Progressive supranuclear palsy: neuropathologically based diagnostic clinical criteria

In their excellent retrospective clinicopathological study of 12 cases of progressive supranuclear palsy (PSP), Collins et al noted a variety of clinical signs and symptoms beyond those in the original description of this disorder. They proposed an algorithm for the clinical diagnosis of PSP, a definite diagnosis of which had been made during life only in eight of 12 of their patients. In principle, we agree with their clinical criteria and results, based on a retrospective clinicopathological study of 24 cases of PSP from the files of the Ludwig Boltzmann Institute of Clinical Neurobiology, Vienna, and the R Escourroule Neuropathology Laboratory, Paris. In this material, a definite diagnosis had only been made in 12 of 24 of the cases. Our diagnostic criteria were as follows: (a) onset over age 40; (b) progressive course of a non-familial disease; (c) duration less than 10 years; (d) postural instability or falls without specific etiology; (e) akinesia and rigidity; (f) supranuclear ophthalmoplegia including down gaze abnormalities; (g) dysarthria or pseudobulbar palsy; (h) frontal lobe-like symptoms; (i) lack of focal lesions on CT; (j) improvement or levodopa treatment. Almost all of these diagnostic criteria are identical to those used by Collins et al, except for reticulosis or dystonic arm, sitting “en bloc”, and Babinski’s signs, which were not seen in most of our patients. Based on these diagnostic items, we concluded that a clinical diagnosis of PSP was probable when nine of 10 criteria were present, whereas in the absence of two signs or symptoms, the diagnosis was considered “possible”. The retrospective evaluation of these criteria allowed identification of 88% of the cases; 18 being “probable”, and three “possible”. These data seem of interest, as in other recent postmortem series of PSP, only a small percentage fulfilled currently accepted clinical diagnostic criteria—for example, seven of 17 (41%)—whereas the remainder who lacked these criteria had alternative clinical diagnoses. In another recent series the clinical diagnosis of PSP had been made in eight of 13 histopathologically confirmed cases (61%); whereas 13 brains (54%) showed concomitant pathological changes of Alzheimer’s or Parkinson’s disease. In view of the clinical heterogeneity of PSP and some difficulties in the postmortem diagnosis of typical, atypical, and combined PSP—the last feature by the presence of typical neuropathological changes of PSP together with findings that are diagnostic of other neurologists—this is an evaluation of the sensitivity and specificity of clinical and neuropathological criteria of PSP in a larger series of histopathologically confirmed cases seems mandatory.

KURT A JELLINGER
CHRISTIAN BANCHER
Ludwig Boltzmann Institute of Clinical Neurobiology,
Lains-Hospital, Vienna, Austria
JEAN-JACQUES HALYW
MAURICE VERNY
R Escourroule Neuropathology Laboratory,
Inserm U 360, Hôpital de la Salpêtrière,
Paris, France


Collins et al reply:
We appreciate the comments of Jellinger et al and echo their appeal for a more comprehensive clinicopathological study of cases of PSP. It is reassuring that our findings are similar to those of other recent series quoted.1,2 We were also impressed by the wide range of clinical and pathological features in these cases and sought to introduce some order into the classification of PSP by restricting our analysis to cases only with typical histopathological features at postmortem. The resulting paradigm, however imperfect, provides a useful working framework in which to place patients with clinical features suggestive of PSP, until a more specific biological marker is available.

S J COLLINS
JE AHLSKOG
JE PARISSI
D M MARAGANORE
Mayo Clinic,
200 First Street Southwest,
Rochester, MN 55905, USA


Most of the big names in vascular dementia came together at the second symposium on Age and Ageing Disorders which took place in August 1993 in Stockholm. It was organised by the Gamla Tjänarinnor Foundation and the presentations form the 17 chapters of the book which appears as a special issue of Dementia. Vladimir Hachinski opens with a marvelous chapter. Having put vascular dementia on the map in the mid-1970s he now retreats and argues that the term has outlived its usefulness. Vascular is too generic and dementia too restrictive. The traditional concept of dementia is epitomized by SDAT; a disease of old age in which progressive memory failure is not associated with global cognitive impairment. Furthermore, the condition is irreversible. Vascular disease rarely, if ever, gives rise to such a condition. The cognitive deficit is multi-focal and not necessarily progressive. The patients usually have deficits in motor function which are not seen in SDAT. For Hachinski the concept of vascular dementia is redundant but the range and variety of cognitive impairment due to various vascular causes demands further description and investigation. After all, vascular disease is potentially reversible and to some extent reversible.

Erikjuntius describes the 1993 NINDS-AIREN criteria for vascular dementia. Here dementia was defined as a cognitive decline from a previously higher level of functioning and manifested by impairment of memory in two or more other cognitive domains. The different types of ischaemic lesions which can cause dementia include multiple large vessel stroke, single strategically