PAPER

Visuospatial functions in atypical parkinsonian syndromes

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Objectives: Visuospatial deficits have been occasionally reported but never systematically studied in atypical parkinsonian syndromes. The interpretation of existing studies is complicated by the possible influence of motor and frontal executive deficits. Moreover, no attempt has been made to distinguish visuoperceptual from visuospatial tasks. The aim of the present study was to assess visuoperceptual and visuospatial abilities in three atypical parkinsonian syndromes while minimising the influence of confounding variables.

Methods: Twenty patients with multiple system atrophy (MSA), 43 with progressive supranuclear palsy (PSP), and 25 with corticobasal degeneration (CBD) as well as 30 healthy age matched controls were examined with the Visual Object and Space Perception Battery (VOSP).

Results: Visuospatial functions were intact in MSA patients. PSP patients showed mild deficits related to general cognitive decline and the severity of oculomotor symptoms. The CBD group showed the most pronounced deficits, with spatial tasks more impaired than object based tasks. Performance on object based, but not spatial, tasks was related to general cognitive status. The extent of the visuospatial impairment could not be predicted from disease duration or severity.

Conclusion: Visuospatial functions are not consistently impaired in atypical parkinsonian syndromes. The degree and pattern of impairment varies across the diseases, suggesting that the observed deficits could have a different neural basis in each condition. The distinction between the object based (''ventral stream'') and the space oriented (''dorsal stream'') processing might be useful in the interpretation of visuospatial deficits in parkinsonian syndromes, especially in CBD.

In contrast to the extensive studies of visual cognition in Parkinson's disease and dementia with Lewy bodies,[1] little is known about this domain in the atypical parkinsonian syndromes: progressive supranuclear palsy (PSP), multiple system atrophy (MSA), and corticobasal degeneration (CBD). In MSA, some studies reported no significant visuospatial abnormalities,[2] while others suggested a decline with the disease progression.[3] In PSP, early studies based on the analysis of a small number of patients reported deficits in visual search, scanning, and attention.[4–6] A recent study comparing PSP with Alzheimer's disease (AD) patients found the prevalence of visuospatial deficits in PSP (60%) to be substantially higher than in AD (38%).[7] CBD is often associated with difficulties in visuospatial processing[8–10] but their frequency and pattern remain unclear.

The assessment of visuospatial functions in atypical parkinsonian syndromes has to overcome a wide range of confounding variables. Rigidity, bradykinesia, tremor, micrographia, ataxia, and apraxia make tasks involving drawing or copying difficult. The psychomotor slowing has an impact on timed tasks, dysarthria, and speech apraxia on verbal responses. Deficits in attention, task switching, and working memory influence the results of any complex testing. While no single test can eliminate all the confounding variables, the Visual Object and Space Perception Battery (VOSP)[11] offers a set of tasks that minimise the influence of motor, attentional, mnestic, and executive functions. It also distinguishes between object and space processing, a distinction of considerable anatomical interest since object processing is associated with temporal (''ventral stream'') and space processing with parietal (''dorsal stream'') lobe function.[12]

METHODS

We examined 88 consecutive patients with atypical parkinsonian syndromes (20 with the clinical diagnosis of MSA,[13] 43 of PSP,[14] and 25 of CBD[15]) and 30 age matched controls recruited through the Medical Research Council (MRC) volunteer panel. The global cognitive status was assessed with Mini-Mental State Examination (MMSE).[16] As PSP and MSA are both associated with imbalance, we assessed their severity using the five level gait/balance staging component from the PSP rating scale.[17] In CBD imbalance occurs later, so we based our severity judgement on a four level scale assessing extrapyramidal features and apraxia (1, involvement of one limb; 2, two limbs; 3, all limbs; 4, wheelchair bound). The eye movement abnormalities in PSP were assessed as following: 1, vertical gaze slowing; 2, vertical gaze limitation; 3, vertical gaze paresis, horizontal gaze involvement; 4, complete paresis of vertical and horizontal gaze.[18]

All patients and controls were administered the VOSP.[11] It consists of a visual screening tests and four tests of object recognition and space. The stimuli are presented as black and white pictures and drawings; the required response is single word naming or pointing to the correct item. No drawing or copying is required. We selected three object based (incomplete letters, silhouette naming, and object decision) and three spatial (dot counting, number location, and cube analysis) tasks. We omitted the progressive silhouettes, a test based on two items only and the position discrimination, a subtest with a chance level of 50%, leading to a performance at the ceiling level in the majority of patients. The data were analysed using SPSS (SPSS Inc, Chicago, IL, USA).

Abbreviations: AD, Alzheimer's disease; CBD, corticobasal degeneration; MMSE, Mini-Mental State Examination; MSA, multiple system atrophy; PSP, progressive supranuclear palsy; VOSP, Visual Object and Space Perception Battery.

RESULTS

There were no significant age differences between the three disease groups and controls: MSA 65.9 (SD 8.2), PSP 69.1 (SD 5.6), CBD 67.7 (SD 7.3), controls 68.2 (SD 7.2). The average disease duration in the MSA group (5.1 (SD 2.8)) was significantly longer than in PSP (3.5 (SD 2.3)) and CBD (3.7 (SD 1.7)) ($p<0.05$), but the disease severity of MSA and PSP was identical (2.9 (SD 1.0)). In the CBD group, where a different scale was used, the disease severity was 2.3 (SD 0.8). Expressed as the percentage of the highest score the severity of all three diseases was between 57.5 and 58%.

A series of one-way ANOVAs revealed significant group effects for all subtests (table 1) but with variable patterns of intergroup differences on post hoc pairwise comparisons. There was no significant difference between the controls and the MSA patients on any of the subtests. The PSP patients were impaired in comparison to controls and MSA patients on ''silhouette naming'' and ''number location'' only. In addition, they were impaired in comparison to controls on ''dot counting''. The CBD group was worse than controls and the MSA group on every subtest except ''object decision'' and worse than the PSP group on ''incomplete letters'', ''dot counting'', and ''cube analysis''.

We then categorised the individual patients' scores as impaired, or preserved, relative to the published cut-off scores for VOSP subtests. The differences between the groups are illustrated in figure 1. In the MSA group the proportion of impaired patients varied between 0 and 5% and never exceeded 10%. The PSP group showed a higher proportion of impaired patients overall, but only for ''silhouette naming'' and ''number location'' did this exceed 33%. In the CBD group the percentage of patients impaired varied between 28% and 52% percent, with the spatial tests more often impaired (44%–52%) than the object based (28–38%).

Finally, we examined the relation between the clinical data and VOSP performance in the PSP and CBD groups (MSA patients, not impaired in 85% of cases, were not analysed further). In the PSP group a significant correlation was found between the degree of VOSP impairment (number of subtests failed) and the MMSE (rho = 0.480, $p = 0.01$), disease duration (rho = 0.271, $p = 0.04$), and severity of eye movement abnormalities (rho = 0.550, $p<0.001$). In contrast, disease severity did not correlate with VOSP results. MMSE correlated with impairment on both object based (rho = 0.314, $p = 0.02$) and space based (rho = 0.484, $p = 0.001$) tests, although the latter correlation was stronger. The degree of eye movement abnormalities also correlated with the performance on object (rho = 0.378, $p = 0.006$) and space based (rho = 0.330, $p = 0.002$) tasks. In the CBD group only MMSE (rho = 0.531, $p = 0.005$), but not disease duration or severity correlated with the VOSP score. However, when examined separately, only the object (rho = 0.603, $p = 0.01$) but not the space based tasks correlated with MMSE. All other correlations were non-significant.

DISCUSSION

Our results demonstrate that visuospatial deficits are differentially distributed in atypical parkinsonian syndromes. No visuospatial impairment was detected in the MSA group, which is particularly remarkable given that our sample included patients with long disease duration, high levels of physical disability, and evidence of general cognitive dysfunction (MMSE<23). In neuroanatomical terms our data suggest that extensive basal ganglia pathology, as documented in MSA,[19] is not always associated with visuospatial impairment.

Table 1 Group analysis of VOSP subtests (mean and standard deviation) in the three patient groups and in the control group

Subtest (max score)	Controls	MSA	PSP	CBD	F
Incomplete letters (20)	19.2 (0.8)	19.4 (0.9)	17.9 (3.2)	14.9 (6.5)*†‡	7.91
Silhouette naming (30)	21.4 (2.9)	20.9 (3.6)	17.2 (5.7)*†	14.2 (6.4)*†	11.45
Object decision (20)	16.9 (2.3)	18.6 (2.2)	17.1 (2.6)	15.4 (4.3)†	4.43
Dot counting (10)	9.9 (0.3)	9.8 (0.4)	8.6 (2.2)*	6.9 (3.1)*†‡	13.03
Number location (10)	8.9 (2.8)	9.6 (1.1)	7.1 (2.7)*†	5.5 (3.9)*†	8.67
Cube analysis (10)	9.3 (1.5)	8.9 (1.8)	8.0 (2.1)	5.1 (3.4)*†‡	17.4

*Impaired relative to controls.
†Impaired relative to MSA patients.
‡Impaired relative to PSP patients.
In all six subtests the differences between the groups are significant with a $p<0.01$.

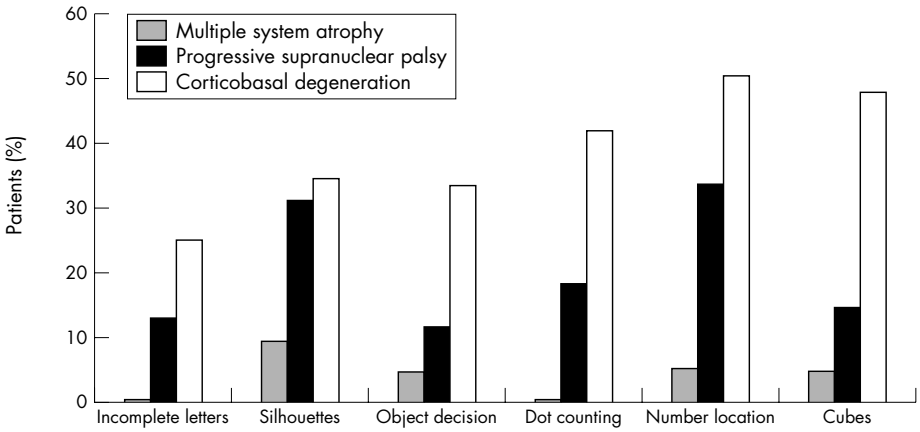

Figure 1 Percentage of patients impaired on VOSP subtests.

The impairment in the PSP group was practically confined to two subtests: ''silhouette naming'' and ''number location''. The first of them was the only subtest requiring naming. As the distorted figures are not immediately recognisable, the usual strategy used by most controls is to produce several possible answers and then select the most likely one. The subtest can, therefore, be influenced by a reduced verbal output and generation of concepts, documented in PSP patients through their pervasive reduction in verbal fluency.[7 10] This interpretation is supported by the good performance of the PSP group on ''object decision'', a task assessing a similar aspect of visual object processing, but not requiring a verbal response. The other impaired subtest, ''number location'', was the only subtest requiring a vertical shift of attention—difficult for PSP patients because of the vertical gaze palsy and the visual scanning deficits preceding it.[6] Apart from these two tasks the PSP patients performed remarkably well, including difficult tasks (''cube analysis') sensitive to visuospatial dysfunction in AD.[20] The extent of impairment was related to the MMSE, disease duration, and degree of oculomotor impairment but not to overall disease severity. The observed deficits are likely, therefore, to reflect more general cognitive decline, related to the severity of the oculomotor but not motor symptoms.[18]

By far the greatest impairment was observed in CBD patients. This cannot be sufficiently explained by a general cognitive decline alone: many VOSP impaired patients had normal MMSE. In contrast, normal VOSP results have been documented in the context of a severe dementia[21] suggesting that the two tests can dissociate. Moreover, whereas in the PSP group the spatial and object based tasks were equally affected and both correlated with the MMSE score, the spatial deficits in the CBD group were more pronounced and did not correlate with the MMSE. We suggest, therefore, that this pattern reflects an early involvement of the ''dorsal stream'' with its anatomical substrate in parietal lobe pathology.[22] The visuospatial deficits might be further aggravated by impairment in number knowledge.[23] Both types of deficits have been associated with parietal lobe dysfunction[24] and further studies are needed to determine the exact relation between them.

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