

Looking beyond the eyes: visual impairment in posterior cortical atrophy

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A 66-year-old, left-handed man was seen in our specialist clinic, complaining of difficulty reading, counting coins, using his mobile phone, and getting dressed. Otherwise, he was managing at home alone and going out independently, albeit with difficulty using stairs and walking on uneven ground. His problems had started 6 years earlier with difficulty reading and fastening his buttons and zips. At that stage, having seen opticians, he was referred to an ophthalmologist and underwent cataract surgery; however, his visual problems continued and got worse. He had no medical or family history of neurological disease.

On physical examination, we noticed he had difficulty locating objects in space in the left lower hemifield. Visual acuity, fundi, pupillary responses, and extraocular movements were normal. We also noticed bilateral finger myoclonus and symmetrically brisk reflexes with flexor plantars. Tone, power, and sensation of all limbs and his trunk were normal. On cognitive testing, he scored 25/30 on the Mini-Mental State Examination, losing points for recall of three words, writing, reading, and drawing intersecting pentagons. He also struggled to perform simple calculations and had limb dyspraxia even after we accounted for his visual problems.

In view of these findings, we suspected an underlying neurodegenerative disorder. Blood investigations for reversible causes of dementia were either negative or normal. An MRI of his brain showed extensive atrophy of the parietal and occipital lobes with so-called knife-like thinning of the gyri (figure). We made a diagnosis of posterior cortical atrophy (PCA), on the basis of the clinical presentation and the findings on imaging. The patient was offered

cerebrospinal fluid (CSF) analysis and amyloid PET—to investigate further the underlying cause of the PCA—but he decided he did not want any additional tests.

PCA is a syndrome characterised by progressive impairment of visual processing and other functions of the occipital and parietal lobes including praxis, calculation, and spelling. Typically, patients present to health professionals with PCA symptoms before the age of 65 years. PCA is usually caused by Alzheimer’s disease, and is not thought to occur with an autosomal dominant pattern of inheritance. Other causes of PCA include dementia with Lewy bodies, corticobasal degeneration, and, very rarely, prion disease. CSF analysis—to look for amyloid and tau proteins—or amyloid PET imaging can provide evidence to corroborate the diagnosis of Alzheimer’s disease.

Patients with PCA are often referred to optometrists and ophthalmologists in the belief that they have a primary ocular abnormality, leading to a delay in diagnosis of the syndrome. It is therefore important that PCA is considered in late middle-aged patients presenting with progressive visual symptoms and normal visual acuity. Our case illustrates the need for awareness that visual problems in patients might not have a cause within the eye.

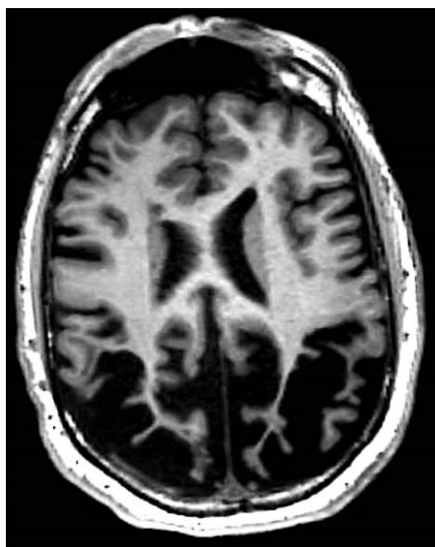


Figure. Axial T1-weighted MRI scan shows marked atrophy of the occipital and parietal lobes