mic slow wave discharges on the right at 1–2 Hz, occasionally associated with spikes, and more clearly epileptiform activity on the left. Intraventricular diazepam abolished this activity on the left, but spared that on the right. The initial diagnosis was of pneumonia with complex partial status epilepticus. On the following day the EEG record was dominated by widespread, irregular, rhythmic complexes with a period of approximately two seconds. Unenhanced CT scan appearances had not, however, changed since 1988. A lumbar puncture was performed, but routine biochemical and cytological analysis of the CSF showed no abnormalities. The CSF was under a pressure of 24 cm of water and contained 52×10⁶/L leucocytes (93% lymphocytes) with normal CSF protein and normal CSF glucose ratio. A further lumbar puncture two days later showed worsening abnormality and treatment was started with intravenous Acyclovir, rifampicin, isoniazid and pyrazinamide. Over the following nine days her reflexes deteriorated and she died.

A polymerase chain reaction for the amplification of HSV-1 DNA was carried out post-mortem using the primer set described by Klenk et al. with the following conditions: 28 cycles of denaturation at 94°C for 30 sec, annealing at 50°C for 30 sec, extension at 72°C for 30 sec. The second PCR sample, taken two days before, was negative. Neither oligonuclear bands of total IgG nor antigen-specific oligonuclear bands were detected in either CSF sample. Viral culture and viral titers were negative in the first CSF sample; in the second sample viral culture was again negative but a weak IgG response to herpes simplex virus 1 was detected by ELISA.

Post mortem examination confirmed the presence of a right petrous meningioma. The brain was swollen and soft, with uncal and cerebellar tonsillar herniation. Temporal and insular cortex were involved in a marked meningocoeephaptitis, with necrotising venulitis and perivascular infil-

tration by lymphocytes and macrophages. Immunoperoxidase staining revealed a very striking positive reaction for herpes simplex virus antigens in neurons, macrophages, and many cells whose nature could not be identified.

This complex case emphasises the importance of prompt treatment with Acyclovir where there is clinical suspicion of the diagnosis of HSE. In general the diagnosis should be considered in any patient with fever and depression of consciousness: suspicion should be heightened by accompany-

Figure Second round PCR products, on ethidium bromide stained gel, showing a positive result in lane 3 (patient’s CSF) and lanes 9 and 10 (positive controls). Lane M contains molecular weight maker.

MATTERS ARISING

Accuracy of clinical diagnosis of idiopathic Parkinson’s disease

I read the paper by Hughes et al in the journal with much interest and wish to compliment them for their work.

The percentage of inaccurate clinical diagnosis of idiopathic Parkinson’s disease (IPD) in their study is identical to that which we reported last year. Their observations, however, are different from our study in several respects. For example, the largest subgroup of patients (43%) which were erroneously diagnosed as having IPD by Hughes et al had progressive supranuclear palsy (PSP), in contrast, all the necropsy proven PSP cases in our study were recogni-

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Hughes et al reply:
We thank Professor Raijput for his interest and contribution to the discussion of our study. It is unclear why progressive supranuclear palsy (PSNP) comprised such a high percentage of patients clinically misdiagnosed in our study, while in their series all necropsy cases of PSNP were recognized before death. A pure akinetic syndrome may be the common manifestation of PSNP. The UK Parkinson's Disease Society Brain Bank (PDSSB) receives donor tissue from Parkinsonian patients throughout the UK. Criteria for inclusion into the scheme, patients are examined annually by 70 neurologists and geriatricians associated with the Brain Bank and information is recorded according to a standard format. Despite the use of diagnostic criteria it is impossible to completely standardize diagnostic practice across such a group of assessor. The stage of disease when patients are examined is clearly important in studies of this type. Clinical diagnoses used in our present study were all made within 12 months of death, at the time of the last assessment, and during or after 1986. All patients were considered specifically to have PD rather than a less well-defined Parkinsonian syndrome.

We agree that no diagnostic criteria for PD are fool-proof and have subsequently analysed the clinical features of our cases in terms of their diagnostic value. By using selected criteria (asymmetrical onset, no atypical features, and no possible aetiology for another Parkinsonian syndrome) the proportion of PD cases identified was increased to 93%, but at the expense of excluding 32% of pathologically confirmed cases. Twelve of 100 cases of histologically confirmed PD examined at the PDSSB had atypical clinical features according to Brain Bank diagnostic criteria for this disease. More than half of these cases had no other associated neuropathological findings that could explain the atypical features. These findings suggest that studies based on consultant diagnosis of Parkinson's disease will include patients without the disease as well as excluding some who subsequently satisfy the histological criteria. As a result, results from clinical trials and epidemiological studies may be distorted.


Matters arising

BOOK REVIEWS
All titles reviewed here are available from the BMJ Bookshop, PO Box 295, London WC1H 9TE. Prices include postage in the United Kingdom. In some cases (usually for members of the British Forces Overseas), but overseas customers should add £2 per item for postage and packing. Payment can be made by cheque in sterling drawn on a United Kingdom bank, or by credit card (Mastercard, Visa or American Express) stating card number, expiry date, and your full name.


At the present time there is some fascinating progress in the definition and territorial demarcation of neurodegenerations causing dementia. The scale is rather split between some among clinicians and pathologists to identify new diseases such as corticobasal degeneration and the causes of frontal lobe degeneration, and primary progressive aphasia. In contrast, molecular genetics is tending to lump diverse phenotypes together in the prion disorders and familial Alzheimer’s disease. Real progress in other areas, notably Pick’s disease, is to some extent lacking. This condition is finding a more critical definition by the effect of an eponym, because some examples of non-Pick body Pick’s disease are undoubtedly other things, such as corticobasal degeneration. The remaining chapters in the book cover the neuropathology of unusual dementias, dementia and motor neurone disease and Lewy body dementia: so not, as you might have thought, small print causes of dementia as implied by the title, nor a fully comprehensive account of these disorders, but mostly areas of real progress and new knowledge.

The introduction provides a paragraph on the clinician’s approach to a patient with dementia. This is a useful summary, to which collection, storage and analysis of genetic and pathological material could be added. Many of these diagnoses remain neuropathological ones, and their genetic implications are still uncertain. In addition, peculiar phenotypes of these neurodegenerations can mimic almost any other. Future developments should justify this encouragement to obtain a post mortem diagnosis, if not for the family, for future patients and families. Storage of DNA will provide a valuable reference for many of the new and child and considers the question of ictal violence and the attribution of serious crime to epileptic activity.