Unexplained spastic paraplegia

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Summary

Of 330 Black patients admitted to hospital with paraplegia, the causes in 33 remained unexplained after investigation. These patients had features of predominant corticospinal tract degeneration with lesser degrees of sensory loss. This series of patients is similar to some reported previously from South Africa and from other tropical countries. Among several possible causes the most likely is a toxic dietary factor.

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Neurology in the tropics and subtropics is beset by spinal cord disorders of various types. A number of these disorders remain unexplained. The object of this study was to assess the prevalence and nature of unexplained paraplegia in Natal Blacks and to determine whether these cases have features in common among themselves and in comparison with other series reported previously. Possible causes of the condition were examined.

Patients and methods

During the period of this study (January 1979 - May 1982) 330 patients were admitted to medical, surgical and orthopaedic wards at King Edward VIII Hospital, Durban with a diagnosis of paraplegia as a result of a spinal cord lesion. The final diagnoses in these patients are set out in Table I.

TABLE I. FINAL DIAGNOSIS IN PATIENTS PRESENTING WITH PARAPLEGIA*

Diagnosis	No. of patients	%
Trauma	**78**	**23,6**
Tuberculosis of vertebrae	**47**	**14,2**
Tuberculous arachnoiditis	**8**	**2,4**
Primary tumours	**29**	**8,8**
Metastatic tumour (known primary)	**14**	**4,2**
Metastatic tumour (undiagnosed primary)	**8**	**2,4**
Congenital vertebral deformity	**20**	**6,1**
Myelitis	**19**	**5,8**
Spondylotic myelopathy	**12**	**3,6**
Vascular cord lesion	**22**	**6,7**
Meningovascular syphilis	**20**	**6,1**
Tabes dorsalis	**2**	**0,6**
Thoracolumbar disc lesion	**11**	**3,3**
Portosystemic shunt myelopathy	**4**	**1,2**
Bilharzial myelopathy	**3**	**0,9**
Idiopathic myelopathy	**33**	**10,0**
	330	100

*Patients found to have peripheral neuropathy were excluded.

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No cause for the paraplegia could be found in 33 patients, and it is this group which forms the basis of this study. There were 23 males and 10 females, with a mean age of 42 years (range 13 - 61 years). All the patients were Black and the majority (29) came from rural areas. In all the condition had been insidious in onset, with a mean duration of symptoms of 10 months (range 1 - 36 months). All complained mainly of weakness and stiffness of the lower limbs, and abnormal physical findings were confined to the lower extremities. Details of the main signs and symptoms in these patients are set out in Table II.

TABLE II. MAIN CLINICAL FEATURES

	No. of patients	%
Symptoms		
Weakness and stiffness of legs	**33**	**100**
Subjective sensory change	**16**	**48,5**
Incontinence of urine	**14**	**42,4**
Physical findings		
Corticospinal tract involvement	**33**	**100**
Segmental sensory impairment	**12**	**36,4**

Where possible the history included details of diet, alcohol consumption, smoking and occupation. The findings are listed in Table III.

TABLE III. SOCIAL HISTORY

		Total*
Occupation		
Unemployed	**9**	
Manual (farm) work	**10**	**23**
Clerical	**4**	
Diet		
Traditional	**21**	
Vegetarian	**1**	**26**
Western	**4**	
Alcohol consumption		
None	**12**	
Social drinker	**8**	**24**
Heavy	**4**	
Cigarette smoking		
None	**15**	
1 - 10/d	**7**	**24**
> 10/d	**2**	

*Detailed information was not available for all patients.

In all cases evaluation of the mental state, fundi, cranial nerves, cerebellum and posterior columns revealed no abnormalities. General examination revealed no specific abnormalities and, in particular, the patients did not show the stigmata of malnutrition.

Investigations

Plain radiographs of the dorsolumbar spine were followed by myelography and found to be normal in all cases. In all cases the cerebrospinal fluid was examined at the time of myelography. The results are summarized in Table IV. Immunoglobulin studies (including electrophoresis for oligoclonal immunoglobulins) were undertaken in 2 patients seen early in 1982. The results were normal.

TABLE IV. EXAMINATION OF CEREBROSPINAL FLUID

	No. of patients
Increased cell count	0
Protein (g/l)	
< 0,4	16
0,4 - 0,7	7
0,7 - 1,0	5
> 1,0	5
Raised globulin levels	11
Positive fluorescent treponemal antibody test	0

Haematological studies (haemoglobin level, mean cell volume, erythrocyte sedimentation rate) revealed no statistically significant abnormalities when compared with the results in controls at the hospital. A total eosinophil count of over 400 x 10^9/l was found in 12 patients, but in view of the relatively high incidence of eosinophilia in our general patient population this was not considered significant.

Serological tests for syphilis (VDRL and the fluorescent treponemal antibody test) were performed on the serum and CSF and were found to be negative in all cases.

Vitamin B_{12} estimation was carried out in 12 patients. The mean level was 438 pg/ml (range 125 - 840 pg/ml), the normal range in our laboratory being 200 - 1 000 pg/ml. Serum folate levels were all within normal limits.

Active pulmonary tuberculosis and urinary schistosomiasis were excluded in all cases by the usual routine screening methods.

Discussion and conclusion

The first question raised by these observations is whether these patients have sufficient points of similarity to warrant their description as a nosological entity, or whether they represent a random assortment of cases of paraplegia due to different unrecognized causes.

The cause of paraplegia can be difficult to diagnose, even given the most sophisticated equipment. In reviewing cases of 'spastic paraplegia of uncertain aetiology' seen at the National Hospital, Queen Square, London, Marshall[1] wrote: 'it is a chastening thought that, despite our best diagnostic endeavours, half of the group of 52 patients still defied diagnosis ten or more years after the onset . . . '. Of those patients with unexplained causes who ultimately came to autopsy, one-third were found to have multiple sclerosis (MS), one-third had cord compression from tumours or prolapsed discs and the remainder had a heterogeneous group of conditions.

Nevertheless, our patients have so many features in common that it is felt that they do represent a nosological entity. This conclusion is supported by the close similarity of these patients to those previously reported from Natal.[2]

This syndrome is characterized by spastic weakness of the lower limbs which is of insidious onset and associated with variable segmental sensory loss. It is confined to Blacks, almost all of whom come from rural areas where traditional dietary habits are the rule. The syndrome reflects a spinal cord disorder with a predilection for the corticospinal tracts and, to a variable extent, the lateral spinothalamic tracts. The exact pathological nature of the condition is not known as the condition is not fatal, and consequently autopsy studies have not been carried out. Recognition of the syndrome depends on the exclusion, as far as is possible, of known causes of spinal cord lesions which produce similar effects.

We also found that most of the patients became ambulant with the aid of physiotherapy although they were still disabled. Where follow-up has been possible the condition does not seem to have progressed to any significant degree, and no episodes of complete remission or relapse have been reported. The fact that the condition is self-limiting and sometimes improves to a limited extent tends to exclude causes of cord compression from the differential diagnosis, as does the uniformly negative myelography. Common systemic infections and infestations such as tuberculosis, syphilis and bilharzia may affect the spinal cord. In the previous series[2] the frequency of eosinophilia led to a suspicion of parasitic disease involving the spinal cord. Similar eosinophilia was found in the present study, but this probably reflects the parasitic load among this population. Tuberculosis and syphilis were excluded by the relevant investigations.

TABLE V. MAIN CORD SYNDROMES REPORTED PREVIOUSLY

Geographical area	No. of cases	Main clinical features	Supposed cause	Author
Natal, South Africa	41	Spastic paraplegia, minor sensory loss	?	Cosnett[2]
Witwatersrand, South Africa	61	Spastic quadriparesis, minor sensory loss, mental disturbance	Malnutrition, pellagra	Grieve *et al.*[4]
Central India	Endemic	Lathyrism, spastic paraplegia, no sensory loss	*Lathyrus sativus* in diet	Prasad and Sharan[5]
South India	45	Spastic quadriparesis, peripheral sensory loss	?	Mani *et al.*[6]
Jamaica	181	Spastic paraplegia, some posterior column loss, peripheral neuropathy	? Syphilis or yaws	Montgomery *et al.*[7]
Jamaica	25	Sensory ataxia, optic atrophy, nerve deafness	?	Montgomery *et al.*[7]
Tanganyika	7	Sensory ataxia, optic atrophy nerve deafness	Malnutrition	Haddock *et al.*[8]
Nigeria	375	Sensory ataxia, optic atrophy	Cyanide intoxication from cassava in diet	Osuntokun[9]

The family histories of our patients have not revealed a genetic predisposition to the disease. In Europe or North America these patients might have been diagnosed as having MS. It is recognized that a small proportion of patients with MS may lack a history of remission and relapse; however, it is unlikely that the patients in this series have MS, especially in view of the fact that the more common forms of MS are virtually unknown among South African Blacks.

Unexplained neurological disorders affecting the legs have been described in many tropical and subtropical countries.[3] Table V lists the main features of important examples of these. They fall into two groups: the spastic group (like the present series of cases) and an ataxic group. Where likely causes have been identified these are either malnutrition or toxic dietary factors.

The patients in our series did not show clinical stigmata of malnutrition. The normal Vitamin B_{12} and folate levels support this conclusion. The evidence seems to point more towards a toxic dietary factor. The occurrence of the syndrome in a single population group and its preponderance among those of rural origin with traditional dietary habits lends weight to this hypothesis. The clinical features of these patients from Natal resemble those of the patients with lathyrism in central India.[5] Lathyrism is caused by a toxic substance contained in the seeds of *Lathyrus sativus*, which is used as a staple diet, especially in times of famine. This legume is not grown for human consumption in Natal. It is, however, possible that similar toxins may occur in local foodstuffs or in herbs used for medicinal purposes.

In the final analysis, therefore, this syndrome of unexplained paraplegia appears to be a distinct nosological entity. Its aetiology may be multifactorial but, on circumstantial evidence, one or more toxic dietary factors are most prominent among the suspected causes.

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