, information about the present condition could be obtained in 15 by using a structured proforma with details of "activities of daily living" and "functional neurological status". Two patients were lost to follow up.

The follow up data is shown in Table IV. It is seen from the results that in nearly two thirds of patients with either motor disability or visual deficit, recovery was satisfactory. Remissions and relapses occurred in 78.6 per cent of MS 'probable' group.

Table IV - Follow-up data of 65 patients of Multiple Sclerosis

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Discussion

Although multiple sclerosis was described more than 100 years ago by Charcot, there is no confirmatory diagnostic test, hence diagnosis is necessarily based on clinical criteria. The difference in prevalence rates reported from different countries may be partly due to differing diagnostic criteria. By using standardised criteria and data coding, it was observed that MS in Orientals has a low prevalence rate compared to Caucasians, [6] confirming previous observations [10].

During the period of 22 years, 65 cases of multiple sclerosis (MS 'probable' and 'Neuromyelitis optica') were seen at the National Institute of Mental Health & Neuro Sciences, giving an average of 3 new cases an year. The percentage of MS cases considered in relation to total Neurology admissions (23724) was 0.3 per cent confirming the low incidence. Cases fulfilling diagnostic criteria of MS "Possible" were not included, since a variety of other neurological conditions may also have similar features; additionally it would be safer to exclude these cases in "low risk" regions like India.

Neuromyelitis optica formed 13.8 percent (9 patients) of the total MS cases, confirming the common occurrence of this clinical form in Asian countries (3-13%) as against in Western countries where it was almost absent [11]. The most common type of MS was the optic-spinal form. The frequent occurrence of visual symptoms at onset and during the course, spinal cord involvement and relatively less common evidence of cerebellar involvement, are noteworthy features in our series, as well as in other series reported from Asia, compared to the Western countries [2], [4], [9], [11].

Another feature of interest is the symptom of 'tonic spasms'. Our figure (12.5%) is similar to those reported from Japan [8]. However, other reports from Northern and Western parts of India indicate a much lower incidence ranging from 1.9 to 6.2 percent [2], [4].

It has been stated that individuals from higher socioeconomic strata are more prone to develop MS, however our observations are not in conformity [12]. The large majority of patients belonged to the low

socioecononac strata (72.3%), the incidence being proportionate to the general pattern of socioeconomic structure of the society.

Except for the differences mentioned, the natural history of MS in Orientals and Caucasians in general, is not different [6], [11]. Similarly, based on autopsy studies, Seitelberger [13] found that there was no essential difference in MS in Asia or West in topographical distribution of lesion. In the Eastern cases however, the pathological changes were found to be more severe leading to loss of axis cylinders, partial necrosis, and spongy state. These features were particularly evident in optic chiasma and spinal cord, explaining the higher incidence of neuromyelitis optica and opticospinal form of MS in East.

The reasons for the racial differences in the incidence, clinical and pathological features in Orientals and Caucasians are not known. Further in depth studies are warranted

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