Motor neuron disease in blacks

Epidemiological observations in Natal

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Summary

A series of 86 black, Indian and white patients with motor neuron disease were analysed retrospectively. Although the material does not allow statistically valid conclusions, there are sufficient cases among blacks to allow two *prima facie* observations in this population group: (*i*) motor neuron disease has an earlier age of onset than in whites and Indians; and (*ii*) more patients come from peripheral and rural areas than would be expected in prevailing circumstances.

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We reported earlier that motor neuron disease occurs more commonly among blacks than Parkinson's disease, which is relatively rare in this race group.[1] The hypothesis that these conditions, and other neuronal abiotrophies, are the result of previous subclinical neuronal insult and subsequent age-related neuronal attrition[2] adds pertinence to epidemiological observations. For this reason we have examined the occurrence of motor neuron disease more closely.

Patients and methods

Case summaries of 95 patients diagnosed as having motor neuron disease were collected over 9½ years. These patients were of all races and had been admitted at least once to one of the four major public hospitals in Durban — King Edward VIII, Addington, R.K. Khan and Wentworth. Before 1984 patients were seen during neurological consultation in general wards. Since then most have been admitted to the Neurology Unit at Wentworth Hospital for assessment. After excluding 9 patients with doubtful diagnosis, there were 86 patients in whom the diagnosis of motor neuron disease was reasonably certain — 59 were black (36 men and 23 women), 16 white and 2 coloured (9 men and 9 women) and 9 Indian (8 men and 1 woman) and the mean age at onset, estimated from age at presentation and duration of symptoms, was 47,45 years in blacks and 54 years in whites and Indians. The patients had full clinical assessment and laboratory and radiological investigations, including myelography when necessary. After 1984 most patients had electromyographic studies, these were performed in 45,3% of the total series (39 patients). Because of geographical and socio-economic factors, and the unavailability of specific treatment, only 25 patients (29%) were seen after discharge from hospital. This lack of follow-up places certain restrictions on diagnosis in some cases.

It must be emphasised that this report does not profess to indicate the total prevalence of motor neuron disease in this area. However, because the patients were collected randomly, without any selection, and diagnosed by the same group of doctors, they broadly reflect the prevalence in the various race groups. The 86 patients in whom there was little doubt of the diagnosis were classified retrospectively according to their dominant features, into three types of motor neuron disease.[3] Those black patients with a combination of upper motor neuron (UMN) and lower motor neuron (LMN) signs, with a lesser degree of bulbar involvement in some, were classified as amyotrophic lateral sclerosis (ALS) — 27 men (75%), 12 women (52%). Those with predominant affection of bulbar muscles, including those of LMN type, were classified as progressive bulbar palsy (BP — 5 men, 6 women) and those with predominant LMN involvement of limb and trunk muscles were labelled progressive muscular atrophy (PMA — 4 men, 5 women). For whites and Indians combined the distribution was as follows: ALS — 15 males, 6 females; PMA — 1 male, 2 females; BP — 1 male, 2 females. There was, of course, considerable overlap between these types in some cases. Patients with clinical signs of pure UMN affection, who might have been considered to have primary lateral sclerosis, were not included in this series. This clinical picture is very similar to that of the hitherto unexplained myelopathy[4] that prevails in this region and may be associated with HTLV-I infection.[5] A few patients with pure localised LMN affection, who might have had early motor neuron disease, were excluded because it was not possible to perform follow-up studies.

Results

The principal findings are shown in Fig. 1. All races showed a preponderance of men, which accords with world experience.[6,7] The male/female ratio was 3:2 in the case of blacks, and in whites and Indians combined. No specific effort was made to elicit a history of injury or antecedent illness. The relatively high incidence of trauma (stab wounds 3 cases, fracture or other trauma 9 cases), hypertension (7 cases) and pulmonary tuberculosis (3 cases), which was recorded routinely in the case summaries, reflects the frequency of these conditions in the black population. Table I shows an attempt to relate these cases to the total population from which they are drawn[8] in order to provide a standard for comparison. The areas concerned — KwaZulu (8 cases), Transkei (8 cases), and other peripheral and rural areas of Natal (17 cases) — usually drain to Durban for specialist neurological services. Some black patients do not seek medical attention, and many with chronic neurological disease are probably not referred to large centres. The number of cases among whites and Indians is reduced by some who receive private medical attention. What these figures show is that motor neuron disease is by no means unknown among blacks and, allowing for failure to reach specialist medical centres, the disease may be as common among blacks as among whites and Indians.

The most remarkable feature revealed by this series is the earlier age of onset of motor neuron disease among blacks, with a peak in the 4th decade, whereas the greatest incidence in most other countries and among our white and Indian patients (Fig. 1) is 2 decades later. A further unexpected

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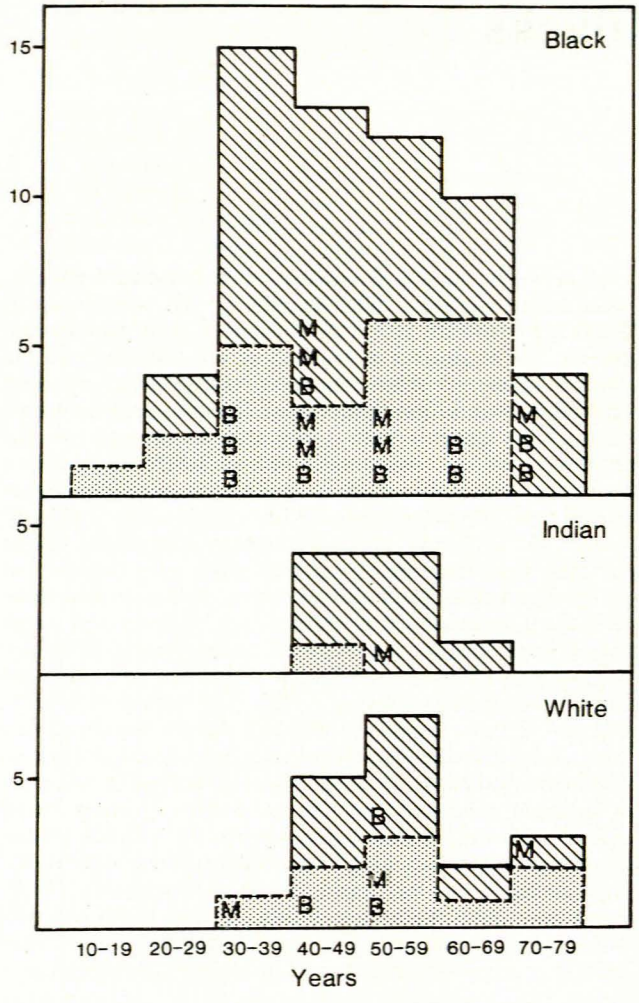

Fig. 1. Distribution of cases of motor neuron disease according to race, sex and age of onset (cross-hatched = men; stippled = women; M = case of PMA; B = case of BP; all other cases were classified as ALS.

finding is that 33 black patients (56%) were referred from areas outside the greater Durban area. These were from Transkei, KwaZulu and peripheral hospitals in Natal. This indicates distinct bias towards rural domicile, particularly since some patients who gave metropolitan addresses may have come from the country. This finding is not in keeping with the average hospital population or with the common assumption that rural patients seldom reach metropolitan hospitals. No evidence of clustering or familial incidence was found, but this would be difficult to establish without a field survey. Regarding the type of motor neuron disease, Fig. 1 shows the expected preponderance of ALS in all race groups. There is, however, a relatively greater proportion of BP and PMA among women, especially blacks.

TABLE I. RELATION OF MOTOR NEURON DISEASE TO TOTAL POPULATION

Race	Total population	MND cases	Rate/ 100 000
Blacks	**6 750 000**	**59**	**0,88**
Whites, coloureds	**662 000**	**18**	**2,7**
Indians	**659 000**	**9**	**1,4**

MND = motor neuron disease.

Discussion

The most striking finding is the early age of onset of most cases of motor neuron disease among blacks. The asymmetrical distribution of age incidence (Fig. 1), with a bias towards youth, is unlike any condition in which advancing age plays a part in its aetiology. The pattern for whites is more in keeping with this concept, and is more in accord with world experience.[6,7] Possible explanations for this unusual distribution among blacks are that a causative factor becomes operative at an earlier age, that we are dealing with a different condition or that the graph represents the superimposition of two conditions, one of which is motor neuron disease as seen among other races. In a population in which poliomyelitis has been very prevalent, the suspicion may arise that this early onset motor neuron disease is of the post-poliomyelitis type. Recent evidence from Britain has shown a relationship between past notification rates for poliomyelitis (1931-1939) and recent mortality from motor neuron disease (1969-1978).[9] Most of our young black patients with motor neuron disease showed the classic features of ALS with UMN involvement. They also tended to present at an advanced stage, unlike the benign appearance of those cases that have been reported after poliomyelitis.[10] Other virus infections may also be incriminated. HTLV-I appears to be associated with some of the unexplained spastic paraparesis that is prevalent in this region, but evidence has not shown that it is responsible for motor neuron disease, either here or elsewhere.[11]

A further observation is that, among blacks, motor neuron disease appears to be more prevalent in rural and peripheral areas than would be expected from relative population density and the composition of average hospital population. Surveys in other parts of the world have not shown excess of motor neuron disease among farm workers but there has been a suggestion that exposure to animal carcasses and consumption of large quantities of milk may be influential in the aetiology of motor neuron disease.[7] The only consistent association that has been shown is one with mechanical injury. However, the high rate of trauma among our patients is probably not unusual for this ethnic group.

These observations are offered in the full realisation that they will not stand close methodological scrutiny. The material suffers from the defects inherent in any random collection of hospital admissions. Nevertheless, it is felt that they should not be ignored on this account. They may provide pointers toward avenues for more definitive research into the cause and pathogenesis of this mysterious and tragic disease.

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