SHORT REPORT

Alzheimer’s disease in a patient with posterior cortical atrophy

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Abstract
Posterior cortical atrophy (PCA) is characterised by slowly progressive dementia with cognitive and perceptual deficits suggestive of bilateral parieto-occipital disease. A case is reported of a patient with PCA and neuropathological findings consistent with Alzheimer’s disease.

Benson et al.1 recently described posterior cortical atrophy (PCA) in five patients who showed slowly progressing dementia with initial alexia and visual agnosia, followed by Balint’s syndrome, Gerstmann’s syndrome, constructional apraxia, spatial disorientation, and transcortical sensory aphasia. All patients had bilateral parieto-occipital atrophy. On neuropathological examination one patient showed bilateral parietal subcortical gliosis and another showed bilateral parietal congophilic angiopathy, neuritic plaques (NPs) and neurofibrillary tangles (NFTs).2

Our patient with neuropathologically verified Alzheimer’s disease (AD) presented with cognitive deficits restricted to functions served by the posterior parietal and occipital lobes.

Case report
The patient was a 48 year old, right handed woman with 12 years of formal education, who had a three year history of slowly progressive visual loss and writing difficulties. Her father had had dementia and a sister had multiple sclerosis. One year before the examination she gradually became unable to read and write, developed problems in manual skills and calculation, and showed episodes of spatial disorientation. Her main visual complaint was piecemeal perception and transient disappearance of objects. Although she could identify objects by sight, her relatives reported that she behaved like a blind person as she could not navigate her room without colliding with objects. These disturbances led to early retirement. Her relatives also noticed that she developed an anxiety disorder in the absence of prominent language or memory deficits. On neurological examination she was alert and oriented. Though ocular fundus, visual fields, oculocephalic movements, and optokinetic responses were all normal, saccadic eye movements to the left were slow. Visual acuity was difficult to assess due to abnormal fixation, and her corrected visual acuity for both eyes was 6/10, 8/10, and 4/10 in three successive evaluations. Results of the rest of the neurological examination were unremarkable.

On neuropsychological examination her Blessed dementia scale score was 5.5 points, and the Mini-Mental State Examination score was 22 points. On the WAIS,3 there was a remarkable dissociation between verbal IQ (87 points) and the performance IQ (0 points) (the patient could not perform any of the subtests). On the Weschler memory scale,4 her memory quotient was below average (80 points); she showed severe impairment of visual memory for geometric designs, but her immediate verbal recall and overall verbal memory were only marginally defective. Language was fluent and grammatically correct but contained occasional semantic paraphasic substitutions. Auditory comprehension and sentence repetition were normal. She had echolalia, however, impaired object naming, and defective word list generation. On the Western aphasia battery,5 she scored 84-4 points, and the profile was consistent with a mild anomic aphasia. She also showed letter-by-letter reading, impaired

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Figure. SMI 34 MAb staining showing moderate immunostaining of neuritic plaques and neurofibrillary tangles (magnification ×400).
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Neuropathology

After informed consent and to help in the diagnosis of the underlying neurological disorder a brain biopsy specimen was obtained from the right parietal cortex (Brodmann's area 7). Specimens were stained with haematoxylin-eosin, PAS, MacMunn, Congo red, and Bielchowski silver impregnation. Immunocytochemistry was performed with a monoclonal antibody against paired helical filament (SMI 34 MAb). There was mild neuronal loss and a mild lipofuscin accumulation. Congo red and Bielchowski silver impregnations showed abundant neuritic plaques (NPs) (mean plaque count 8-6 per 250 x field) and occasional neurofibrillary tangles (NFTs). There were also some immature NPs but no granulovacular degeneration. SMI 34 MAb immunostained both NPs and NFTs (figure). There were no Pick bodies, Lewy bodies, subcortical gliosis, or status spongiosus.

Discussion

The syndrome of PCA does not necessarily imply Alzheimer's pathology as it may also be seen in cases of Pick's disease, progressive subcortical gliosis, and Creutzfeld-Jakob disease (Heidenhain's type).2 Cognitive and perceptual deficits of similar characteristics to PCA, however, have been most often reported in the early and middle stages of AD, possibly reflecting an initial, selective patho-

8 Hooper HE. The Hooper visual organisation test. Los Angeles: Western Psychological Services, 1958.

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