COMMENT & CRITIQUE

Unilateral neglect in a patient diagnosed with frontotemporal dementia and parkinsonism linked to chromosome 17

Frontotemporal dementia and parkinsonism linked to chromosome 17 (FTDP-17) is characterised clinically by dominantly inherited cognitive decline, behavioural changes and motor impairment (1). Herein, we report a patient with FTDP-17 who presented with prominent hemispatial neglect, a syndrome that has not been reported in previous studies.

A 58-year-old right-handed man was diagnosed with FTDP-17 and a subsequent sequence analysis revealed a P301L mutation in the tau gene (*MAPT*). He had a 10-year history of apathy and behavioural disturbances and developed left-sided parkinsonism after seven years. Rigidity and tremor were initially levodopa-responsive. Neurological assessment revealed left-sided rigidity and postural tremor, vertical nystagmus and primitive reflexes. Electroencephalography was normal. Head magnetic resonance imaging showed a right temporal lobe atrophy with temporal horn enlargement. Cerebral blood flow measured by rCBF SPECT with ^{99m}Tc-ECD showed right hypoperfusion of the frontal lobe (52–68% of cerebellar perfusion), inferior and anterior parts of the right temporal lobe and posterior parietal lobe including the parieto-occipital area.

On initial neuropsychological examination performed 10 years after symptomatic disease onset, the patient displayed severe executive deficits with disinhibition and echopraxis, suggestive of atypical subcortical dementia. Over the following 6-month period, global cognitive function assessed with the Mini Mental State Examination (2) declined from 28/30 to 24/30. He scored 82/100 on the revised version of the Addenbrooke's Cognitive Examination (ACE-R) (3) and 98/144 on the Mattis Dementia Rating Scale (4). A unilateral neglect syndrome was suspected based on a history of

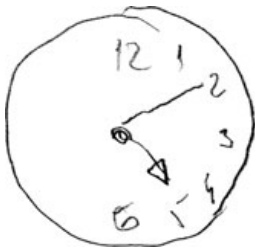

Fig. 1. Clock drawing task from ACE-R (3).

left-sided neglect during bathing, shaving and dressing, and on lateralised abnormalities observed when performing the clock-drawing test from ACE-R (Fig. 1). Subsequently, a comprehensive neglect-oriented test battery was administered, which consistently indicated unilateral neglect.

The Schenkenberg test revealed a significant 6.77 mm deviation to the right on average. Abnormalities were also observed using a visual search trial (5), during which 20 target objects are put in the centre of a rectangular board, one at a time. Each time, the patient is instructed to identify the same object as the target as quickly as possible. Additional targets are randomly distributed in all four quadrants (five in each quadrant). The longest reaction times occurred in the upper left quadrant (mean = 42 s), similar reaction times in the lower left and upper right quadrants (mean = 16 s and 18 s respectively), and the shortest reaction times for the lower right quadrant (mean = 9 s). In a letter cancellation task from the Behavioral Inattention Test, he made three omission errors in the first, second and third columns. In the scattered letter cancellation task, he made five omission errors in the left lower quadrant and two omission errors in the left upper quadrant. In all paper-pencil tasks, reversed order of execution (from the right side to the left) was observed. The Functional Neglect Assessment Battery (FNAS) (5) was administered to assess the impact of neglect symptoms on daily life function. The FNAS battery includes tasks referring to personal space (e.g. shaving, using a comb and putting on gloves and glasses) and peripersonal space (e.g. dealing cards, laying the table and scanning a newspaper in search of symmetrically distributed headlines). Results showed significant impact of symptoms in the category of personal space. In writing a short dictated text, the patient had a slight tendency to neglect the left side of the paper. There was a progressive deviation of the text to the right. In a reading task, only one word on the left was omitted.

Hemispatial neglect has occasionally been reported in patients with frontotemporal dementia, particularly those with corticobasal degeneration (CBD) (6–9); however, it has not been highlighted in FTDP-17. As observed in CBD, our patient presented with neglect on the left side associated with contra-lateral brain atrophy and hypoperfusion (6,8,9). Neglect in our patient was more pronounced in personal space and less evident in peripersonal space, in contrast with most common clinical observations (10).

This report shows that unilateral neglect symptoms may affect the results of cognitive tests that focus on executive dysfunction and utilise visuospatial components (like the maze or tower tasks). Moreover, asymmetrical neglect of personal hygiene in FTDP-17 patients may stem from hemispatial dysfunction, and should not be interpreted as a psychiatric symptom. We therefore suggest that patients diagnosed with FTDP-17 should undergo testing for unilateral neglect, especially if right-sided asymmetry is observed on neuroimaging.

Emilia J. Sitek[1,2], Ewa Narożańska[2], Jarosław Sławek[1,2], Dariusz Wieczorek[3], Bogna Brockhuis[4], Piotr Lass[4], Mirosława Dubaniewicz[5], Barbara Jasińska-Myga[6,7], Matt Baker[8], Rosa Rademakers[8], Zbigniew K. Wszołek[6]

https://doi.org/10.1111/j.1601-5215.2009.00367.x Published online by Cambridge University Press

[1]Department of Neurological and Psychiatric Nursing, Medical University of Gdansk, Poland;
[2]Department of Neurology, St. Adalbert Hospital, Gdańsk, Poland;
[3]Department of Rehabilitation, Medical University of Gdansk Gdańsk, Poland;
[4]Department of Nuclear Medicine, Medical University of Gdańsk, Poland;
[5]Department of Radiology, Medical University, Gdansk, Poland;
[6]Department of Neurology, Mayo Clinic, Jacksonville, Florida, USA;
[7]Department of Neurology, Aging, Degenerative and Cerebrovascular Disorders, Medical University of Silesia, Katowice, Poland; and
[8]Department of Neuroscience, Mayo Clinic, Jacksonville, Florida, USA

Dr Emilia J. Sitek
Department of Neurology, St. Adalbert Hospital, Al. Jana Pawla II 50, 80-462 Gdansk, Poland.
Tel: 0048 58 768 46 61;
Fax: 0048 58 340 92 90;
E-mail: emiliasitek@amg.gda.pl

Acta Neuropsychiatrica 2009: 21:209–210

DOI: 10.1111/j.1601-5215.2009.0367.x

References

1. Foster NL, Wilhelmsen K, Sima AAF, Jones MZ, D'Amato CJ, Gilman S. Conference Participants. Frontotemporal dementia and Parkinsonism linked to chromosome 17: a consensus conference. Ann Neurol 1997;**41**:706–715.
2. Folstein MF, Folstein SE, McHugh PR. "Mini-mental state". A practical method for grading the cognitive state of patients for the clinician. J Psychiatr Res 1975;**12**:189–198.
3. Mioshi E, Dawson K, Mitchell J et al. The Addenbrooke's Cognitive Examination Revised (ACE-R): a brief cognitive test battery for dementia screening. Int J Geriatr Psychiatry 2006;**21**:1078–1085.
4. Mattis S. Mental status examination for organic mental syndrome in the elderly patient. In: Bellack R, Karasu B, eds. Geriatric psychiatry. New York: Grune & Stratton, 1976;77–121.
5. Wieczorek D, Jodzio K. Comprehensive unilateral neglect assessment in brain-damaged individuals. Stud Psychol 2002;**40**:151–171.
6. Mitani K, Uchihara T, Tamaru F, Endo K, Tsukagoshi H. Corticobasal degeneration: clinico-pathological studies on two cases. Rinsho Shinkeigaku – Clin Neurol 1993;**33**:155–161.
7. Sato M, Nagata K, Yokoyama E, Hirata Y. Corticobasal degeneration: a combined clinical, PET and pathological study. Rinsho Shinkeigaku – Clin Neurol 1996;**36**:
293–299.
8. Kleiner-Fisman G, Black SE, Lang AE. Neurodegenerative disease and the evolution of art: the effects of presumed corticobasal degeneration in a professional artist. Mov Disord 2003;**18**:294–302.
9. Nagaoka K, Ookawa S, Maeda K. A case of corticobasal degeneration presenting with visual hallucination. Rinsho Shinkeigaku – Clin Neurol 2004;**44**:
193–197.
10. Robertson IH, Halligan PW. Spatial dimensions of neglect. In: Robertson IH, Halligan PW, eds. Spatial neglect-a clinical handbook for diagnosis and treatment. Hove: Psychology Press, 1999;26–34.