JOURNAL OF APPLIED COGNITIVE NEUROSCIENCE
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Short Survey

Corticobasal syndrome as a focal variant of Alzheimer's disease

Síndrome corticobasal como variante de la enfermedad de Alzheimer

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How to cite this article:
Mariani, F., Damian, A., Pino, S., Dansilio, S. & Ferrando, R. (2022). Corticobasal syndrome as a focal variant of Alzheimer's disease. *Journal of Applied Cognitive Neuroscience*, 3(2), e00284513.
https://doi.org/10.17981/JACN.3.2.2022.06

Manuscript received on 14th September 2022
Accepted on 29th November 2022

Abstract

We present a 60-year-old woman with a 2-year history of progressive anterograde memory deficit, that evolved with a typical corticobasal syndrome with extrapyramidal signs and left superior limb dystonia. 18F-FDG and 11C-PIB PET/CT studies supported the diagnosis of Alzheimer's disease associating a corticobasal syndrome.

Palabras clave: Enfermedad de Alzheimer; síndrome corticobasal; depósitos amiloides; deterioro de la memoria; demencia

Resumen

Presentamos una paciente, mujer de 60 años, con un déficit progresivo de memoria anterógrada de 2 años de evolución que desarrolló un síndrome corticobasal típico, con signos extrapiramidales y distonía de miembro superior izquierdo. Los estudios PET/CT con 18F-FDG y 11C-PIB fueron compatibles con el diagnóstico de enfermedad de Alzheimer con síndrome corticobasal asociado.

Keywords: Alzheimer's disease; corticobasal syndrome; amyloid PET; memory impairment; dementia

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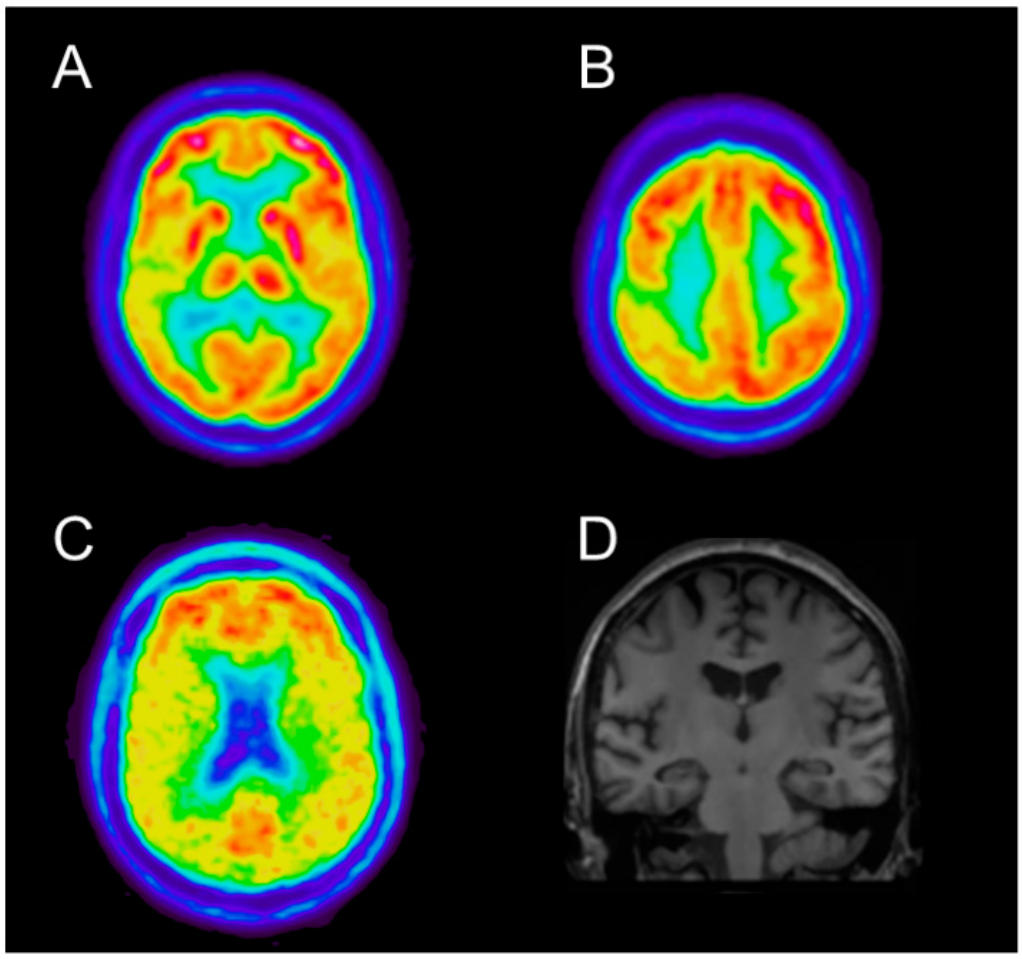
Introduction

We report a woman of a 60-year-old with a 2-year history of progressive anterograde memory deficit affecting episodic memory and loss of strength in the left upper limb without dystonic component. She had a maternal family history of late-onset dementia and Parkinson´s disease and a medical history of high blood pressure. She added depressive symptoms and received pharmacological treatment with bisoprolol and escitalopram. Neurological examination was unremarkable. A formal neuropsychological evaluation (see table 1) revealed a mesotemporal amnesic syndrome, elements of executive dysfunction, incipient ideomotor apraxia and mild visuo-constructive disintegration. Repercussion in instrumental activities of daily life with anosognosia was also present.

A PET/CT study with 18F-FDG was performed, which showed severely asymmetric right temporoparietal, perirolandic, striatal and thalamic hypometabolism (Figure 1). 11C-PIB PET/CT showed significant cortical amyloid deposits (C). MRI presented bilateral hippocampal atrophy corresponding to a medial temporal lobe atrophy (MTA) score of 1 (D), abnormal in the age range < 65 years.

Figure 1. *FDG-PET with asymmetric temporoparietal, perirolandic, striatal and thalamic hypometabolism (A and B). Positive amyloid PET (C). Bilateral hippocampal atrophy on MRI, with MTA score of 1 (D).*



Source: Authors.

In the follow-up (4 years later) she presented a severe cognitive impairment with a CDR score of 2. The amnesic syndrome remained stable, but significant worsening was evidenced in other cognitive domains (Table 1). The patient also presented a typical corticobasal syndrome with extrapyramidal signs: limb rigidity, limb myoclonus, left superior limb dystonia, limb apraxia, alien limb phenomena, decreased left-sided sway and paretic gait. She reported less use of the left hemibody, requiring assistance for basic activities of daily life. Increased depressive symptoms were evident, with anxiety and severe weight loss, that were treated with duloxetine and alprazolam. These clinical and imaging features were consistent with an atypical evolution of AD (Alladi et al., 2007).

Although AD has a characteristic clinical presentation (Calderon-Garcidueñas & Duyckaerts, 2018; Hodges et al., 2006; Traykov et al., 2007) some patients may present with atypical symptoms including focal signs, that confirm the heterogeneity of the disorder (Kramer & Miller, 2000). Within the presentation and evolution variants of AD, corticobasal syndrome with apraxia, sensory deficits and alien limb phenomenon has been previously described (Armstrong et al., 2013) (Constantinides et al., 2019). Other symptoms include asymmetric parkinsonism, dystonia, and different patterns of cognitive dysfunction (Ali & Josephs, 2018). Structural and functional imaging studies usually show asymmetric cortical and subcortical abnormalities, involving parietal cortex, perirolandic region and basal ganglia (Ali & Josephs, 2018; Di Stasio et al., 2019). This pattern was present in the 18F-FDG images of our patient years before the onset of the corticobasal syndrome. AD should be considered as a possible diagnosis in patients presenting an amnesic profile with motor signs and asymmetric deficits (Kramer & Miller, 2000). Amyloid PET may play an important role in supporting AD diagnosis in these patients.

TABLE 1. *Neuropsychological assessment.*

	2018	2022	$\bar{X}$
MMSE (Folstein et al).	27/30	10/30	-
CDR	Not performed	2	-
ADAS - COG	Not performed	43.3/70	12[1]
Attention and Executive Functions			
Digit reversal	3	3	5
Trail Making A	55"	57"	35"
Trail Making B	199"	> 240"	78"
Memory			
Digit span	5	4	7
Logical Memory (Inmediate/Delayed)	2/0,5	1.5/0	
Total RALVT	31	26	50
Interference	5	5	10
Ideomotor Praxis	3/6	1/6	-
Visuconstructive	Impaired[*]	Impaired	-
Language (Boston)	Normal	Disartria, anomias and random paraphasias	-

[1] Cutting score. [*]Topological transposition of details. Source: Authors.

Contributor Roles

Francesca Mariani: Conceptualization, formal analysis, investigation, writing original draft.
Andres Damián: Conceptualization, formal analysis, investigation, writing original draft, writing review/editing.
Pablo Duarte: Supervision, writing review.
Stephanie Pino: Investigation.
Sergio Dansilio: Supervision, writing review, editing.
Rodolfo Ferrando: Supervision, writing review, editing.

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