Short report

Multiple sclerosis presenting as chronic atypical psychosis

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SUMMARY Two patients with multiple sclerosis are reported in whom psychopathological symptoms were present for many years. An exact psychiatric classification of the clinical syndrome was not possible. At no time during the illness were abnormal neurological findings observed. Multiple sclerosis was proven following necropsy in one patient. In the second patient a possible diagnosis of multiple sclerosis was made. This patient was found to have multiple lesions in the white matter which were evident on MRI and in the isoelectric focusing (IEF) of cerebrospinal fluid oligoclonal bands were found.

Multiple sclerosis is usually diagnosed on the basis of the typical clinical course with dissemination in time and space. The difficulty of diagnosis lies in the great clinical variability of the disease. Discreet neurological symptoms are often overlooked or misinterpreted. The clinical picture is often masked by hysterical manifestations.¹ Although psychopathological features in multiple sclerosis have been reported,^{3,6,9,13,16} the presence of exclusively psychiatric symptoms is not generally considered to be indicative of multiple sclerosis and consequently is not usually further investigated.

The unusual clinical course of multiple sclerosis in two patients will point to the isolated occurrence of psychopathological symptoms in the disease and in addition the value of the new diagnostic possibilities, in particular MRI and CSF investigation of isoelectric focusing.

Patient 1

In 1958, two years following the death of his schizophrenic mother, a man aged 24 years stopped going to work and showed impairment in routine daily functioning. He occasionally became afraid and locked himself in his room,

searching the walls for secret entrances. He was examined in 1960 by a district medical officer and was described as mentally defective, totally dependent, and lacking initiative. During the first psychiatric hospitalisation, the patient complained of headache and insomnia. He continued to be totally dependent and slow in action. The patient was observed to have poorly defined orientation, indicating disturbances of visual cognitive performance. In contrast to the lack of both initiative and emotion, there was abnormal psychomotor behaviour. There were no abnormal neurological or EEG findings. There were no abnormal findings in the pneumoencephalogram, but a lymphomonocytic pleocytosis (20 cells/mm³) and a relative increase in globulin were found in CSF from the cisterna magna.

Despite the history, hebephrenia was diagnosed. An accessory encephalitis was suggested based upon the CSF findings, without a causal relationship being made to the psychopathological syndrome. The clinical picture improved slightly and CSF findings became normal after 4 weeks' steroid therapy.

The patient was admitted to various psychiatric hospitals from 1960 until his death in 1985, apart from being released twice, for a few months. He was initially described as lacking initiative, morose, obstinate, grumbling, at times aggressive and difficult to manage. He became more violent with fellow patients and nursing staff. The patient constantly asked to be discharged or given holiday leave. He attempted to escape at every possible opportunity. The patient continually claimed not to be mentally ill. The patient's family was not willing to care for him. Treatment with potent neuroleptics led to pronounced acute dyskinesia. The patient thereafter refused the medication and expressed fears of being poisoned. In 1970, the patient's complaints of visual disturbances and fears of going blind were regarded as paranoid. Acute psychotic epi-

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Received 18 November 1986 and in revised form 26 August 1987.

Accepted 29 August 1987

Fig 1 Case 1: Acute demyelination with perivascular inflammation and astrocytic gliosis in frontal white matter. (Klüver-Barrera $\times 100$.)

sodes of a mostly paranoid hallucinatory nature occurred repeatedly throughout. A severe hypokinetic-rigid syndrome developed during the long-term high dose neuroleptic therapy. In the last years of the illness, during an attempt to discontinue therapy, the patient developed increased agitation with fear, aggressive behaviour and megalomania. The patient died in such an agitated state due to acute right ventricular failure without apparent underlying clinical disease. The last psychiatric diagnosis was the residual phase of schizophrenia. There were no reports of abnormal neurological findings at any time, aside from the medication induced disturbances.

The post-mortem findings were of a typical multiple sclerosis with multiple larger and smaller areas of demyelination with pronounced gliosis and distinct perivascular lymphocyte infiltration in frontal white matter bilaterally. An approximately 1×1 cm large area of demyelination was also found in the pons (figs 1 and 2).

Patient 2

A previously healthy 42 year old woman first complained in November 1981 of feelings of fear, pressure in the breast, heaviness in the legs and sleep disturbance. The patient was hospitalised in a psychiatric clinic 2 weeks later. The patient

screamed and spoke in a confused way. There were alternating phases of psychomotor restlessness, aggression, insomnia, loud unintelligible monotone wailing with stuporous states. At times a hysterical reaction was considered. It was not possible to classify the patient's illness according to psychiatric criteria. There were no abnormal neurological findings, or in the EEG or CT.

Treatment with various neuroleptics and antidepressants did not effect the patient's clinical state. On the contrary, extra-pyramidal motor symptoms developed, lastly in the form of a hyperkinetic syndrome with distally accentuated irregular choreiform movements of the limbs. The psychopathological disturbances remained constant, apart from minimal fluctuation, over the years. New symptoms did not appear. Finally there were pronounced disturbances in drive and emotion.

MRI (spin-echo mode 0.5 tesla, fig 3) in August 1984, showed multiple signal intensive T-2 accentuated foci in the periventricular white matter of the cerebral hemispheres. A lumbar puncture yielded no abnormal CSF findings apart from evidence of oligoclonal bands on isoelectric focusing. A high dose steroid treatment did not improve the patient's clinical state. The number of foci in the MRI decreased, as well as the signal intensity.

Fig 2 Case 1: Inactive old plaque with complete loss of myelin, relative axonal preservation and gliosis in pons. No inflammation. (Klüver-Barrera $\times 250$.)

Discussion

Encephalitis, particularly involving the temporal lobe, limbic system and brain stem, presenting as psychopathological symptoms occur often and are individually difficult to differentiate from endogenous psychosis or neurotic disturbances.^{5 11 15 17}

Davison and Bagley reviewed the literature concerning psychiatric changes in multiple sclerosis in 1969.² Multiple sclerosis, in general, involves an emotional disturbance without a direct relationship to the duration of the disease. Good correlation does exist between the duration of the disease and intellectual deficits, which must be classified as residual symptoms of chronic encephalitis.¹² The correlation of defined psychopathological syndromes of differing grades with the disease attacks has only been rarely described and only in single case histories.^{6 9 13} Generally, neurological disturbances, indicating the diagnosis, develop during the course of the disease.

The course of the disease in our patients, which was not truly progressive or with definable attacks, and the absence of abnormal neurological findings, did not lead to initial diagnosis of multiple sclerosis. It

can, of course, not be excluded that transitory, discreet neurological disturbances, which occur often in multiple sclerosis, were either overlooked or misinterpreted.

Multiple sclerosis was definitely diagnosed in patient 1 following necropsy. Abnormal CSF, as found in multiple sclerosis, was already present during the first hospitalisation. The absence of neurological abnormalities was the only diagnostic information available at that time. Additional diagnostic methods, which could have identified *in vivo* multiple foci, were not then available. The later single occurrence of visual disturbance with fear of going blind was probably a correlate of a retrobulbar neuritis. Here again, additional diagnostic techniques could have clarified the clinical diagnosis. Important aspects, for instance the clearly pathological results in the CSF, were most probably ignored, in view of the clinically predominant behaviour disorder and the few acute neurological symptoms.⁷

Patient 2 initially presented as having masked depression. The quick change in the psychopathological findings led to the diagnosis of a neurotic-hysterical disturbance and also to more

Fig 3 Case 2: MRI spin echo scan showing multiple foci of T2 accentuated increased signal intensity in the white matter and in periventricular region.

detailed intensive diagnostics. MRI surprisingly indicated multiple foci in the white matter. This, taken together with the abnormal CSF, must be interpreted as a laboratory-supported definite multiple sclerosis according to the criteria of Poser *et al.*¹⁴

Neurological disturbances, which suggest classical multiple sclerosis, were absent in both patients. It was not possible to classify the disease states according to the individual psychopathological findings alone. Psychopathological symptoms, as described in temporal encephalitis,¹⁵ were found in both patients.

The non-specific psychopathological symptoms should be emphasised. The disturbance of initiative and mental confusion in patient 1 is reminiscent of the "depersonalisation" described by Strian.¹⁵ In patient 2, the quick fluctuating psychopathological syndrome resembled the descriptions of Matthews⁹ patient with multiple sclerosis and Strians' patients with temporal encephalitis. Both patients reacted very sensitively to neuroleptics, possibly another indication of organic brain damage.

Chronic progression with primarily neurological symptoms is not uncommon in multiple sclerosis. A benign course with clinically silent lesions discovered unexpectedly at necropsy are well known.⁴ Our patients demonstrate that multiple sclerosis can also present as a chronic psychiatric disease. Discreet neurological symptoms can therefore be concealed by the primary psychopathological syndrome or they can also be completely absent. In the past, the diagnosis

of multiple sclerosis was most frequently found to be in error in patients in large psychiatric hospitals,¹⁰ probably due to limited diagnostic facilities. Improved diagnostic certainty has evolved following the introduction of MRI and the use of isoelectric focusing for detecting oligoclonal bands in CSF.

The use of these methods in the future will lead to earlier diagnosis, as well as to a broadening of the clinical spectrum of this CNS disease. Psychiatrists must also utilise these new methods in order to diagnose, at an early stage, such uncommon forms of disease.

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