The Assessment and Management of Psychiatric Symptoms in Prion Disease: An Illustrative Case Series

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Introduction

Prion diseases are rare neurodegenerative conditions. There are three aetiological groups: sporadic, inherited and acquired. The disease typically progresses over several months to death. However, some forms of the disease have a much longer duration. All forms are associated with complex and wide-ranging neuropsychiatric symptoms.

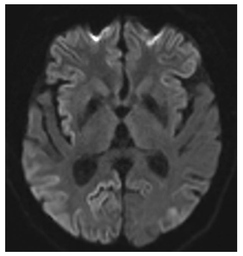

Figure 1. *Axial diffusion-weighted MRI showing typical changes of sCJD with high signal in the cortical ribbon and striatum*

The most common form of prion disease, Creutzfeldt-Jakob disease, has excellent diagnostic tests including MRI brain scan and CSF seed amplification assay.

For neuropsychiatric symptoms, psychological and pharmacological treatments are offered on a case-by-case basis depending on clinical presentation and treatment needs of the individual. Management can also be informed by guidelines for more common dementias.

Aim

In this clinical overview and case series, we aim to demonstrate the varied onset, nature and course of psychiatric symptoms in prion disease, as well as the diverse treatment approaches required.

Method

Since 2008, 1158 patients have been followed up over the full clinical course by prospective cohort study.

We provide an overview of psychotropic medication use in prion disease.

We present a series of profiles that typify the common presenting symptoms and management approaches.

Results

- **6%** of cohort patients with sporadic CJD (sCJD) presented with predominantly psychiatric/behavioural symptoms at the time of enrolment into cohort.
- **26%** of cohort patients were prescribed either antidepressant or antipsychotic medication.
- **13%** of cohort patients were prescribed benzodiazepine medication with a behavioural/psychiatric indication.

Antidepressant use

- The most commonly used antidepressants were citalopram, sertraline and mirtazapine.

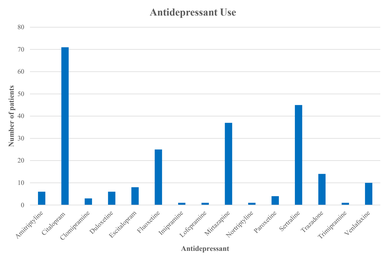

Figure 2. *Antidepressant use in the National Prion Monitoring Cohort*

Antipsychotic use

- The most commonly used antipsychotics were quetiapine, risperidone, olanzapine and haloperidol.

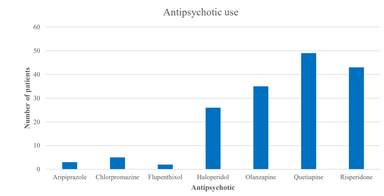

Figure 3. *Antipsychotic use in the National Prion Monitoring Cohort*

Results

Benzodiazepine use for psychiatric/behavioural symptoms. NB: Clonazepam is commonly used to treat myoclonus. This indication was excluded.

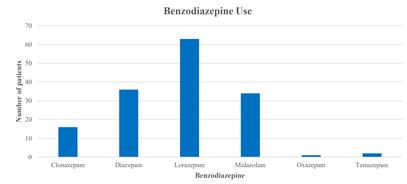

Figure 4. *Benzodiazepine use in the National Prion Monitoring Cohort*

Illustrative Profiles

Profile 1

Diagnosis: sCJD
Demographics: 60 year old male
Psychiatric/behavioural symptoms: Early visual hallucinations, including of animals. During disease progression increasing agitation including episodes of violence towards staff members at care home.

Management:
Biological – delirium screen and lorazepam PRN for agitation.
Psychological – distraction techniques, engagement in singing.
Social – moved to care home environment, training session on CJD offered to care home staff.

Profile 2

Diagnosis: Inherited prion disease (6-OPRI)
Demographics: 50 year old male
Psychiatric/behavioural symptoms: History of reckless conduct in younger years including fighting and polysubstance misuse. As disease progressed, episodes of violence and aggression in response to frustration. In late stages, some resolution of violence due to decreasing mobility.

Management:
Biological - use of regular risperidone and PRN lorazepam.
Psychological – de-escalation techniques, clear boundaries.
Social – move to appropriate nursing home environment, consideration of wider family also at-risk of 6-OPRI.

Illustrative Profiles

Profile 3

Diagnosis: At-risk of inherited prion disease
Demographics: 25 year old female
Psychiatric/behavioural symptoms: Anxiety regarding onset of illness and decision about genetic testing.

Management:
Biological – thorough neurological examination, cognitive testing and reassurance.
Psychological – genetic counselling. Neuropsychology input for management of anxiety symptoms.
Social – Signposted to CJD Support Network.

Conclusion

- The neuropsychiatric manifestations of prion disease are varied.
- There is both marked variation in symptoms across individuals at illness onset, and within an individual as the disease progresses.
- Behavioural and psychological symptoms are common and can prove challenging to manage.
- The profiles presented in this series demonstrate potential approaches to symptom management.
- It is important to consider environmental modifications, psychological support, and where necessary careful use of psychotropic medication.
- Going forwards, standardised treatment protocols, adapting those in use for common disorders, may prove useful.

Acknowledgements

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