MULTIPLE SCLEROSIS IN A SOUTH AFRICAN-BORN WHITE MAN AND WOMAN

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It has long been realized by South African doctors that disseminated, or multiple, sclerosis is extremely uncommon among the South African-born, although it is relatively frequently seen among immigrants from Europe.[1] In fact, there have been some South African physicians and neurologists who hold that multiple sclerosis never occurs among the South African-born who have not visited Europe. No authenticated case of multiple sclerosis has yet been described in a South African Bantu. For these reasons we thought it would be interesting to report on 2 South African-born Whites who were diagnosed during life as having multiple sclerosis and in whom the diagnosis was confirmed at autopsy.

CASE 1

History

C.J.W. was born at Ladybrand in the Orange Free State in 1906. He was Afrikaans-speaking and he was a bachelor who had never been abroad. By profession he was a primary teacher in the Orange Free State until 1945. He lived in the Dewetsdorp district until 1958 when he was admitted to the Oranje Hospital, Bloemfontein.

When he was 20 years old he had blurring of vision or actual blindness for a short time. In 1945, at the age of 39, he developed severe blurring of vision which lasted for 3 days and did not recur. This was followed by progressive mental deterioration so that he had to retire from his work as a teacher, but otherwise he was able to live a fairly normal life on his mother's farm. In 1950 he found difficulty in walking because of weakness in his legs, and he developed poor control of his bladder. In 1953 he suffered from severe paroxysmal right-sided trigeminal neuralgia which became so severe by 1955 that he was referred to Drs. Kerr, Allen and Mendelow at the Princess Nursing Home, Johannesburg.

The following is an extract of a report from the Princess Nursing Home, where he was investigated during August/September 1955:

'Right trigeminal neuralgia—2 years.
Incontinence.
Difficulty in walking—5 years and several months.
Mental retardation—10 years.

Pallor of both optic discs; no field defect.
Increased tendon reflexes and clonus present.
Plantar reflexes, normal.
Incoordination and ataxia, e.g. on finger, nose and heel-knee test.
Some loss of proprioception of both big toes.
Depression of vibration sense in both lower limbs.
Speech slurred.
Memory and powers of concentration poor.
Bilateral impairment of visual acuity and primary optic atrophy present.
Fine, non-sustained horizontal nystagmus.
Mild right lower facial weakness.
Excruciating paroxysmal pain of whole right face and forehead, precipitated by touching face just anterior to the right ear.

Air-encephalogram showed dilatation of ventricles and widening of sulci, suggesting a moderate degree of cerebral atrophy.

Blood count. A mild, normochromic anaemia with some anisocytosis, Hb. 12·7 G per 100 ml., colour index 0·92, RBC 4,400,000, and leucocytes 11,600 (polymorphs 67.5%, mononuclears 6%, lymphocytes 23%, eosinophils 2·5%, and mast cells 1%).

Sedimentation rate 11 mm. in 1 hour (normal 1 - 10 mm.).

Cerebrospinal fluid on 2 consecutive days: pressure 80 mm. water, WR negative, lymphocytes 1 - 3 per cu.mm., protein 50 - 90 mg. per 100 ml., sugar 56 - 66 mg. per 100 ml., and chloride 720 - 730 mg. per 100 ml.

Section of right auricular nerves and alcohol injection of right 5th ganglion successfully performed on 13th and 19th September 1955.

Result. Free of pain and complete anaesthesia of the 2nd and 3rd branch and depression of right corneal reflex. Residual urine 14 oz., hence indwelling Foley's catheter. Urine contained no porphyrin. Blood urea 33·3 mg. per 100 ml.'

After 1955 there was periodic improvement during which time he was able to manage the Native shop on his mother's farm. By April 1958, his mental state had again deteriorated and he was admitted to the Oranje Hospital, Bloemfontein.

Physical Examination on Admission to the Oranje Hospital, 1958

He was confused and his intimate habits were faulty. He was disorientated for time and subject to hallucinations—he heard voices speaking to him from the ceiling. He was incontinent of urine and required an indwelling Foley's catheter. He had bilateral temporal pallor of the optic discs and nystagmus. His speech was explosive with difficult articulation. He was not paralysed, but all his movements were ataxic. The deep tendon reflexes were increased. The abdominal reflexes were absent. Ankle and knee clonus were present in both legs. Both plantar responses were extensor. He could not stand or walk without assistance. The cerebrospinal fluid Serum Eagle test and the Serum Kahn test were negative. The cerebrospinal fluid was under normal pressure, but the protein was increased to 60 mg./100 ml.; 5 lymphocytes were present per cu.mm.

Shortly after admission he had 3 epileptic convulsions; he had no more attacks and remained mentally euphoric. He could give an account of himself and his family, but was only partially orientated. His memory was defective for recent events and vague for past events. He required the Foley's catheter for the first 3 months, after which he could empty his bladder naturally, although he suffered from frequency and urgency of micturition. From April 1959 until his death in December 1963 he showed a progressive mental deterioration. He was euphoric and his memory became markedly impaired. His physical state also deteriorated, and he again became incontinent of urine in April 1962, and incontinent of faeces in October 1962. For most of this period he was confined to a chair during his waking hours. In August 1963 he again had epileptic seizures and his general condition worsened steadily until December when he developed pneumonia from which he died.

Summary of Pathological Examination

Macroscopic examination of the brain showed numerous greyish plaques of apparent demyelination, most numerous in the sub-cortical white matter of both hemispheres (Fig. 1). Similar plaques were also observed in the pons (Fig. 2).

Microscopic examination showed these plaques to consist of clear-cut partial or complete demyelination (Fig. 3). In the pons, lesions were present in the tegmentum as well as in the basis pontis. Gliosis was prominent in all these lesions, but surviving nerve fibres traversing the lesions could be demonstrated.

The spinal cord was not submitted for examination.

The pathological features in this brain were entirely in keeping with the diagnosis of multiple sclerosis.

CASE 2

Abstract of History

Mrs. J.C. (SAIMR 13278/64), a White female aged 36 years at death, was born in Barkly West, Cape Province, and had never left the borders of the Republic. She had been treated privately by Dr. H. Isaacs who, in a personal communication, has indicated that he made the diagnosis of multiple sclerosis in the first half of 1960.[2] In 1962 she was first examined in the outpatient department of the Johannesburg General Hospital where a spastic paraplegia, weakness of the hands, paraesthesiae in hands and feet, and difficulty in swallowing were recorded. Fasciculation was observed and a diagnosis of amyotrophic lateral sclerosis was made. The last stage in her illness began in September 1964, when she suddenly collapsed at work and complained of severe headache. Examination showed a subarachnoid haemorrhage, atrophy of the lateral half of one optic disc and bilateral extensor plantar responses. Abducent nerve weakness was noted, and after some improvement she collapsed again and died about 2 weeks later.

Summary of the Pathological Examination

There was a fairly extensive subarachnoid haemorrhage and a large intracerebral haematoma, both of which could be traced to a ruptured saccular aneurysm of the left middle cerebral artery in the stem of the lateral sulcus. Some greyish areas were seen over the posterior surface of the upper part of the cord, and on section scattered greyish-white areas were seen in the parenchyma on both the right and left sides over much of the length of the cord. A single very small similar subcortical greyish area was noted in the left frontal area. Microscopically these areas were for the most part sharply defined foci of demyelination, not localized to anatomical tracts, associated with gliosis in nearly every instance and they contained axons (Figs. 4, 5). Other microscopic areas were present in a subependymal position in the occipital lobes and in the tegmentum of the pons (Fig. 6). One optic nerve showed myelin pallor. The character and distribution of the lesions are consistent with multiple sclerosis.

An unexpected finding was 2 rounded dark lesions of a different character in the vermis of the cerebellum, the larger measuring 1·1 cm. in diameter. They were formed of neoplastic glial cells, the one resembling a cystic astrocytoma and the other containing what appeared to be oligodendroglial elements separated by pools of albuminous fluid and red cells.

Fig. 1

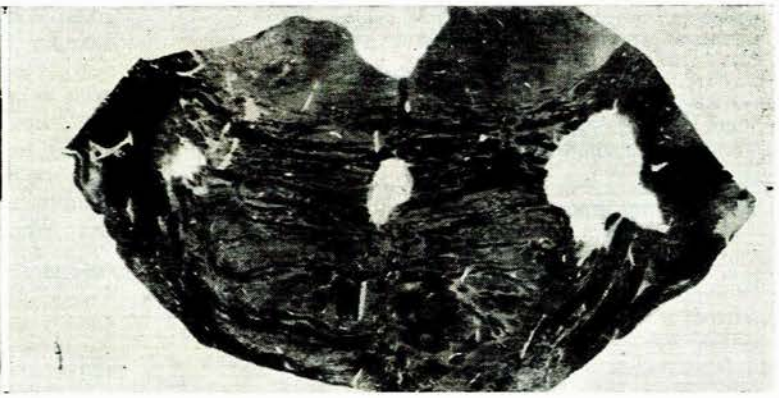

Fig. 2

Fig. 1. Typical sub-cortical demyelinated plaques (Kluver-Barrera). *Fig. 2.* Clear-cut demyelinated plaques in pons (Kluver-Barrera).

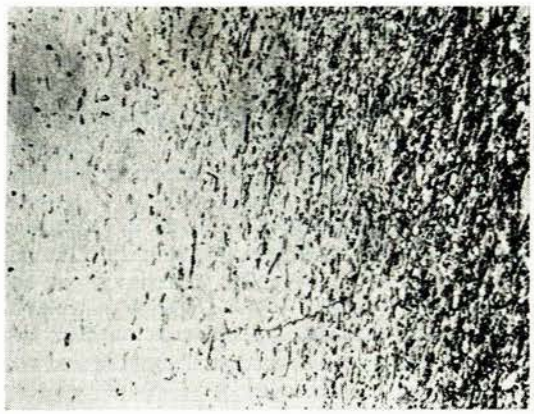

Fig. 3

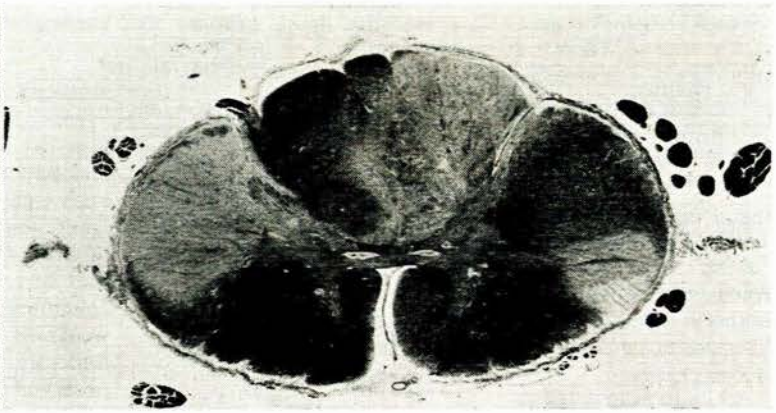

Fig. 4

Fig. 3. Edge of plaque showing complete demyelination (Kluver-Barrera). *Fig. 4.* Upper cervical cord showing areas of demyelination. Note involvement of part of the posterior horn of the grey matter on one side (Kluver-Barrera).

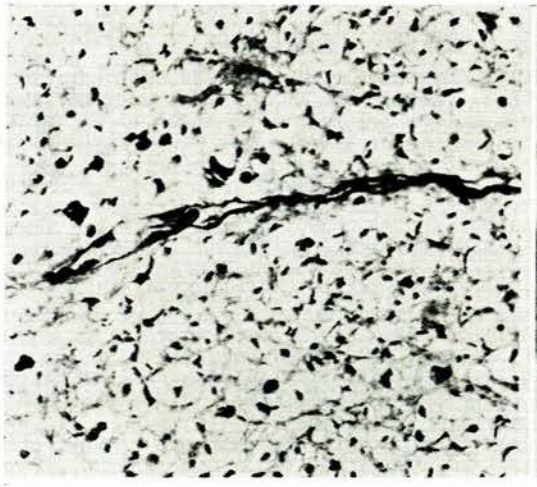

Fig. 5

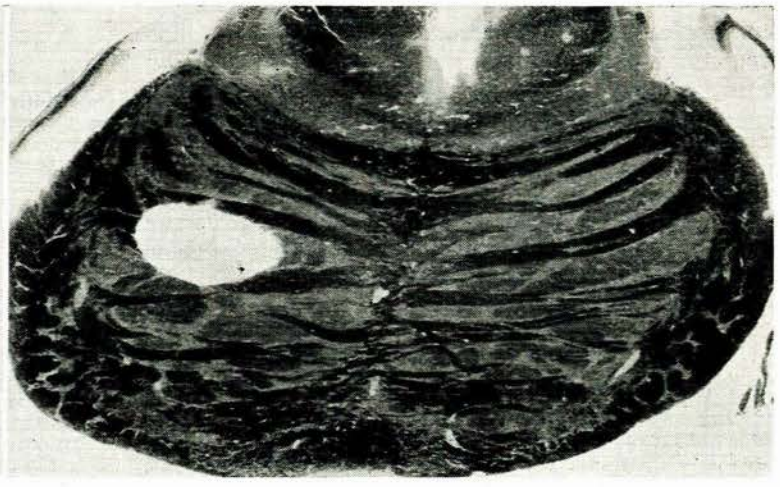

Fig. 6

Fig. 5. Persisting axons in one of the pale lateral areas (Bielschowsky x 480). *Fig. 6.* The faint background pattern of the nuclei pontis is visible in the sharply demyelinated area (Kluver-Barrera).

Comment

Clinically this patient would seem to fit best into the so-called spinal form of multiple sclerosis manifesting as a paraplegia without bladder involvement.[3] When the spinal forms are followed up for long enough, the upper part of the central nervous system is usually affected. The small lesions in the pons, hemispheres and optic nerve suggest that this was beginning to occur and the evolution of the disease was stopped abruptly by rupture of the cerebral aneurysm. The cerebellar neoplastic lesions are most unexpected and, not being in close proximity to a plaque, are probably only a coincidence; however, the association of glioma and multiple sclerosis has been noted before.[4] The cerebellar lesions and certain other clinical and pathological features will be fully described in another publication.

SUMMARY

The history, the reported clinical examinations and the autopsy findings all confirm that these South African-born Whites, who had not left South Africa and had lived all their lives in South Africa, suffered from multiple sclerosis.

We should like to thank Dr. Geoffrey Dean, of Port Elizabeth, and Dr. A. M. Lamont, the Commissioner of Mental Health, who encouraged us to publish this paper. Thanks are also due to Mr. E. Rosenberg, Drs. A. G. Hefer and Peter Jacobs for access to the case notes. The Director of the South African Institute for Medical Research has given permission to publish.

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