Matters arising


Hughes et al reply:

We thank Professor Raiput for his interest and contribution to the discussion of our study.1 It is unclear why progressive supranuclear palsy (PSNP) comprised such a high percentage of patients clinically misdiagnosed as having Parkinson's disease (PD) in our study, while in his series all necropsy proven cases of PSNP were recognised before death.2 A pure akinetic syndrome was the second most frequent manifestation of PSNP.3 The UK Parkinson's Disease Society Brain Bank (PDSSB) receives donor tissue from Parkinsonian patients throughout the UK. Of those examined for the scheme, patients are examined annually by one of 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 clearly impossible to completely standardise diagnostic practice across such a group of assessors. The stage of disease when patients are examined is clearly important in studies of this type. The 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.4 By using selected criteria (asymmetrical onset, no atypical features, and no possible aetiology for another Parkinson 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.5 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, thus results from clinical trials and epidemiological studies may be distorted.


BOOK REVIEWS

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At the present time there is some fascinating progress in the definition and territorial demarcation of neurodegenerations causing dementia. How many positional dilemmas are encountered among clinicians and pathologists to identify new diseases such as cortico-basal 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 erection, because some examples of non-Pick body Pick's disease are undoubtedly other things, such as cortico-basal degeneration. The remaining chapters in the book cover the neuropathology of unusual dementias, dementia and motor neurone disease and Lewy body dementia: so far, 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 postmortem diagnosis, if not for the family, for future patients and families. Storage of DNA will provide a valuable future resource for work in new genetic studies. The chapter dedicated to an overview of neuropathology is valuable as these disorders are sufficiently uncommon to allow a critical mass of cases to be seen clinically and pathologically by one individual, and yet the pathology can be objectively compared by an experienced neuropathologist. Refreshingly, the pathology consists largely of updates, understandable for the non-specialist, and extensive reviews are not included.

I found minor inaccuracies in the book, mostly reflecting very recent progress in knowledge. This speed of progress is encouraging, and until recently was not a feature of neurodegenerations. This book is well worth reading by those wanting an update or introduction to the subject; this is certainly the case soon as the book will be superseded in due course.

WRG GIBB


It has been the experience of most neurologists on appointment to receive instructions from solicitors to examine and give an opinion by way of report on claimants undertaking civil action for injuries sustained in domestic, social or work situations.

Unfortunately many young neurologists when first approached, have had neither advice nor instruction on the preparation of reports and the implications of a medicolegal assessment. Nor have they been professionally offered the pitfalls which arise between writing a report and submitting to a cross examination in the high court. Often the prospect is disturbing. With the increasing civil litigation in the western world and a specific increase in claims of medical negligence, most of us will be invited to undertake assessments of claims. In some departments of neurology, a feature of the post graduate training is an introduction to this aspect of the neurologists work, but these are few.

For many years there has been a need for published advice. This is now provided by Dr Bell's book. Medico-legal assessment of head injury. He addresses the duties of the medical expert and the court's expectations. In a chapter almost certainly written for the lawyers, he describes the anatomy and pathology of the head and the major consequences of the brain damage. In his chapter on the syndromes of regional brain injury he finds space for a useful account of the effects of extension-flexion injury to the cervical spine and quotes some important figures and references to an acceptable estimation of prognosis. He assesses the literature on the prediction of post traumatic epilepsy both in the adult and child and considers the question of ictal violence and the attribution of serious crime to epileptic activity.