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Short Survey

Corticobasal syndrome as a focal variant of Alzheimer's disease

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Abstract

We present A 60-year-old woman with a 2-year history of progressive anterograde memory deficit, with a A 18F-FDG PET/CT and 11C-PIB PET/CT studies performed. In the follow-up she presented a typical corticobasal syndrome with extrapyramidal signs and left superior limb dystonia. These clinical and imaging features are consistent with a focal presentation of AD.

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

Resumen

presentamos el caso de una mujer de 60 años, con un déficit progresivo de memoria anterógrada de 2 años de evolución. Se realizó estudios 18F-FDG PET/C y 11C-PIB PET/CT compatibles con EA. En la evolución presentó un síndrome cortico basal típico, con signos extrapiramidales y distonia de miembro superior izquierdo. Tanto los hallazgos de imagen, como la presentación clínica son consistentes con una presentación focal de EA.

Keywords: Alzheimer's Disease; corticobasal syndrome; Amyloid deposits; memory impairment; dementia

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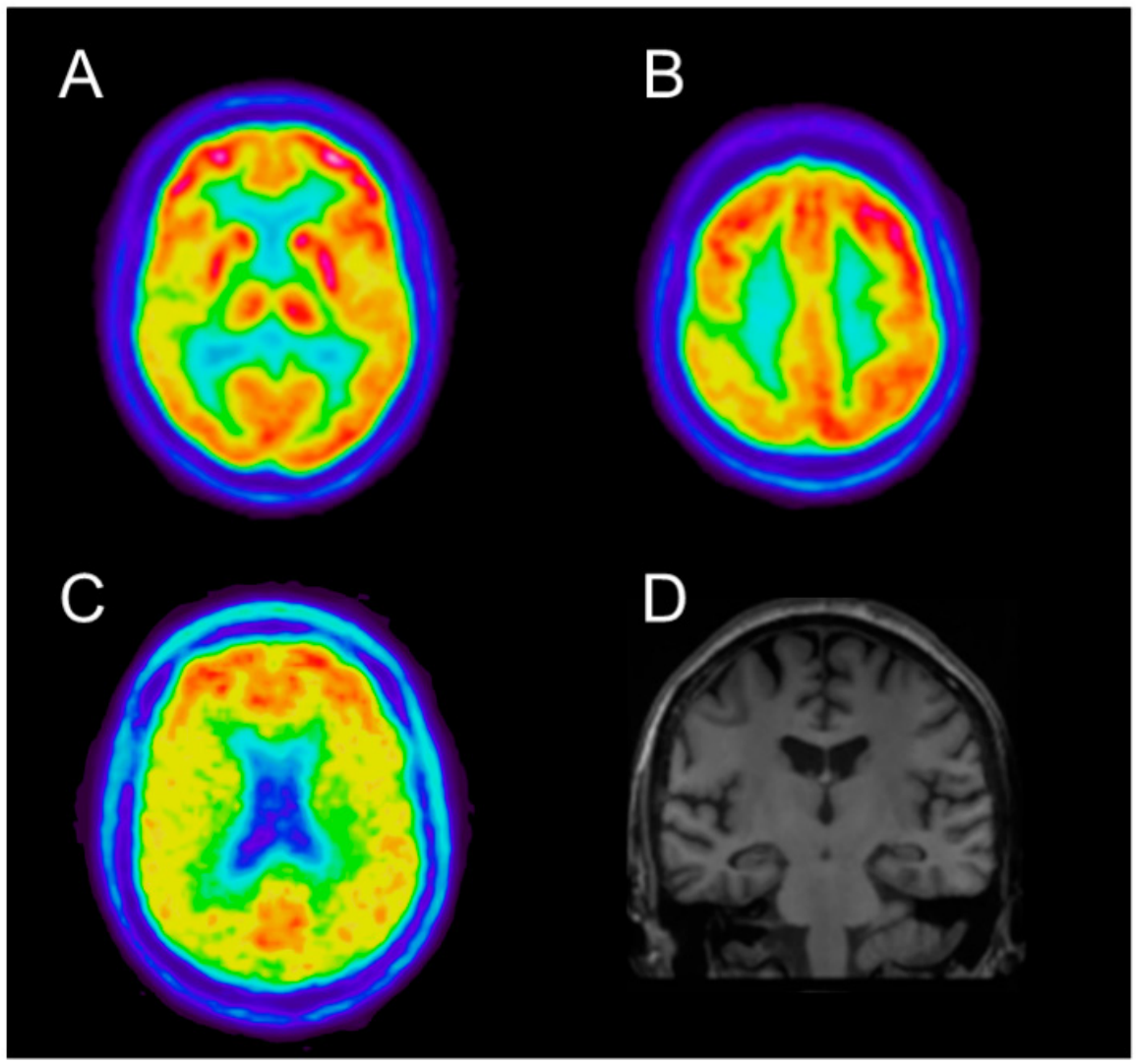
We report the case of a 60-year-old woman with a 2-year history of progressive anterograde memory deficit that affects episodic memory and loss of strength in the left upper limb. Neurological examination was unremarkable. A formal neuropsychological evaluation revealed mesotemporal amnesic syndrome, executive dysfunction, visuoconstructive disintegration and semantic lexical deficit. Repercussion in instrumental activities of daily life with anosognosia was also present.

A 18F-FDG PET/CT study was performed, which showed severe asymmetric right temporoparietal and right thalamic hypometabolism (Fig. 1A and Fig. 1B). 11C-PIB PET/CT showed significant cortical amyloid deposits (C). MRI showed bilateral hippocampal atrophy (D). In the follow-up she presented a typical corticobasal syndrome with extrapyramidal signs and left superior limb dystonia. These clinical and imaging features are consistent with a focal presentation of AD (Alladi et al., 2007).

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



Source: Authors.

Although AD has a characteristic clinical presentation (Calderon-Garcidueñas & Duyckaerts, 2018; Hodges et al., 2006; Traykov et al., 2007) macroscopic atrophy affects the entorhinal area and hippocampus, amygdala, and associative regions of the neocortex. The locus coeruleus is depigmented. The deposition of Aß is first made of diffuse deposits. Amyloid focal deposits constitute the core of the senile plaque which also comprises a corona of tau-positive neurites. Aß deposits are found successively in the neocortex, the hippocampus, the striatum, the mesencephalon, and finally the cerebellum together with the pontine nuclei (Thal phases some patients may present with atypical symptoms including focal signs, which confirm the heterogeneity of the disorder (Kramer & Miller, 2000). Within the focal variants of AD, the corticobasal syndrome with apraxia, sensory deficits and alien limb phenomenon has been previously described (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 in these patients, involving parietal and perirolandic regions as well as basal ganglia (Ali & Josephs, 2018; Di Stasio et al., 2019). AD should be considered as a possible diagnosis in patients presenting with focal signs and asymmetric motor deficits (Kramer & Miller, 2000).

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