Bilateral chorea following severe traumatic brain injury treated with risperidone

Daniel Krasna , Erica Montgomery, Jacob Koffer, Miriam Segal

Physical Medicine and Rehabilitation, Moss Rehabilitation Hospital, Elkins Park, Pennsylvania, USA

Correspondence to
Dr Daniel Krasna;
krasna.daniel@gmail.com

Accepted 27 April 2021

SUMMARY

A functionally independent man in his 20s with a history of intellectual disability and epilepsy and family history of Huntington's disease suffered a severe traumatic brain injury. Postinjury, bilateral chorea rendered him dependent for all activities of daily living. Risperidone provided a significant reduction of chorea, decreasing the overall burden of care. Movement disorders are a common sequela of brain injury. Currently, there are no best treatment guidelines for chorea in patients with brain injury. To the authors' knowledge there have been no case reports describing the effects of brain injury on patients with a primary movement disorder. Risperidone was an effective treatment in this case. Further research is needed to establish guidelines for treatment of movement disorders following brain injury and to better understand the effect of brain injuries on primary movement disorders.

BACKGROUND

Post-traumatic movement disorders are common neurological sequelae of severe brain injuries. It is estimated that 22% of patients with severe traumatic brain injuries develop movement disorders.[1] Moreover, several post-traumatic movement disorders often co-occur and often in the context of hypertonia and weakness making diagnosis difficult. Post-traumatic chorea is rare.[2]

Chorea is characterised by brief, sudden, unpredictable and large movements of the face and extremities. It often affects the proximal muscle groups and leads to flailing of the arms and legs. Ballism and hemiballism are subcategories of chorea. Ballism is violent, unpredictable flailing of the bilateral arms and legs, whereas hemiballism tends to involve only one limb or the ipsilateral arm and leg. These hyperkinetic movements are due to impaired inhibitory basal ganglia output, and can follow structural injury of the subthalamic nucleus, lenticular nucleus, thalamus and parietal cortex-subcortex.[3]

Medications most effective in the treatment of chorea and ballism are those that interfere with central dopaminergic function.[4] Neuroleptic drugs such as haloperidol, risperidone and chlorpromazine block the function of dopamine, whereas tetrabenazine, a monoamine uptake inhibitor in presynaptic neurons, depletes the amount of dopamine present in the synapse. Tetrabenazine is a first-line agent for the treatment of chorea and ballism due to the rapid onset of action and low side effect profile. However, tetrabenazine is difficult to prescribe because it is only approved by the Food and Drug Administration for patients with confirmed Huntington's disease.[5] Despite these medications, a treatment algorithm for post-traumatic chorea and ballism movement disorders does not exist.

CASE PRESENTATION

A 29-year-old man with a history of intellectual disability, epilepsy since age 9 years, and family history of Huntington's disease in his mother suffered a severe traumatic brain injury after falling during a seizure. A CT head demonstrated small acute subdural haemorrhage and subarachnoid haemorrhage. He subsequently had an additional seizure in the emergency department, was intubated for airway protection, and treated with midazolam and levetiracetam.

His sister reported that he had been enrolled in special education classes and completed high school but did not know his baseline IQ score. He never worked due to a short attention span and collected social security for intellectual disability. About 3 years prior to his brain injury he had a slow, but progressive decline in his speech and cognitive abilities. Two years prior to his injury, his neurologist reported the development of choreoathetoid movements that did not impact him functionally. Despite the concern for Huntington's disease, the family declined testing. However, he was still verbal, cooked his own meals and did not require assistance for his activities of daily living (ADLs). He lived with his father who was able to provide additional support for instrumental activities of daily living (iADLs), such as medications and finances.

One year prior to his brain injury, the patient had a period of altered mental status at home in the evening that persisted into the next morning. He was sent to the hospital where it was presumed he suffered a seizure with postictal state after normal neurological workup. He was given clobazam 10 mg daily, levetiracetam 1500 mg two times per day and valproic acid 500 mg two times per day. On discharge from this stay he required a wheelchair for mobility and assistance with ADLs and was discharged to Skilled Nursing Facility for a few months before returning home. The patient made slight gains but a home health aid was needed for additional support to return to home. Moreover, his linguistic function was impaired. He was able to vocalise basic wants and needs but had difficulty communicating specifics. Outpatient neurology referred the family to a geneticist who again recommended testing for Huntington's disease but this was not completed.

To cite: Krasna D, Montgomery E, Koffer J, *et al*. *BMJ Case Rep* 2021;**14**:e241929. doi:10.1136/bcr-2021-241929

Immediately after his traumatic brain injury he became dependent for all ADLs, iADLs and ambulation. Clobazam was suspended in the acute care hospital and valproic acid was increased to 650 mg two times per day. He could not propel his wheelchair and became non-verbal. His movement disorder was exacerbated and phenotypically resembled severe chorea and ballism involving all extremities and trunk. His wheelchair required extensive padding in order to prevent self-injury. The bed was also modified with rails and padding to prevent injury. His movement disorder interfered with his care, ambulation, safety, and precluded a safe discharge home. The family did not have the capacity to maintain his safety in the home. Placement in a skilled facility was also difficult due to the high burden of care and restraints required to maintain safety. He remained in a rehabilitation hospital for over 4 months while medications were trialled and movement disorder specialists were consulted to best manage his movement disorder.

INVESTIGATIONS

Further investigation gathered while at the rehabilitation hospital showed that the patient's mother and multiple first order relatives were diagnosed with Huntington's disease. The patient's mother passed away before the age of 50 years. There was concern that the patient likely had Huntington's disease prior to his brain injury, however genetic testing was not pursued by the family. Testing was again offered to the family during his rehabilitation stay but could not be completed due to logistics. Repeat CT head 3 months after his brain injury showed stable volume loss and resolution of haemorrhages.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for this patient was broad. Initially, there was concern for a post-traumatic movement disorder. While chorea is possible as a sequela of brain injury, it is far less common than other movement disorders in this patient population.[2] Thus, when it became clear that the movements exhibited by this patient were choreiform, other causes were investigated.

Family history, the patient's preinjury movement disorder, rapid functional decline as well as the presence of bilateral symptoms were all more consistent with chorea from a genetic disorder, most likely Huntington's disease. However, it appeared the patient had a significant increase in chorea frequency and severity after brain injury. Thus, the working diagnosis was the patient had an accelerated neurological deterioration secondary to his brain injury.

TREATMENT

Initially, given the unclear aetiology of the patient's movement disorder, multiple medications were tried. Clonazepam, olanzapine and topiramate were all initiated with varying improvements in his chorea and ballism. The benefits of each drug were outweighed by their negative side effects, mainly oversedation, and was each discontinued. Movement disorder specialists at other institutions were consulted who recommended initiation of tetrabenazine. However, since he was not formally tested for Huntington's disease, tetrabenazine was unavailable during his hospitalisation. Risperidone was recommended as an alternative to tetrabenazine. This was suggested as risperidone has been used in case reports and is the preferred atypical antipsychotic among Huntington's disease experts.[5] Risperidone was trialled and titrated up to 2 mg two times per day. Improvement was noted within days of initiating risperidone.

OUTCOME AND FOLLOW-UP

Risperidone significantly decreased the frequency and intensity of the patient's movements without overly sedating him. He was able to keep himself safely positioned in his wheelchair, transfer more easily and ambulate with his physical therapists. It also became easier for his occupational therapists and nurses to administer care. He sang and danced during therapies, which he had not done prior to initiation of risperidone. Eventually, he was safely discharged to a skilled facility.

DISCUSSION

Risperidone is a frequent choice for the management of chorea in patients with Huntington's disease given its efficacy and favourable side effect profile.[5] There are no guidelines for the management of post-traumatic movement disorders, and scant recommendations for post-traumatic chorea.[4]

Most cases of chorea secondary to brain injury are unilateral.[6] This led us to surmise that the patient's symptoms were more due to his underlying Huntington's disease rather than his traumatic brain injury, though the brain injury may have unmasked his movement disorder.

Our literature review did not find any prior cases of a primary movement disorder that was worsened by traumatic brain injury.

Learning points

- Movement disorders need to be evaluated and properly identified for treatment after traumatic brain injury.
- Risperidone was effective in improving the patient's symptoms and decreasing his burden of care.
- Further research is needed in order to establish guidelines for treatment of movement disorders following brain injury and to better understand the effect of brain injuries on primary movement disorders.

Contributors DK, EM and JK contributed to the research and writing of the case report with DK coordinating the efforts. MS served as the attending physician for all the trainees during the patient's hospital stay, guided the care and interventions, and helped edit the case report for content and clarity.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

ORCID iD

Daniel Krasna http://orcid.org/0000-0001-8555-8289

REFERENCES

1 Krauss JK, Jankovic J. Head injury and posttraumatic movement disorders. *Neurosurgery* 2002;50:927–39.
2 Krauss JK, Tränkle R, Kopp KH. Post-Traumatic movement disorders in survivors of severe head injury. *Neurology* 1996;47:1488–92.
3 Fhan S, Jankovic J, Hallett M. Chorea, Ballism, and Athetosis. In: *Principles and practice of movement disorders*, 2011: 335–49.
4 O'Suilleabhain P, Dewey RB, Richard B. Movement disorders after head injury: diagnosis and management. *J Head Trauma Rehabil* 2004;19:305–13.
5 Coppen EM, Roos RAC. Current pharmacological approaches to reduce chorea in Huntington's disease. *Drugs* 2017;77:29–46.
6 Krauss JK, Jankovik J. Movement Disorders After Traumatic Brain Injury. In: Zasler ND, Katz D. I, Zafonte RD, eds. *Brain injury medicine: principles and practice. Demos medical publishing*, 2007.