Clinical diagnosis and diagnostic criteria of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome)

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Summary. Progressive supranuclear palsy (PSP) is characterized clinically by supranuclear gaze palsy, neck dystonia, parkinsonism, pseudobulbar palsy, gait imbalance with frequent falls and frontal lobe-type dementia. In the advanced typical case, when supranuclear gaze palsy and other main features are present diagnosis is relatively easy. Diagnostic problems, though, are frequent in the early stages due to the variable clinical presentation and in those atypical cases in which gaze palsy does not develop or that present as a severe dementing disorder or as an isolated akinetic-rigid syndrome. In this review we summarize the clinical features of PSP and emphasize those aspects helpful in the differential diagnosis with Parkinson’s disease and other motor and cognitive disorders that can pose difficult diagnostic problems. Clinical diagnostic criteria are also discussed and modifications of those currently in used are proposed.

Introduction

Although some reasonably convincing cases of what is now called progressive supranuclear palsy (PSP) had been described before 1964 it was in this year when Steele et al. (1964) clearly established PSP as a separate nosological entity different from other disorders presenting with supranuclear opthalmoplegia, pseudobulbar palsy and parkinsonism and from those associated neuropathologically with neurofibrillary tangles, such as post-encephalitic Parkinson disease and the Parkinson-Dementia complex of Guam. Steele et al. (1964) called the disorder progressive supranuclear palsy, a term that vaguely describes the patients problems but which has survived, particularly in its abbreviated form, PSP, the test of time, unlike other proposed terms. The cardinal manifestations of PSP were clearly outlined by Steele et al. in their original paper (Steele et al., 1964) and reviewed by Steele (1975) and others (Golbe et al., 1988; Kristensen, 1985; Lees, 1987) and include supranuclear gaze palsy, pseudobulbar palsy, prominent neck dystonia, parkinsonism, poor equilibrium and falls and subcortical dementia. The clinical picture is fairly uniform in the advanced, typical case and while each individual feature can be encountered in other
neurological conditions the presentation on a given patient of several of these cardinal features is almost unique for PSP.

Difficult diagnostic problems can occur, though, when trying to diagnose PSP in the early stages. Indeed, the mean duration of PSP before diagnosis in Kristensen's review (1985) of 325 cases collected from the literature was 3.9 years and other authors, such as Pfaffenbach et al. (1972) report an even longer average delay in the diagnosis of PSP after symptom onset (4.5 years). This delay in diagnosis is undoubtedly due to lack of familiarity of physicians with the clinical syndrome, but also to the heterogeneous presentation of the disorder in the early stages and in particular to the fact that the supranuclear ophtalmoplegia which remains the most characteristic clinical sign of the disease, tends to appear late after the onset of symptoms. Brusa et al. (1979), for example, in their review of 75 reported cases with neuropathological confirmation found that in half of these cases, slowness and limitation of both conjugate voluntary and vergence movements occurred 2 to 4 years after the onset of any other symptom. In the early stage then a firm clinical diagnosis is difficult, frequently impossible and one is often satisfied in ruling out other neurological disorders if possible and establishing a diagnosis of possible or probable PSP which only the passage of time will clarify. Unfortunately there are no biological markers or neuroimaging techniques that offer great help with this difficult diagnostic task.

In the brief review that follows we summarize the main clinical features of PSP emphasizing their relative diagnostic value and insisting upon those clinical signs that are helpful in the differential diagnosis with Parkinson disease (PD), and other motor and cognitive disorders that can pose on occasion diagnostic problems. A list of other less common or clinically significant features is also provided in Table 1. We also briefly comment on existing diagnostic criteria and propose a new set of them.

Clinical manifestations

Oculomotor disturbances

The neurological abnormality considered most important for the diagnosis of PSP is the presence of a supranuclear ophtalmoplegia (SNO). Down gaze palsy has to be present for the gaze palsy to be considered of significance since upward gaze impairment can be encountered in PD and other neurological disorders and is present in some normal elderly individuals. Gradually the gaze disturbance involves both vertical and horizontal movements and a complete supranuclear ophtalmoplegia will eventually occur in more than half of the patients. Typically movements to command are more affected than those to pursuit and by definition the oculocephalic movements are preserved. Other oculomotor and eyelid abnormalities occur in PSP (Kristensen, 1985; Lepore et al., 1988; Troost and Daroff, 1977) and are summarized in Table 2. Some of them, such as lid lag, with markedly reduced blink rate or apraxia of lid opening, are characteristic of PSP.