Psychosis and Depression in Spinocerebellar ataxias: 2 Case Presentations

Maureen Cassady MD, Joan Han DO, Jennifer Reid MD, Samantha Latorre MD

Department of Psychiatry, University of Maryland School of Medicine/Sheppard Pratt Health System

Introduction

Spinocerebellar ataxias (SCAs) include a number of genetically diverse diseases, commonly known for their motor presentations, typically of progressive ataxia due to cerebellar Purkinje cell degeneration. However, there is also evidence for increased prevalence of psychiatric symptoms in this population, including both a well described cerebellar cognitive affective syndrome (CCAS) and less common case reports of psychosis. We present two patient cases with diagnosis of SCAs and no known psychiatric history who developed depression and psychosis years following motor symptom onset.

Background

Psychiatric Symptoms in SCAs

- Depression and personality change are the most common psychiatric symptoms in patients with SCAs, though anxiety, psychotic disorders and hallucinations are also found. Depression is commonly reported in around 30% of cases, yet some studies have found an even higher prevalence.[5,6,7,11] Significant cognitive impairment among SCA patients has been noted in 25–50% of cases. [5,6,11]

- The presence of psychiatric symptoms is correlated with basal ganglia involvement. [4,9]

- Symptoms of psychosis are less commonly described, though have been detailed anecdotally in several case reports, particularly symptoms of delusions and paranoia. [6, 12]

Non-motor Function of the Cerebellum

- Schmannman & Sherman described cognitive and affective symptoms, or CCAS, in patients with cerebellar lesions: impairment in executive function, personality changes, visuospatial deficits. [4]

- Idea of “cognitive dysmetria” has been proposed for the cerebellum’s possible role in misregulation of cognitive processing. This theory has been explored in research on schizophrenia. [4,6]

- Functional neuroimaging studies demonstrated disruption in cerebello-thalamo-cortical pathways in SCA patients. [3, 8]

Case Report

Case 1: 53 year-old Caucasian male with history of progressive familial ataxia had his first psychiatric encounter when admitted after a suicide attempt by shooting himself in the face. Prior to the attempt, he developed new paranoid delusions about mold infestation in his bodily fluid and living environment, began responding to internal stimuli, and also ingested Lysol days prior to the gunshot. While medically hospitalized, he was guarded due to ongoing paranoia, and olanzapine 2.5mg nightly was started. Neurologically, he had saccadic eye movements, resting tremor of his upper extremities, and ataxia. Neurology diagnosed the patient with SCA given his family history and clinical presentation. Workup of basic labs, EEG, and CSF studies, were without abnormality. MOCA score was 16/30 during hospitalization. MRI was not obtained due to residual ballistic fragments.

He was admitted for inpatient psychiatric treatment where olanzapine 2.5mg nightly was continued until his delusion gradually resolved. His neurovegetative symptoms improved on dextroamphetamine-amphetamine 5 mg twice daily and mirtazapine 30mg nightly. Unfortunately, there was initial difficulty with consistent psychiatric follow up, and, without medications, his delusions and symptoms of apathy returned. He had limited oral intake, as he thought his food was poisoned, and a passive death wish reappeared. He was admitted inpatient again, started on Zyprexa 10 mg nightly and venlafaxine 75 mg daily, with improvement in mood and paranoia. Since discharge, he is followed collaboratively by neurology and psychiatry pending genetic testing.

Case 2: 62 year-old African American male with diagnosis of SCA3 and confirmed pathogenic ATXN3 gene was recommended for psychiatric evaluation by his neurologist in his early 50s for difficulty accepting his diagnosis and symptoms of depression. He deferred evaluation until paranoid delusions of being followed emerged in his mid 50s. He engaged in therapy but refused pharmacological intervention until auditory and tactile hallucinations developed in his late 50s. He experienced hallucinations of two children belittling him while ejaculating on his lower back which led to passive death wishes.

Through the integrated care of psychiatry and neurology he started sertraline 25mg daily and paliperidone 3 mg nightly. To date, his hallucinations and depression have decreased. Intermittent paranoid delusions continue. However, medication and cognitive behavioral therapy have improved reality testing and patient reported quality of life.

Discussion

These cases illustrate the importance of the evaluation of neuropsychiatric symptoms in patients with SCAs, including both common manifestations of depression and cognitive changes, as well as uncommon symptoms of psychosis. In both of these, cases psychiatric symptoms caused severe distress and life-threatening symptoms.

While depression is common in patients with progressive neurological disorders with functional impairment, the prevalence of other neuropsychiatric symptoms in SCAs suggest psychiatric symptoms are, in some part, a result of the neurodegenerative process. [6, 7]

Literature on psychosis in SCAs is still emerging, and there is no clear evidence for best treatment of these symptoms. There are case reports of psychosis treated with second generation antipsychotics,[12] which may potentially reduce the burden of motor symptoms, though evidence is limited regarding efficacy and side effect profile.

Interprofessional collaboration has been crucial in the care of both patients. In similar cases, we recommend consideration of early involvement of psychiatry for longitudinal evaluation and timely pharmacologic intervention.

References

1. Braga-Neto P, Pedroso JL, Gahelha A et al. Psychosis in Machado–Joseph Disease: Clinical Correlates, Pathophysiological Discussion, and Functional Brain Imaging. Expanding the Cerebellar Cognitive Affective Syndrome. Cerebellum (2016) 15:483–490.
2. Fancellu R, Paridi D, Tomasella C, Panzeria M, et al. Longitudinal study of cognitive and psychiatric functions in spinocerebellar ataxia types 1 and 2. J Neurol 2013; 260:3134-3143.
3. Hoppenbrouwers S, Schutter D, Fitzgerald P et al. The Role of the cerebellum in the pathophysiology and treatment of neuropsychiatric disorders: A review. Brain Research Reviews 2008; 59:185-200.
4. Hoche F, Guell Z, Vangel M, Sherman J, Schmahmann JD. The Cerebellar cognitive affective/Schmahmann syndrome scale. Brain 2018: 141; 248-270.
5. Leroi I, O’Hearn E, Marsh L, Lyketsos, CG et al. Psychopathology in Patients with Degenerative Cerebellar Diseases: A Comparison to Huntington’s Disease. Am J Psychiatry 2002; 159:1306-1314
6. Liszewski CM, O’Hearn E, Leroi I et al Cognitive Impairment and Psychiatric Symptoms in 133 Patients with Diseases associated with Cerebellar Degeneration. J Neuropsychiatry Clin Neurosci. 2004: 16:1, 109-112.
7. Lo RY, Figueroa KP, Pulst SM, Perlman S et al. Depression and clinical progression in spinocerebellar ataxias. Parkinsonism Relat Disorder. 2016; 22:87-92.
8. Philips, JR, Hewedi DH, Eissa AM, Moustafa AA. The cerebellum and psychiatric disorders. Frontiers in Public Health. 2015: 3:66.
9. Rottnek M, Riggio S, Byne W, Sano M, Magolis R, Walker RH. Clinical Case Conference: Schizophrenia in a Patient with Spinocerebellar Ataxia 2: Coincidence of Two Disorders or a Neurodegenerative Disease presenting with Psychosis? American Journal of Psychiatry: 2008. 165;8.
10. Rossi M, Farcy N, Starkstein S, Merello M. Nosology and Phenomenology of Psychosis in Movement Disorders. Movement Disorders Clinical Practice. 2020; 7(2); 140-153.
11. Schmitz-Hübsch T, Coudert M, Tezenas du Montcel S, et al. Depression comorbidity in spinocerebellar ataxia. Mov Disord. 2011;26(5):870-876.
12. Turk KW, Flanagan ME, Josephson S et al. Psychosis in spinocerebellar ataxias: a case series and study of tyrosine hydroxylase in substantia nigra. Cerebellum. 2011; 17(2):143-151
13. Wexler E, Fogel B. New Onset Psychosis in a Patient with Spino-cerebellar Ataxia Type 10. Am J Psychiatry, Letters to the Editor. 2010; 168:12