

# Late appearance of visuo-perceptual difficulties in a patient with long-standing cognitive impairment due to a vascular pathology: an unusual case of PCA or an unusual manifestation of vascular pathology?



Giordano Lacidogna<sup>1</sup>, Giulia Paroni<sup>2</sup>, Davide Scripa<sup>2</sup>, Antonio Daniele<sup>1</sup>

<sup>1</sup>Institute of Neurology, Catholic University, Rome, Italy; <sup>2</sup> Geriatric Unit & Gerontology-Geriatric Research Laboratory, Department of Medical Sciences, Institute of Care and Scientific Research Casa Sollievo della Sofferenza, S. Giovanni Rotondo, Italy

## Objective

Visuo-perceptual difficulties may appear in various disorders and are a remarkable feature in Posterior Cortical Atrophy (PCA), a syndrome characterized by progressive decline in higher-visual object and space processing due to neurodegenerative pathology affecting posterior cortical areas. The present study was aimed at investigating a man who presented with attentional difficulties and difficulties in remembering recent events, and developed visuo-perceptual difficulties 5-6 years after the onset of cognitive decline.

## Case report

In 2003, a 66-year-old man affected by arterial hypertension and mild carotid artery stenosis started to show difficulties in remembering recent events and in spatial orientation, associated to attentional difficulties. In 2006, he started to show mild Parkinsonian symptoms (limb rigidity and akinesia, upper limb tremor) and in 2007 a treatment with levodopa was started. Up to 2011, Parkinsonian motor symptoms did not significantly worsen over time. Up to about 2008, cognitive impairment did not show a notable progression, as suggested by repeated neuropsychological testing. In about 2008, the patient gradually started to show visuo-perceptual difficulties (difficulties in recognizing objects, difficulties in estimating the distance of objects around him) in the absence of remarkable ophthalmological abnormalities. Visuo-perceptual difficulties slowly progressed over the following 4 years. He underwent a longitudinal neuropsychological assessment, including the Visual Object and Space Perception (VOSP) battery.

## Results

In 2005 and 2008, MRI scans showed mild cortical/subcortical atrophy and several small areas of increased signal intensity in periventricular and subcortical white matter on T2-weighted images.

In 2007, neuropsychological assessment showed normal performance on the MMSE (28/30), while the patient was very slightly impaired on tasks of verbal and spatial short-term memory, abstract reasoning and copying of drawings.

In July 2010 (Table), an impairment was detected on tasks of the VOSP assessing recognition of visual stimuli and assessment of spatial relationships. In April 2012, neuropsychological assessment showed a widespread cognitive impairment, mainly characterised by impaired performance on tasks involving presentation of visual stimuli.

In 2006, 99mTc-ECD SPECT showed hypoperfusion in parietal areas bilaterally and in left temporal areas, while DAT-scan SPECT showed bilaterally a reduced uptake in the striatum, mainly involving the left putamen (Figure 1)

In May 2009 and in October 2010 (Figure 2), 99mTc-ECD SPECT showed hypoperfusion in parietal and temporal areas bilaterally and in left occipital areas.

Test	Score range	December 2007	July 2010	April 2012
MMSE	0-30	28	21 (20.86)	20 (19.86)
RAVLT immediate recall	0-75	47 (50.1)	30 (33.1)	13 (16.1)
RAVLT delayed recall	0-15	10 (11.2)	7 (8.2)	2 (3.2)
RAVLT recognition: accuracy	.50-1	0.95	0.92	0.71
Digit span forward & backward	n.a.	4 / 4	5 / 3	4 / 3
Corsi' span forward & backward	n.a.	4/3	Unable to carry out	Unable to carry out
Double barrage: accuracy	.50-1	1.0	Unable to carry out	Unable to carry out
Double barrage time	n.a.	94"	Unable to carry out	Unable to carry out
Copying of drawings	0-12	7 (6.9)	0 (0)	1 (0.9)
Copying of draw. with landmarks	0-70	60 (60)	6 (6)	24 (24)
Raven Matrices '47	0-36	16 (16.1)	7 (7.1)	11 (11.1)
Phonological verbal fluency	n.a.	22 (18.6)	13 (10.6)	Not done
Categorical verbal fluency	n.a.	14	6	9
Naming of objects	0-30	20	23	25
Stroop interference time	n.a.	22 (14.25)	Unable to carry out	Unable to carry out
Stroop interference errors	0-36	5 (4.25)	Unable to carry out	Unable to carry out
VOSP: Screening test		Not done	15	Not done
Incomplete letters	0-20	Not done	7	Not done
Silhouettes	0-30	Not done	9	Not done
Objects decision	0-20	Not done	6	Not done
Progressive Silhouettes	0-20	Not done	11	Not done
Dot Counting	0-10	Not done	4	Not done
Position discrimination	0-20	Not done	9	Not done
Number Location	0-10	Not done	Unable to carry out	Not done
Cube Analysis	0-10	Not done	5	Not done

Table : performanve on neuropsychological tasks since 2007 up to 2012.

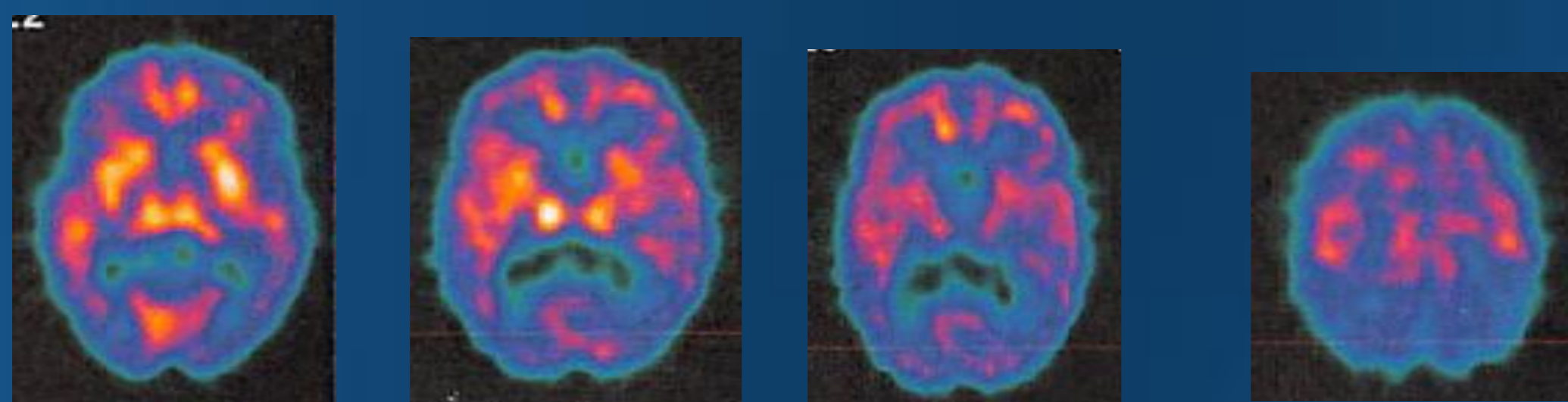


Figure 2: 99mTc-SPECT: hypoperfusion in temporo-parietal areas bilaterally and left occipital areas

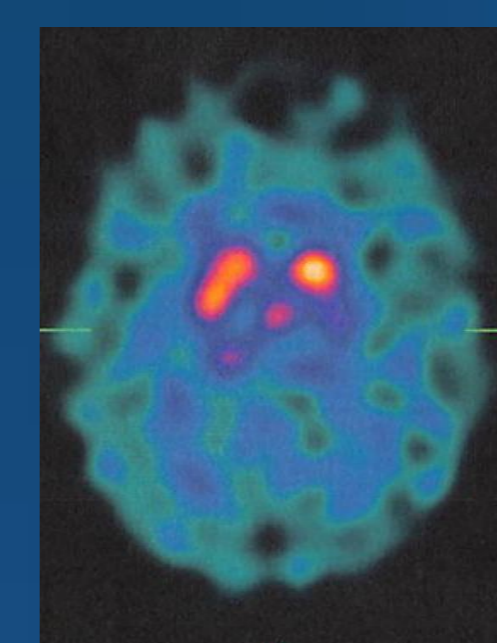


Figure 1: 123I-FP-CIT-SPECT: reduced uptaked in the striatum bilaterally

## Discussion and conclusions

Since in this patient with a vascular pathology visuo-perceptual difficulties emerged 5 years after the onset of cognitive decline, the formulation of a clear-cut clinical diagnosis might be challenging. It seems unlikely that such visuo-perceptual difficulties of this patient might simply be due to his vascular pathology, while we might more likely hypothesize the subsequent clinical appearance of a neurodegenerative pathology. We cannot exclude in this patient the hypothesis of a PCA which might have appeared some years after the vascular pathology: However, notwithstanding with the absence of clinical evidence for visual hallucinations and fluctuating cognition, the association of such pattern of cognitive decline and parkinsonism may suggest the alternative diagnostic hypothesis of a possible dementia with Lewy bodies, also supported by the results of DAT\_scan.

### References.

1. Crutch SJ, Schott JM, Rabinovici GD, et al. Shining a light on posterior cortical atrophy. *Alzheimers Dement*. 2013 Jul;9(4):463-5.
2. Crutch SJ, Lehmann M, Schott JM, Rabinovici GD, Rossor MN, Fox NC. Posterior cortical atrophy. *Lancet Neurol*. 2012 Feb;11(2):170-8.
3. Migliaccio R, Agosta F, Bartolomeo P, et al. Brain networks in posterior cortical atrophy: a single case tractography study and literature review. *Cortex*. 2012 Nov-Dec;48(10):1298-309.