Alzheimer’s dementia, posterior cortical atrophy variant with BPSD

Samuel Amo-Korankye MBBS, MRCPsych, Saheem Gul MBBS, MRCPsych, Sheeba Ninan MBBS, MRCPsych, Mina Patel-Palfreman BSc, CPsychol

The behavioural and psychological symptoms of dementia (BPSD) can have a devastating impact on carers and family. Here, the authors describe a challenging case of a man with Alzheimer’s dementia (AD), posterior cortical atrophy variant (PCA) who presented with severe BPSD, but discuss how non-pharmacological methods were instrumental in helping his management.

Posterior cortical atrophy (PCA) is seen as an atypical and rare variant of Alzheimer’s dementia (AD). It is characterised by a progressive decline in visuospatial, visuoperceptual, literacy and praxic skills with early sparing of language and memory.1

In PCA the main lobes of the brain affected are the parietal, occipital, and temporal. The age of onset of PCA is usually younger and typically falls between 50–65 years.1–3

Although the majority of patients with PCA have the underlying aetiology as AD, other aetiologies include Lewy Body dementia, corticobasal degeneration, and prion disease (see Box 1).1

Behavioural and psychological symptoms of dementia (BPSD) include agitation, depression, apathy, repetitive questioning, psychosis, aggression, sleep problems and wandering.3 According to the available evidence up to 90% of sufferers with dementia experience or present with symptoms suggestive of BPSD during their illness.3–5

Presentation
We present a 73-year-old man with a diagnosis of AD, PCA variant. He was originally diagnosed with AD in 2011 after a 24-month history of short term memory loss and declining topographical memory. His medical history included ischaemic heart disease, myocardial infarction, coronary artery bypass graft (CABG) in 2006, hypertension and benign hypertrophy of the prostate. There was history of previous head trauma. He had a positive family history for dementia in his mother.

An MRI head scan in 2011 showed generalised atrophy with relatively spared temporal lobes. The hippocampus was described as not being particularly atrophic. His cognitive test score was 22/30, losing 4 points on orientation, 3 points on recall and he was unable to draw intersecting pentagons. There was initial diagnostic uncertainty as to whether he had a neurodegenerative condition such as AD or whether the CABG and the use of general anaesthetics had an adverse effect on his cognition. Another differential diagnosis was vascular dementia (VD) due to his extensive vascular risk factors. An amyloid scan was considered but this was unavailable at the time. Cerebrospinal fluid (CSF) analysis was carried out as an alternative and the sample analysis showed an abnormal ratio of amyloid and tau proteins indicating the likelihood of AD. Routine blood tests were normal and so was a test for syphilis.

For initial management, he was treated with anticholinesterase inhibitors. These resulted in unwanted effects such as gastrointestinal problems, an increase in aggressive behaviours and confusion.

He was referred to his local memory service in 2012 due to changes in his behaviour and memory problems. There was a disturbing change in his behaviour in the form of extreme physical aggression. He had punched and kicked his spouse to the floor, throwing her against a wall and trying to smother her. He had also presented with visual acuity problems and problems with depth perception. Anticholinesterase inhibitors did not help with his cognition or BPSD. Diazepam was used on an as required basis in times of crisis. Other treatments included quetiapine and carbamazepine. They all had very little effect on the patient’s BPSD.

As part of his management he was referred to a day centre and allocated domestic carers to help support his family. He was suspended...
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from the day centre due to his aggression and violence towards his carers and other clients.

The patient required inpatient admission in October 2014 after a deterioration in his presentation and increased physical aggression. His cognitive test score on admission was 11/30. A trial of risperidone was commenced whilst an inpatient. This was stopped due to extrapyramidal side-effects (EPSEs). Whilst an inpatient, he was referred to psychological services in December 2014 to help with his general inpatient management. This included a life story work to help understand his background and to educate the family about dementia and BPSD. During this admission he had visuospatial difficulties leading to several falls. At times he appeared not to see properly to his right side. An orthoptist and neuro-ophthalmologist review concluded he had alternating extropia (eye turns out), diplopia, problems with depth perception and he could not complete a drawing of prisms. His intraocular pressure was within normal limits in both eyes, with healthy discs. His visual acuity was poor with a score of 6/48. A full assessment was not possible due to his poor ability to communicate and comply with instructions. A recommendation was made to refer him for a neurology review as his visual problems were thought to be due to a disorder of higher visual function. Due to his progressive visual symptoms and the relative early onset of his cognitive decline his diagnosis was reviewed and the diagnosis of AD with PCA was made. Unfortunately he was unable to tolerate attempts to repeat his head MRI.

He was discharged from hospital to a care home in January 2015 without any regular psychotropic medication.

On being transferred to a care home BPSD continued to be a management problem and had an adverse impact on his care. The relationship between care home staff and the family suffered hindering effective communication and care delivery.

He was referred to psychology once more to help the home manage his BPSD and to prevent his placement at the care home being terminated.

Psychology involvement initially included attendance at meetings to ascertain the key issues, challenges, goals and expectations of his family and carers.

Table 1. Frequency of BPSD evaluated with the neuropsychiatry inventory (NPI) in three European studies. MAASBED: Maastricht Study of Behaviour in Dementia, REAL: Réseaux Alzheimer Français, EADC: European Alzheimer Disease Consortium\(^1\)\(^3\)

<table>
<thead>
<tr>
<th>BPSD</th>
<th>Frequency (N=836%)</th>
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<tbody>
<tr>
<td>Apathy</td>
<td>55.5</td>
</tr>
<tr>
<td>Depression</td>
<td>44.9</td>
</tr>
<tr>
<td>Anxiety</td>
<td>42.0</td>
</tr>
<tr>
<td>Agitation</td>
<td>35.0</td>
</tr>
<tr>
<td>Irritability</td>
<td>30.6</td>
</tr>
<tr>
<td>Aberrant motor behaviour</td>
<td>24.7</td>
</tr>
<tr>
<td>Delusion</td>
<td>22.0</td>
</tr>
<tr>
<td>Appetite</td>
<td>21.4</td>
</tr>
<tr>
<td>Sleep</td>
<td>14.3</td>
</tr>
<tr>
<td>Disinhibition</td>
<td>12.4</td>
</tr>
<tr>
<td>Hallucination</td>
<td>8.5</td>
</tr>
<tr>
<td>Euphoria</td>
<td>6.8</td>
</tr>
</tbody>
</table>

Psychology involvement initially included attendance at meetings to ascertain the key issues, challenges, goals and expectations of his family and carers.

Target areas identified included the use of training, strategies to manage BPSD as well as improve communication and the relationship between his formal and informal caregivers.\(^5\) In the initial stages there was an emphasis on developing a relationship with all parties involved in his care and to ensure effective collaboration. Psychology intervention was phased and included brief behavioural analysis such as direct observation and the use of data collection charts to record behaviours. Information gauged from the initial analysis was used to inform the next phase of the psychological intervention. A two-part training session was subsequently designed and delivered to the caregiving team. The first part of the training was on understanding dementia, BPSD, PCA variant of dementia, using person-centred behaviour that challenges and person-centred care. A management plan was devised in collaboration with staff and family members,\(^7\) this included the development of a set of strategies and resources. These included adjustments and adaptations to the environment, as well as changes to furnishings and lighting.\(^8\)

A series of sessions were planned to identify barriers and strategies to improve communication between the family and the care home staff. The outcome of the first session was positive and opened up opportunities for improved relations and collaborative working. Staff and family explored strategies together with varying degrees of success. The family were encouraged to devise an activity plan. The use of behaviour modelling was encouraged and support was put in place to ensure consistent implementation. The second training session was used for supervision and reflection. Review of progress and ongoing challenges led to further modifications to the management plan.

Lack of consistency in practice and level of staff engagement and motivation were identified as important factors determining the success or failure of interventions. The staff training and communication sessions have been identified as the most helpful of all the psychology interventions. The introduction of one-to-one care has resulted in improved management.
of the patient’s aggression and BPSD. There were improved relations between the family and staff. Overall, despite his BPSD not resolving completely, staff members felt better able to manage the BPSD with the support of the family.

A request for extra support in the form of extra carers was also placed with the local health funding authority. It was felt that this would be necessary to ensure that the patient’s needs could continue to be met in this care home. The local health funding authority agreed for additional staffing to care for him.

**Conclusion**

In 2014, a study looked at whether there was any difference in BPSD in those with PCA and other forms of dementia such as AD. Their analysis of 20 patients with PCA and another 20 patients with AD found BPSD prevalence rates in the PCA group to be up to 95% compared with 85% of AD with the most occurring symptoms being anxiety and apathy (Figure 1). The occurrence of BPSD in most dementias irrespective of type is usually given as about 90%. Some have investigated whether there are different symptoms of BPSD associated with different types of dementia. One study in 2012 mostly compared BPSD symptoms from AD and vascular dementia (VD). The article found some differences in the neuropsychiatric syndromes in the different types of dementias.

Further studies compared the BPSD symptoms of PCA and AD found the commonest symptoms to be depression, irritability, apathy and anxiety for both groups with little difference between the two populations. A study in 2014 confirms this although a suggestion is made that the PCA group from their analysis showed higher rates of anxious patients and apathy.

BPSD ranges from mild problems such as fidgeting, and wandering to the extremes of verbal and physical aggression. BPSD poses major difficulties in the day-to-day care of patients and is likely to impair the quality of life of both patient and caregiver. It is associated with poor quality of life and increased morbidity and mortality.

The treatment of BPSD is complex and a good management plan should reflect and include all possible causative factors. Current guidelines suggest the use of non-pharmacological approaches first (see Box 2). Non-pharmacological approaches such as tailored psychosocial interventions that involve family and carers have been reported to help with BPSD.

Antipsychotics are often used in the management of BPSD. However, there are real concerns about adverse health risks such as extrapyramidal side effects, falls, cerebrovascular events, further cognitive decline and increased mortality. Studies have also shown that the impact on BPSD is limited or shows little positive impact.

This case highlights the importance of tailor-made approaches in the management of patients with...
dementia suffering with BPSD. The use of psychological formulation and the importance of psycho-education of family and staff in the non-pharmacological management of BPSD. The case also highlights the importance of implementing management strategies with a multidisciplinary approach to promote effective and sustainable outcomes. From the published literature the non-pharmacological approaches that are the most effective include educating and supporting carers, stress management and how to identify and manage difficult behaviour. Increasing the physical activity of dementia sufferers, improving communication, simplifying the immediate surroundings, music and simplifying tasks have been shown to help.

The use of non-pharmacological measures has been instrumental in helping the patient’s carers and family cope with our patient’s dementia and BPSD. Although non-pharmacological measures have not completely treated his challenging behaviours, they have helped prolong his residing at the care home where he had been at risk of needing to leave. The case also highlights PCA, a rare variant of AD, which can present with diagnostic difficulties. This is mostly due to the lack of awareness of the condition and also due to the multiple causes of the syndrome. The patient in this case study had PCA diagnosed relatively late. It is important to correctly diagnose such cases as it has the potential to help improve patient outcomes. We hope this case helps highlight the need for a higher index of suspicion in patients who present with cognitive decline as well as visuospatial problems. We hope this case also adds to the body of evidence already available on atypical variants of dementia such as PCA.

Dr Amo-Korankye, Dr Gul, Dr Ninan, are all Consultant Psychiatrists, Dr Patel -Palfreman is a specialist care pathway lead for acute and inpatient psychology. Dr Amo-Korankye, Dr Gul and Dr Patel-Palfreman are all at North East London Foundation Trust, UK and Dr Ninan works with Lancashire Care NHS Foundation Trust.

Declaration of interests
No conflicts of interest were declared.

References