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Visuospatial Deficit and Apraxia in Posterior Cortical Atrophy (PCA): a Single Case Follow-up Study

Lucio D'Anna*, **Pier Luigi Dolso, Simone Lorenzut, Anna Perelli, Barbara Zanchettin, Mariarosaria Valente and Gian Luigi Gigli**

Centre for Neurodegenerative Disorders, Neurology Clinic, University of Udine Medical School, Udine, Italy

***Corresponding author:** Lucio D'Anna, Centre for Neurodegenerative Disorders, Neurology Clinic, University of Udine Medical School, Udine, Italy, Tel: 393492530016; E-mail: luciodanna@hotmail.it

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Abstract

Posterior cortical atrophy (PCA) is a rare neurodegenerative syndrome characterized by insidious onset and selective, gradual decline in visuospatial and visuoperceptual skills.

We report a typical case of a patient with posterior cortical atrophy who presented initially with visuospatial deficit and apraxia. The aim of this report is to present this rare dementia subtype as a relevant differential diagnosis in respect to other neurodegenerative syndromes.

Introduction

Posterior cortical atrophy (PCA) is a rare neurodegenerative syndrome characterized by insidious onset and selective, gradual decline in visuospatial and visuoperceptual skills. Benson et al. in 1988 [1] for the first time described this rare but disabling dementia syndrome, in which visuo-spatial and visuo-perceptual impairments are the main clinical manifestations, often associated with alexia, prosopagnosia, environmental disorientation, features of Balint's syndrome and Gerstmann's syndrome [2]. Memory and verbal fluency are relatively well-preserved. Age at onset is typically before 65 years old [3].

Alzheimer's disease (AD) seems to be the most common underlying cause of PCA, however it has been described in associations with Huntingtons' disease, Lewy body dementia, Creutzfeldt-Jakob disease, cortico-basal degeneration and subcortical gliosis [4].

We report a typical case of a patient with PCA who presented initially with visuospatial deficit and apraxia. The aim of this report is to present this rare dementia subtype as a relevant differential diagnosis in respect to other neurodegenerative syndromes.

Case Summary

A 60-year-old right-handed woman was admitted to the Centre for Neurodegenerative Disorders, University of Udine, Italy, presenting progressive dyspraxic difficulties, deficits in writing and visuoperceptual problems since the last eight months.

At the age of 40 years, she was diagnosed as affected by breast tumour and underwent surgical intervention, followed by chemotherapy. Since the age of 45 has been treated for hypothyroidism.

Her family history was negative for any neurodegenerative condition, psychiatric disease or substance abuse. Her parents died at the age of 76 and 69 years, respectively, without cognitive deficits and his 77-year-old sister was still living and in good health.

The patient, who completed only the fifth grade of the elementary school, retired at the age of 58 years from her occupation as an industrial worker. Two years later, her husband reported the onset of mood deflection. In the last eight months she has progressively lost her ability to draw and write and developed problems with dressing, washing, using cutlery and handlings tools.

At admission, the neurological examination revealed signs of left visual neglect, optic ataxia and ocular apraxia. No alien limb phenomenon was documented. She scored 24/30 at Mini-Mental State Examination (MMSE).

Neuropsychological testing revealed severe ideomotor and ideational apraxia and visuospatial problems. The copy of a simple figure was impossible. Writing was impaired. Executive functions were compromised and a clear neglect was evident. Memory and fluent speech was intact, with a clear difficulty in naming tasks (Table 1).

Laboratory tests were within normal range. Structural neuroimaging demonstrated bilateral cortical atrophy, mainly located in the occipital and parietal lobes (Figure 1). Brain structural changes were confirmed by Single Photon Emission Computerized Tomography (99mTc SPECT), and extensive hypoperfusion in the parieto-occipito-temporal regions, mainly affecting the right hemisphere, was documented (Figure 2). Cerebrospinal (CSF) analysis was consistent with AD pattern, with decreased Ab42 (426 pg/mL, cutoff >500 pg/mL) and increased Tau (607 pg/mL, cut-off<350 pg/mL) and phospho-Tau (71 pg/mL, cut-off<60 pg/mL, as suggested by manufacturer, Kit-ELISA: Innotest Phospho-Tau) levels. Electroencephalography showed theta waves on posterior regions, bilaterally. In agreement with current clinical criteria, a diagnosis of PCA was made [5,6]. Namely, it was defined as biparietal variant according to the Alladi et al. classification [7]. The patient started therapy with transdermal rivastigmine (4.6 mg/24 h). At one-year follow-up, the neuropsychological examination showed that visuospatial praxis, writing and executive functions were significantly worsened. Language and verbal memory, largely preserved in the previous examination, were also significantly worsened (Table 1 and

Figure 3A-3C). The patient had become totally dependent in her daily living activities.

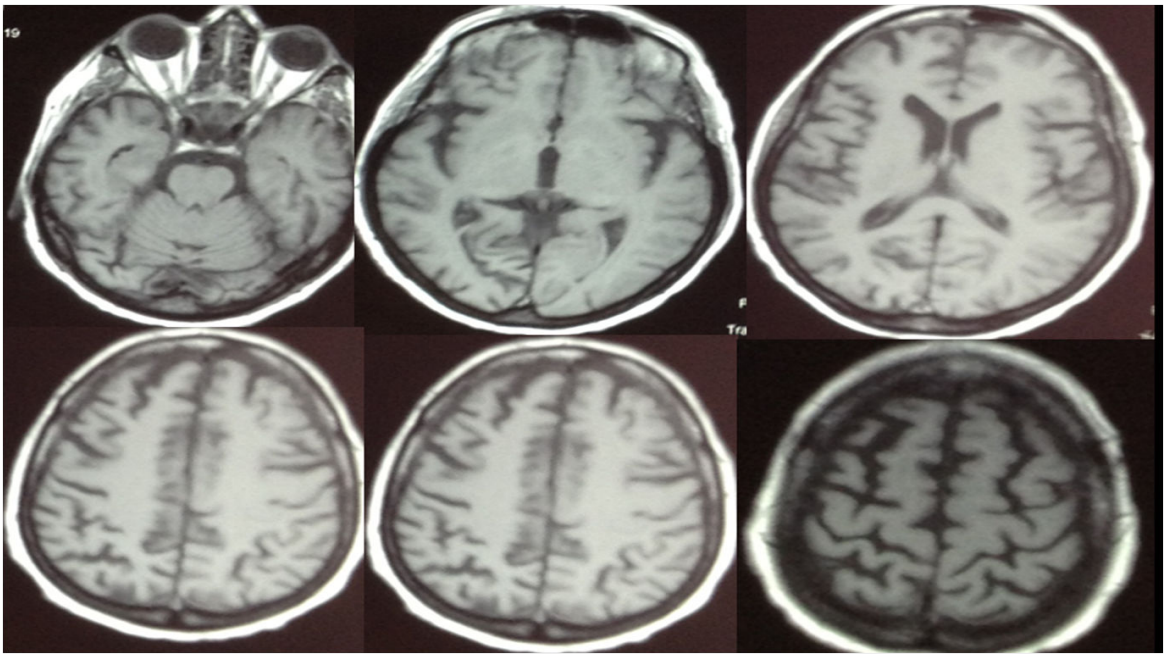

Figure 1: MRI scan performed at admission.

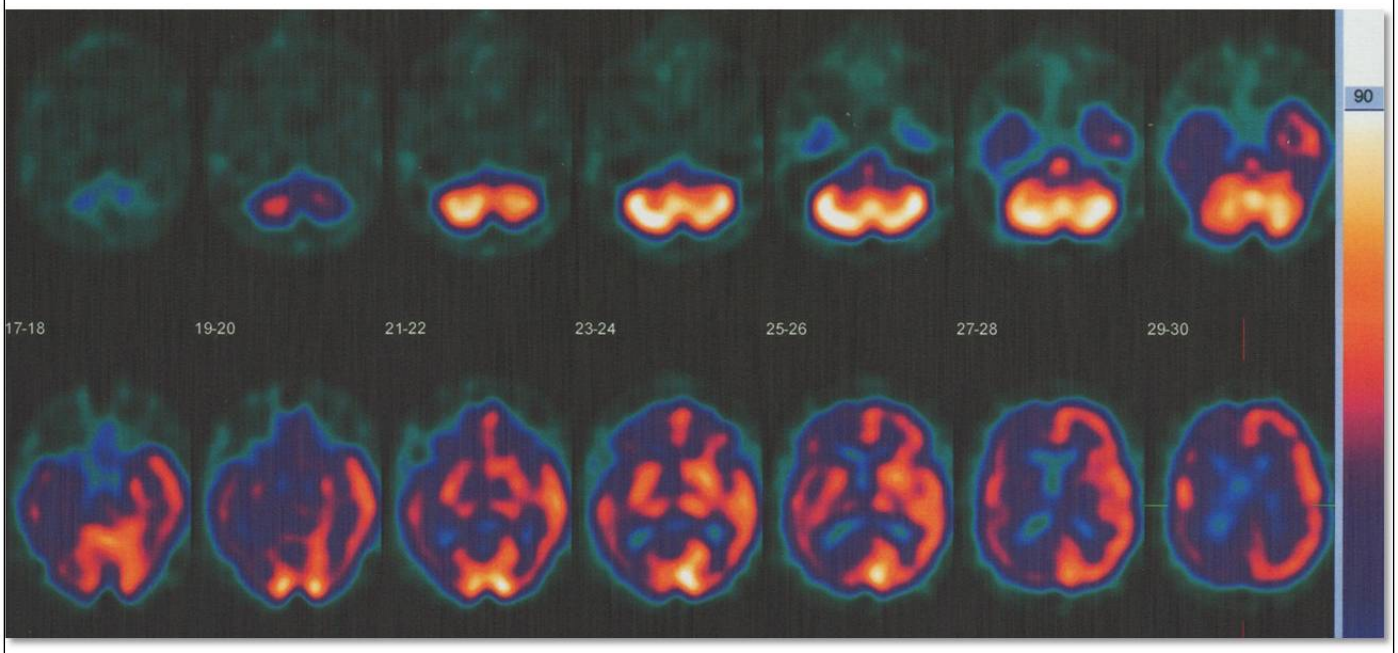

Figure 2: Single Photon Emission Computerized Tomography (99mTc SPECT) study at admission.

Figure 3A

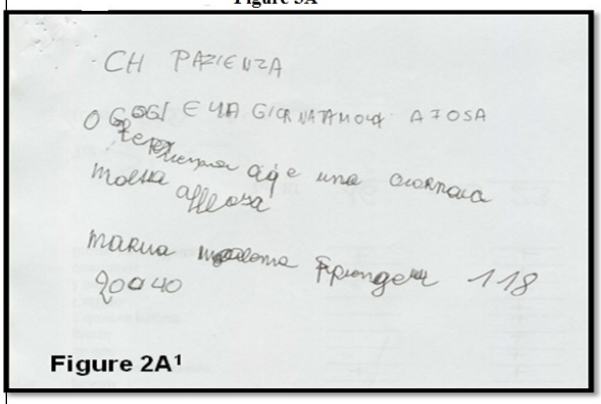

Figure 2A¹

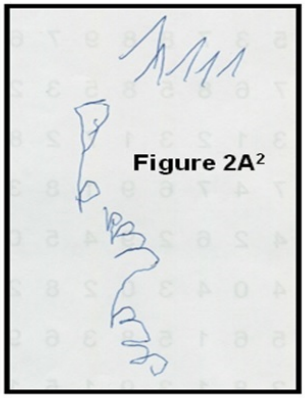

Figure 2A²

Figure 3B

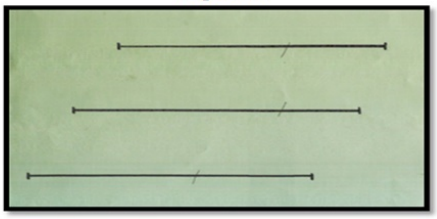

Figure 3C

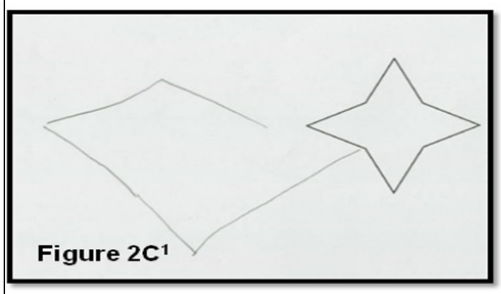

Figure 2C¹

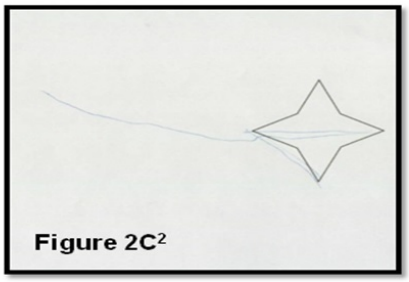

Figure 2C²

Figure 3: Neuropsychological study. **Figure 3A:** Writing test performed at admission (Figure 2A1) and one year later (Figure 2A2); **Figure 3B:** Visually neglect test performed at admission; **Figure 3C:** Shape Copy test performed at admission (Figure 2C1) and one year later (Figure 2C2).

Cognitive Domains and Test	**(Correct Score) Baseline evaluation**	**(Correct Score) Follow up evaluation (1yr)**	**Decrease rate**	**Qualitative assessment**
Spatial Orientation **Temporal Orientation**	5/5 5/5	1/5 1/5	80% 80%	Down Down
Language				
Denomination (AAT)	17/30	0/30	Compromised	Down
Phonological Fluency (Carlesimo et al.,) [16]	20 (27.9)	2(9.9)	64.51%	Down
Semantic Fluency (Novelli et al.,) [17]	31(39)	4(12)	69.23%	Down
Reading				Down
Writing				Down
Intelligence				
Raven's Coloured Progressive Matrices (Carlesimo et al.,) [16]	10/36 (13.9/36)	Not valuable		Down
Short Memory				
Short Spatial Memory_ Corsi's Test (Orsini et al.) [18]	5 (5.5)	0/5	Compromised	Down

Short Verbal Memory_ Digit Span Forward (Orsini et al.,) [18]	5 (5.5)	2 (2.5)	54.54%	Down
Working Memory				
Digit Span Backward (qualitative)	2	0	Compromised	Down
Verbal Long Memory				
Rey's 15 word-test (Carlesimo et al.,) [16] Immediate Recall: Delayed Recall:	33 (39.1) 9 (10.8)	7(13.1) 0	66.49% Compromised	Down Down
Story Recall (Novelli et al.,) [17]	19.5 (22)	0	Compromised	Down
Executive Functions				
Attentional Matrices (Spinnler and Tognoni) [19]	15(18/60)	Not valuable	Compromised	Down
Trail Making Test (Giovagnoli et al.,) [20] TMT-A TMT-B	123" Interrupted	Not valuable Not valuable	Compromised Compromised	Down Down
Apraxia, Perception and Visuo-Contructional Ability				
VOSP	Not valuable	Not valuable	Compromised	
Shape Copy (Carlesimo et al.,) [16]	0/12 (0/12)	0	Compromised	Not Valuable
Ideo-motor praxis (De renzi et al.,) [21]	59/72	45/72	23.72%	Down

Table 1: Cognitive assessment performed at admission and one year later. The numbers in parenthesis indicate the correct score according to age and education of the patient.

Discussion

There is a growing number of reports of atypical focal cortical presentations of AD in patients with relatively younger onset, suggesting that diagnosis of AD needs to be considered even in patients who present with focal dementia without significant memory loss. Morevover, it is well established that the most frequent focal cortical presentations af AD are PCA, corticobasal syndrome (CBS), behavioural variant frontotemporal dementia (bvFTD), progressive non-fluent aphasia (PNFA) (or a mixed aphasia) and semantic dementia (SD) [7].

PCA is a focal cortical presentation of AD, atypical for its early age at onset and because a single cognitive domain, not related to memory, is predominantly affected [3].

Establishing clinical diagnosis of PCA involves a two-step process; at first, patients should meet basic criteria based on McMonagle et al. [5], and then they can be furtherly classified into three main variants: biparietal syndrome (apraxia, visuospatial problems, agraphia, Balint's syndrome with preserved basic perceptual abilities, object recognition and reading), occipitotemporal syndrome (alexia, apperceptive agnosia and/or prosopagnosia) and visual variant (primary visual failure and impairment of basic perceptual abilities) [7].

We studied a 60-year patient with PCA and relatively short disease duration. Her clinical, cognitive and neuroradiologic findings indicate a predominantly right hemisphere dysfunction, resulting in dyspraxic difficulties, deficits in writing and impairment of visuo-spatial functions. Migliaccio et al. [8] showed that these deficits result from a selective posterior pattern of grey matter atrophy and from a damage to specific white matter pathways composing the visuo-spatial network.

CSF profile of the patient was typical for AD (characterized by alteration of ABeta42, t-tau and p-tau) [9]. The presence in our patient of the three pathological biomarkers of neurodegeneration confirm that in most in cases PCA can be considered a focal cortical variant of AD.

Previous literature studies support the hypothesis that a greater load of amyloid plaques and neurofibrillary tangles in the cerebral tissue are correlated with the earlier clinical symptoms of PCA patients and with the progressive cognitive decline [10,11], Since many patients with PCA show posterior neuritic plaques and neurofibrillary tangles at autopsy, the term PCA was often used interchangeably with AD or "the visual variant of AD." [12]. Further studies showed the uniformity of the clinical profile of PCA, which is distinct from that of typical amnestic AD [5] and demonstrated that numerous conditions can mimic symptoms of PCA, including other types of progressive dementias, such as cortical basal syndrome (CBS), dementia with Lewy bodies and prion disease, in which presentation with apraxia and prominent visuospatial deficit is also common [13].

On the basis of these reports, this syndrome should be not considered exclusively a variant of AD, but rather a distinct nosologic entity with its behavioral profile dictated by the topography of

pathology, more than its histology, and with its own diagnostic criteria.

Ruling out AD may also prevent inappropriate treatment, renouncing to use someone of the many medications available for AD, which are unnecessary for the conditions mimicking the symptoms of AD. Furthermore, whereas it is well known that a progressive amnestic syndrome in an elderly subject is highly predictive of AD pathology, in younger patients (under age 65) atypical, focal clinical syndromes which spare the long term memory may not evoke immediately the diagnosis of dementia disorder, and delay its correct treatment. For example, the visual variant of PCA can sometimes be misattributed to ocular causes, such as cataract or macular disease [11].

Our patient started therapy with transdermal rivastigmine (4,6 mg/24 h), and at one-year follow-up we observed a significant deterioration not only of the neuropsychological skills, but also in her daily living activities. Acetylcholinesterase inhibitors are usually reported as ineffective in the literature with, only a few cases suggesting some clinical benefits in PCA patients [14]. As far as we know, no clinical trials have been published.

In conclusion, our case report highlights the importance of making an accurate diagnosis of AD in clinical practice (and especially of ruling out AD) in order to reduce potentially inappropriate treatments which may slow the progression of the symptoms and disability of AD but are useless for other conditions [15].

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