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, Gertsman'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 evaluation her Blessed dementia scale³ score was 5.5 points, and the Mini-Mental State Examination⁴ score was 22 points. On the WAIS,⁵ 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,⁶ 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,⁷ 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 $\times 400$).

reading comprehension, right-left discrimination, and finger localisation as well as acalculia and agraphia. She showed all the features of Balint's syndrome, including simultanagnosia, ocular motor apraxia, and impaired visually-guided hand movements. Colour recognition and discrimination of unfamiliar faces were preserved, but she had impaired visual recognition of objects (20% correct) and familiar faces (30% correct). She also did poorly (more than 2 SDs below the standardised scores) in other complex visual perceptual tests (Hooper visual organisation test, visual form discrimination, and judgement of line orientation), and on constructional tasks (three dimensional block construction).^{8,9} She also showed ideational apraxia, dressing apraxia, spatial disorientation, and loss of mental imagery for colours, objects, animals, and spatial associations. An electroencephalogram showed bilateral slow wave activity over posterior brain regions. Magnetic resonance imaging (MRI) revealed cortical atrophy restricted to the posterior parieto-occipital areas, with no evidence of atrophic changes in fronto-temporal regions (figure).

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, McManus, Congo red, and Bielschowski 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 Bielschowski silver impregnations showed abundant neuritic plaques (NPs) (mean plaque count 8.6 per 250 × field) and occasional neurofibrillary tangles (NFTs). There were also some immature NPs but no granuvacuolar 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 Creutzfeldt-Jakob disease (Heidenhain's type).² Cognitive and perceptual deficits of similar characteristic to PCA, however, have been most often reported in the early and middle stages of AD,¹⁰⁻¹³ probably reflecting an initial, selective patho-

logical involvement of the occipital and posterior parietal association areas. A recent neuropathological study of a subpopulation of AD patients presenting with a Balint's syndrome revealed prominent histopathological changes in parieto-occipital regions with little involvement of other neocortical areas (such as the prefrontal cortex) that are usually affected in typical cases of AD.¹⁴

Hof and co-workers recently suggested that Balint's syndrome in patients with AD results from a specific regional and laminar distribution of NPs and NFTs in the occipito-parietal association cortex with preferential degeneration of the dorsal occipito-parietal visual pathway.¹⁴ These structures were presumably affected in our patient. The coexistence of visual object agnosia, prosopagnosia, and letter-by-letter reading, however, reflects additional degeneration of the ventral occipital-temporal visual pathway. Finally, the occurrence of spatial disorientation, dressing apraxia, ideational apraxia, anomia, and Gertsmann's syndrome suggests involvement of the adjoining inferior parietal lobes.

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