Posterior Cortical Atrophy

An Atypical Variant of Alzheimer Disease

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KEYWORDS

• Neuropsychiatry manifestations (NPM) • PCA • Young-onset dementia • Apathy • Depression • Posterior atrophy • Visual impairment • Peer support

KEY POINTS

- Patients with posterior cortical atrophy (PCA) present an atypical phenotype different from typical (amnesic) Alzheimer disease (AD) in that it is characterized by midlife onset, progressive visual dysfunction, and focal posterior (occipital and parietal) atrophy; hence, neuropsychiatric phenomena also differ from those of typical AD.
- The neuropsychiatric profile in PCA is often overlooked and merits attention because of its impact on the patients' quality of life and prognostic implications.
- Apathy, anxiety, depression, and irritability are the most common neuropsychiatric manifestations (NPM) in PCA.
- Neuropsychiatric examination is an essential tool for discriminating PCA caused by AD and dementia with Lewy bodies.
- Focused interviewing targeting NPM must be included in the clinical interview, and validated neuropsychiatric scales should be added to neuropsychological assessments.
- Individualized therapy including cognitive-behavioral therapy is valuable in PCA.
- Support groups are powerful intervention tools for patients and families.

INTRODUCTION

Posterior cortical atrophy (PCA) is a clinical syndrome characterized by progressive loss of visual processing and other posterior brain functions (including reading,

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Abbreviations

AD	Alzheimer disease
CBT	Cognitive-behavioral therapy
DLB	Dementia with Lewy bodies
LOAD	Late-onset Alzheimer disease
NPM	Neuropsychiatry manifestations
PCA	Posterior cortical atrophy
PCA-AD	Posterior cortical atrophy–Alzheimer disease
PCA-DLB	Posterior cortical atrophy–dementia with Lewy bodies
VH	Visual hallucinations
YOAD	Young-onset Alzheimer disease

calculation, and navigational orientation) and atrophy of the parietal, occipital, and occipitotemporal cortices.[1] Alzheimer disease (AD) is the most common underlying pathologic state (up to 78% of patients with PCA having pathological confirmed AD[1]) with alternative causes including dementia with Lewy bodies (DLB), subcortical gliosis, corticobasal degeneration, and prion-associated disease.[1–4] There are no epidemiologic studies of PCA, but it has been estimated that PCA may account for 5% to 10% of young-onset AD (YOAD) presentations.[5] Age at onset is usually lower in PCA than in typical (amnesic) AD, with most patients with PCA experiencing their first symptoms in their 50s or early 60s.[6,7]

Patients with PCA report difficulties in reading, driving, navigating, and identifying objects.[1,6–8] In many senses these patients behave as if blind, regardless of their preserved visual acuity and absence of ophthalmologic impairment. Very often they are referred by ophthalmologists, as visual difficulties are commonly their first and main complaint. Deterioration in other cognitive domains comes over time, degrading posterior functions, such praxis, calculation, and spelling first, whereas episodic memory, insight, and anterior functions (such as attention and executive functions) are relatively preserved until later in the disease. Although research on the neurologic, cognitive, and neuroimaging characteristics of PCA have increased during the last 2 decades, the neuropsychiatric manifestations (NPM) have received little attention and are consequently poorly characterized. More than 80% of patients with typical AD have some kind of neuropsychiatric disorder over the course of the disease[9]; these rates are even higher in DLB.[10] In short, NPM have proved to be highly prevalent in patients with dementia, are a domain of great complexity, and have important implications for diagnosis, treatment, and prognosis.[10–14] Studying NPM in atypical phenotypes of AD is particularly challenging because the prevalence of these forms is low and missed diagnosis common. Furthermore, in the case of syndromes in which specific clinical features are particularly salient and striking (such as visual disturbances in PCA), other regular features (eg, depression or delusions) may be overlooked.

NEUROPSYCHIATRIC MANIFESTATIONS AND CLINICAL PICTURE

In this review, the authors examine the evidence concerning similarities and differences in the patterns of NPM expressed by individuals with PCA and typical AD. One pertinent factor is the younger age at onset of PCA compared with typical AD. The data regarding the prevalence of NPM in YOAD (cases with onset before 65 years of age) and late-onset AD (LOAD) are equivocal, as some studies report a higher prevalence of NPM in YOAD and others in LOAD.[12,15–17] The problem of interpretation is that these studies generally have small samples, and the YOAD samples are likely to include other atypical AD phenotypes (such as frontal-variant AD, which mimics