A longitudinal study of wo patients with slowly progressive behavioural variant of frontotemporal lobar degeneration

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BACKGROUND AND OBJECTIVE

Behavioural changes with gradual onset associated with impairment in social/occupational functioning may be an early manifestation of Fronto-Temporal Dementia (FTD).
The present study was aimed at investigating two patients with a history of behavioural changes associated with a slowly progressive cognitive decline..

CASE 2

In 2008, a 74-year-old woman, at the age of about 66 years, gradually started to show behavioural changes (apathy, social withdrawal, oppositional behavior). Subsequently, she started to show mild cognitive disturbances (forgetfulness, difficulties in temporal orienting). In 2010, MRI scan showed cortical atrophy, more marked in perisylvian regions bilaterally. In 2010, neuropsychological testing showed impaired performance on tasks of executive functioning and episodic memory. In 2011, SPECT showed perfusion abnormalities in the frontal cortex bilaterally and in the left temporal cortex (Figure 3-4). So far, a quite slow progression of her cognitive impairment was observed (Table).

CASE 1

In 2005, at the age of 62, a 72-year-old man gradually started to show irritability and anxiety. Two years later, the patient showed mild depression and apathy. In February 2007 and November 2009, he underwent MRI scans, which showed mild cortical atrophy, mainly affecting the frontal lobes. In 2007, neuropsychological assessments did not detect any abnormality. Since 2008, the patient became impulsive in some purchases. In 2010, behavioural disinhibition appeared and the patient started to show episodes of verbal (and, occasionally, physical) aggressiveness. Since 2011, he started to show difficulties in planning and carrying out complex daily living activities. Since 2007 up to 2011, repeated SPECT showed perfusion abnormalities in frontal and temporal cortical areas bilaterally, more markedly on the left (Figures 1 and 2)
Since 2008 up to 2012, an impaired cognitive performance was detectable only on tasks sensitive to frontal lobe dysfunction (Table).

TABLE AND FIGURES

TABLE		CASE 1		CASE 2	
Neuropsycological Task/ADL Scale	Cut-off	2007	2012	2010	2014
MMSE	23	29	25	22	17
Raven's Progressive Matrices '47	18.96	31 (30.4)	31 (30.4)	24.0 (26.0)	13.0 (15.6)
Rey immediate recall	28.52	44 (45.3)	31 (32.3)	11 (15.0)	12 (17.9)
Rey delayed recall	4.69	10 (10.6)	2 (2.6)	0 (1.3)	0 (1.9)
Digit Span forward	7+-2	6	7	5	4
Digit Span backward	5+-2	5	5	3	3
Corsi's span forward	7+-2	7	4	Not carried out	2
Corsi's span backward	5+-2	5	5	Not carried out	2
Letter verbal fluency	17.35	27 (23.9)	21 (17.9)	21 (24.2)	12 (15.9)
Semantic verbal fluency	12	19	14	Not carried out	Not carried out
Stroop interference: time	<31.67	30.5 (25)	45 (38.25)	Not carried out	Not carried out
Stroop interference: errors	<2.84	1 (0.5)	0.5 (0)	Not carried out	Not carried out
mWCST: correct criteria	>36 T	10	4 (42T)	Not carried out	Not carried out
mWCST: nonperseverative errors	> 39 T	25	10 (49T)	Not carried out	Not carried out
mWCST: perseverative errors	> 46 T	3	5 (32T)	Not carried out	Not carried out
Temporal rule induction: no of criteria	NA	Not carried out	Not carried out	3 out of 3	1 out of 3
Temporal rule induction: total score	≤ 15	Not carried out	Not carried out	30 (31.5)	40 (41.5)
Copying of drawings	7.18	9 (8.8)	12	10 (10.5)	4 (4.6)
Copying of drawings with landmarks	61.85	66 (65.7)	70	65 (65.7)	57.0 (58.0)
Objects naming	28	30	30	29	15
Action naming	26	-	28	Not carried out	Not carried out

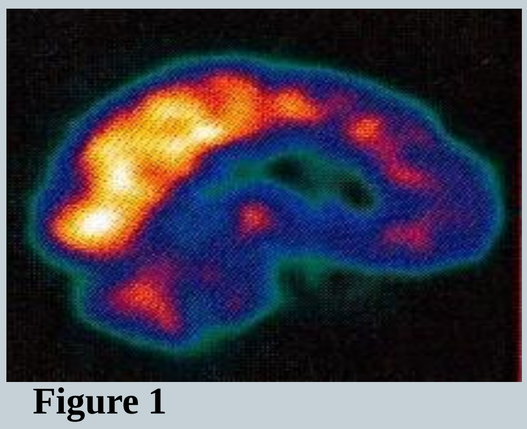

Figure 1

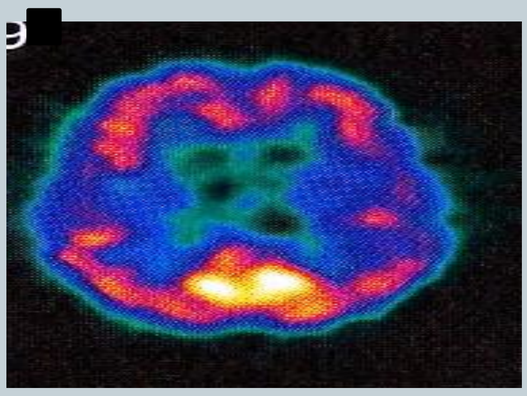

Figure 2

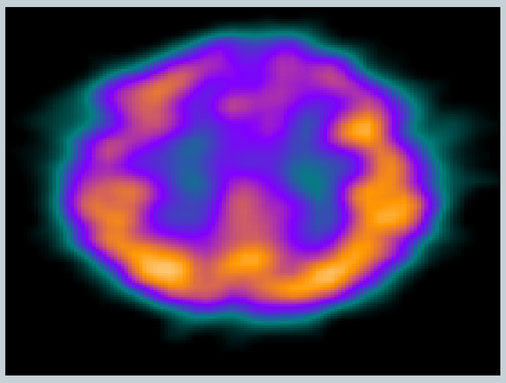

Figure 3

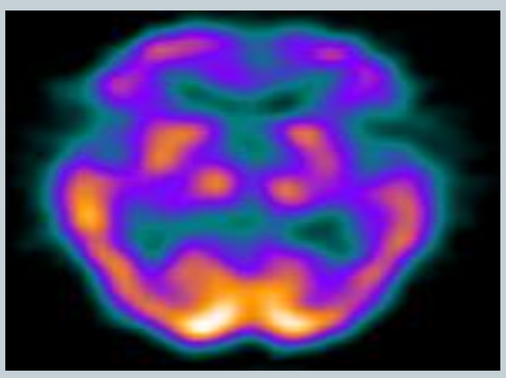

Figure 4

DISCUSSION AND CONCLUSIONS

In both patients, clinical data were consistent with a clinical diagnosis of behavioural variant of FTD with slow progression, a not common variant of FTD. The clinical and neurobiological features of such syndromic variant of FTD are still poorly known and need further investigation.

Reference:
(1) Hornberger M, Piguet O, Kipps C, Hodges JR.. Executive function in progressive and nonprogressive behavioral variant frontotemporal dementia. *Neurology.* 2008 Nov 4;71(19):1481-8