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Short Communication

Pronounced Impairment of Everyday Skills and Self-Care in Posterior Cortical Atrophy

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Abstract. Posterior cortical atrophy (PCA) is a neurodegenerative syndrome characterized by progressive visual dysfunction and parietal, occipital, and occipitotemporal atrophy. The aim of this study was to compare the impact of PCA and typical Alzheimer's disease (tAD) on everyday functional abilities and neuropsychiatric status. The Cambridge Behavioural Inventory-Revised was given to carers of 32 PCA and 71 tAD patients. PCA patients showed significantly greater impairment in everyday skills and self-care while the tAD group showed greater impairment in aspects of memory and orientation, and motivation. We suggest that PCA poses specific challenges for those caring for people affected by the condition.

Keywords: Caregivers, early onset Alzheimer's disease, psychology, questionnaires, self-care, self-report

INTRODUCTION

Posterior cortical atrophy (PCA) is a neurodegenerative syndrome characterized by progressive decline in visuospatial and visuoperceptual skills, literacy and praxis; with parietal, occipital, and occipitotemporal atrophy. Age of onset is typically in the 50 s or early 60 s; while the syndrome is most commonly caused by Alzheimer's disease (AD), cases due to dementia with Lewy bodies and corticobasal degeneration have been described [1, 2].

In contrast to the amnestic predominance of typical AD (tAD), presenting features in PCA commonly include difficulties with visually-guided behaviors such as driving, reading, or locating objects, while insight and episodic memory remain relatively spared [3]. It is likely that these cognitive impairments limit patients' ability to realize, adapt, and monitor interactions with their visual and physical environment. Our characterization of how PCA impacts upon everyday life is informed by clinical experience and patient anecdotes, but there has been little work to quantify or directly compare functional abilities between PCA and tAD.

The Cambridge Behavioural Inventory (CBI) is a carer questionnaire that evaluates a range of everyday skills and neuropsychiatric symptoms. Distinct profiles have been demonstrated for AD, behavioral-variant frontotemporal dementia, and Parkinson's disease [4]. There is high test-retest reliability [5], and a good correlation between its neuropsychiatric items and the Neuropsychiatric Inventory [6]. Since its inception, it has been revised (CBI-R) to reduce the number of items from 81 to 45, while retaining the most informative items for comparing tAD with frontotemporal dementia [7].

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Here, we present the CBI-R profile of PCA and investigate whether it can discriminate between the impact of PCA and tAD upon everyday abilities and neuropsychiatric symptoms.

MATERIALS AND METHODS

Participants

The CBI-R was completed for 42 PCA patients (recruited at the Dementia Research Centre) and 85 tAD patients (data obtained from Dementia Research Centre and Neuroscience Research Australia).

All patients met the respective clinical criteria [2, 8–10]. Participants without a contemporary Mini-Mental State Exam score (MMSE) ($n = 9$), with a low MMSE (<6; $n = 4$), or no disease duration estimate ($n = 11$) were excluded. 32 PCA and 71 tAD patients remained. The majority of respondents were spouses (PCA 91%, tAD 87%). The remainder were other relatives or carers. 50% of PCA respondents were male; 66% were male in tAD.

All PCA patients underwent detailed neurological assessment. While the underlying pathology is uncertain, 10/32 (31%) had molecular pathology markers (5 cerebrospinal fluid, 5 positive amyloid (Florbetapir) PET scans performed as part of another study) that were supportive or compatible with AD pathology. Symptoms indicative of non-AD pathology were present in 7/32 PCA patients (22%; e.g., hallucinations, REM sleep disorder). The project was approved by the NRES Committee London-Queen Square and carried out in accordance with the Declaration of Helsinki.

Stimuli and procedure

The CBI-R contains 45 questions covering 10 domains (number of questions per domain given in brackets): memory and orientation (8), everyday-skills (5), self-care (4), abnormal behavior (6), mood (4), beliefs (3), eating habits (4), sleep (2), stereotypic and motor behaviors (4), and motivation (5). Participants responded by marking whether each item occurred 'never', 'a few times per month', 'a few times per week', 'daily', or 'constantly'.

Data analysis

Responses of 'daily' or 'constantly' were categorized as severe. Logistic regression was carried out on the severe/not severe categorization. Age, MMSE, gender, and disease duration were included as covariates. Separate analyses were carried out at the domain-level and single-item level.

RESULTS

Groups were matched for age (mean [SD]: PCA = 64.7 [8.3], tAD = 67.3 [7.5], $p = 0.11$) and disease duration (mean [SD]: PCA = 4.5 [2.0], tAD = 4.3 [2.3], $p = 0.72$) but not MMSE (mean [SD]: PCA = 16.4 [4.8], tAD = 21.6 [5.1], $p < 0.001$).

The mean proportion of severe deficits in each group and domain is shown in Fig. 1. A significantly higher proportion of PCA than tAD patients were severely impaired in everyday skills ($p < 0.001$) and self-care ($p < 0.001$). By contrast, there were trends for a higher proportion of severe impairment in the tAD than PCA group in stereotypic and motor behaviors ($p = 0.053$) and motivation ($p = 0.067$). The proportion of patients with severe impairments did not differ between groups in the remaining domains. In memory and orientation, both groups had high, but similar, rates of impairment (tAD 51%, PCA 43%). In all other domains, both groups had only a small proportion (<30%) with severe deficits.

In the analysis of individual items, 12 items showed significant differences between groups (see Table 1). PCA patients showed a higher proportion of severe impairments on 7 items from the everyday skills and self-care domains, plus two additional items (in the sleep and eating-habits domains). tAD patients showed a higher proportion of severe impairments on 3 items from the memory and orientation, and motivation domains.

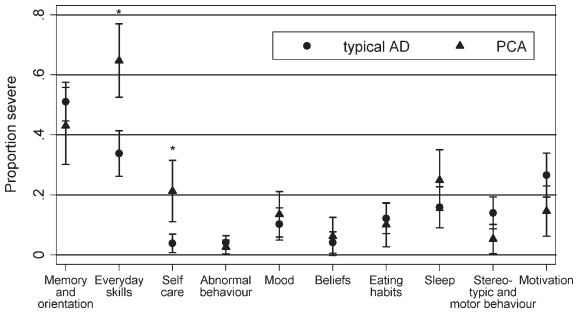

Fig. 1. Mean proportion of severe deficits in each domain of the CBI-R in the typical AD and the PCA patient groups (adjusted for age, MMSE, gender, and disease duration). Bars show 95% confidence intervals. Notable differences are more severe deficits in everyday skills and self-care in PCA patients, and a trend towards more severe deficits in stereotypic and motor behavior, and motivation in tAD patients. Statistically significant group differences are indicated with an asterisk.

Table 1
Individual items in the CBI-R in which there was a significant difference between the proportion of patients rated as showing a severe impairment in the PCA and tAD groups. No shading denotes PCA worse than tAD; grey shading denotes tAD worse than PCA

CBI-R individual items		
Domain	Question	*p* value
Everyday skills	Has difficulties writing (letters, Christmas cards, lists, etc.)	<0.001
	Has difficulties using the telephone	0.002
	Has difficulties making a hot drink (e.g., tea/coffee)	0.003
	Has problems handling money or paying bills	0.007
	Has difficulties using electrical appliances	0.025
Self-care	Has difficulties dressing self	<0.001
	Has problems bathing or showering self	0.042
Sleep	Sleeps more by day than before (cat naps, etc.)	0.020
Eating habits	Table manners are declining, e.g., stuffing food into mouth	0.035
Memory and orientation	Has poor day-to-day memory	0.007
	Asks the same questions over and over again	0.014
Motivation	Fails to maintain motivation to keep in contact with friends or family	0.034

The analysis was repeated with groups matched for MMSE (PCA $n = 32$, MMSE = 16.44; tAD $n = 32$, MMSE = 17.31), yielding similar results. Differences in everyday skills ($p = 0.003$) and self-care ($p = 0.006$) remained significant. With the exception of sleep (PCA greater impairment than tAD in repeated analysis; $p = 0.006$), group differences in the remaining domains were not statistically significant consistent with the original analysis.

Association of CBI-R total scores with MMSE and disease duration

Greater CBI-R score was significantly correlated with lower MMSE in tAD ($R = -0.28$, $p = 0.02$) but not PCA ($R = 0.07$, $p = 0.70$). Neither tAD ($R = 0.19$, $p = 0.12$) or PCA patients ($R = 0.15$, $p = 0.41$) showed a significant association between disease duration and total CBI-R score.

DISCUSSION

This study demonstrates the ability of the CBI-R to differentiate between everyday functional abilities in PCA and tAD. PCA patients showed greater impairment in everyday skills and self-care, while tAD patients showed trends toward greater impairment in stereotypic and motor behaviors, and motivation. At the item level, PCA patients were more likely to have severe impairments in everyday skills and self-care but also in sleep (more sleep during the day) and eating habits (declining table manners), while a greater proportion of tAD patients had severe impairments in day-to-day memory, repetitive questioning, and motivation to maintain social contacts.

The majority of functional deficits highlighted in PCA could be related to visuospatial and visuoperceptual impairments (e.g., writing, using the telephone, making hot drinks) or posterior cortical-dependent impairments in calculation (handling money/paying bills) or praxis (using electrical appliances). The 'declining table manners' item was designed to capture behavioral disinhibition but may reflect practical, visual- and praxis-mediated eating changes (e.g., eating with fingers owing to problems manipulating cutlery and perceiving location of food on the plate). The only item on which PCA patients were rated worse than tAD patients without an obvious posterior cortical functional explanation was the apparent increase in sleep. More detailed analysis of sleep would be required to explore this finding. No patients suffered from obstructive sleep apnea; while it is possible that patients with dementia with Lewy bodies underlying PCA contribute to disruption of sleep, differences were observed only for excess sleep during the day, not disturbed sleep at night.

With regard to neuropsychiatric symptoms, the results point toward relatively spared motivation in PCA, at least in terms of motivation to maintain contact with friends and family. This may relate to what carers describe as a maintained sense of purpose. Differences in motivation between PCA and tAD have not been investigated previously, but patients with tAD have been noted to have high levels of apathy [11, 12] which may relate to the motivation domain in the current study [5]. Our results suggest that PCA patients may have relatively lower apathy, warranting assessment using more detailed measures.

It is notable that while tAD patients were more severely impaired on 2/8 memory and orientation items, the groups did not differ in the memory and

orientation domain overall, both patient groups showing a high proportion of severe deficits compatible with previous findings in tAD [4]. One would have expected PCA patients to have relatively preserved memory consistent with the clinical criteria [2, 8]. This discrepancy may relate to the fact that episodic memory in PCA is relatively spared but not necessarily normal, and that the memory and orientation domain of the CBI-R includes behaviors which could be influenced by visual deficits (such as losing objects and becoming confused in unusual surroundings).

Although carer-rated questionnaires such as the CBI-R are indirect measures, they may offer utility for measuring longitudinal progression over the whole disease course; particularly as visits to specialist centers and detailed neuropsychological or clinical assessment may become infeasible at later stages. An improved understanding of everyday functional abilities and neuropsychiatric symptoms as the disease progresses is critical to ensuring optimal clinical management and care planning.

While PCA patients in this study met syndromic criteria, it is likely that multiple pathologies are represented; further studies are required to examine the effect of pathology on CBI-R profile in PCA. Respondent characteristics may also influence questionnaire completion, therefore the lack of age and education data for respondents in this study is a limitation.

In conclusion, striking deficits in everyday skills and self-care highlight the particular challenges that face PCA patients and those who care for them. We hope that this work is a first step toward a better understanding of these difficulties, and will inform future examination of the differences in care needs, prognosis and impact upon patient and carer quality of life between visual and amnestic presentations of AD and related diseases.

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