Psychosis in Huntington's disease responsive to Electroconvulsive therapy- A case report

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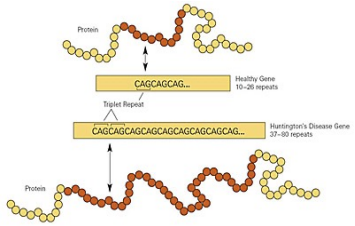
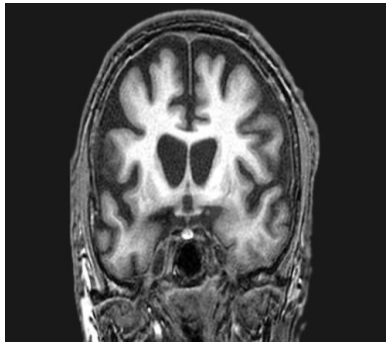
Learning objectives

1. Understand the genetics of Huntington's disease and associated neuropsychiatric symptoms
2. Know the incidence of psychosis as a clinical variant in patients with Huntington's disease and understand the treatment challenges it poses
3. Discuss the role of ECT in managing psychosis in Huntington's disease



Background

Huntington's disease (HD) is an autosomal dominant neurodegenerative disease that is characterized by motor disturbances (chorea, dystonia, bradykinesia, rigidity), cognitive impairment and psychiatric symptoms (depression, irritability/aggression, obsessive compulsive behaviors, apathy, psychosis)[1].



HD results from an unstable and expanded trinucleotide CAG repeat sequence in the Huntingtin (*HTT*) gene on the short arm of chromosome 4, which is expressed as a mutant polyglutamic tract in the protein Huntingtin(Htt).

Mutant Huntingtin protein induces a cascade of cellular changes leading to cell dysfunction and progressive degeneration predominantly in caudate nucleus and putamen. HD typically has a mid-adult onset and a disease duration of 15–20 years. Psychotic symptoms occur in 3%-11% of patients with HD[2]. Neuropsychiatric symptoms can be challenging to treat, are distressing to patients, are burdensome to caregivers and families, and may be predictive of residential care needs.

Case

The patient is a 57-year-old male with a diagnosis of Huntington's disease, who presented initially to the medical hospital with auditory and visual hallucinations, paranoid delusions, impulsivity, depressive symptoms and suicidal ideation. Patient was managed by the Consultation Liaison Psychiatry Service, where multiple medication trials (Quetiapine, Olanzapine, Valproic acid, Escitalopram, Trazodone, Clonazepam) were attempted without benefit. Psychosis continued to worsen and he began refusing food and fluid intake with now accompanying catatonic symptoms (Bush Francis Catatonia Rating Scale Score=6; stupor-1, mutism-2, staring-1, withdrawal-2). Electroconvulsive therapy (ECT) was provided in 14 acute phase treatments resulting in significant improvement in psychosis, catatonia (Bush Francis Catatonia Rating Scale Score=0), depression and overall functioning. He engaged well with the treatment team, was out of bed with assistance, and spent quality time with family and friends prior to his discharge to a long-term care facility. While maintenance phase ECT was recommended, due to COVID-19 restrictions, patient did not receive treatments for about a year. He presented again to the medical hospital with sepsis and during this prolonged admission, began exhibiting worsening irritability, auditory hallucinations and catatonic symptoms (Bush Francis Catatonia Rating Scale Score=10; stupor- 1, mutism-2, staring-1, posturing- 3, withdrawal-3). Multiple medication trials were again attempted. His prognosis was poor and his disease had progressed over the year. He was now bed bound, requiring assistance with feeding and activities of daily living (ADL's). Goals of care meeting was held with family and they hoped that resuming ECT again would provide improvement in quality of life as was evidenced last year. ECT was provided again in 22 acute phase treatments resulting in improvement in catatonic symptoms(Bush Francis Catatonia Rating Scale Score=0), psychosis and general mood. The patient was discharged to his long-term care facility with plans to resume maintenance phase ECT.

Discussion

Psychotic symptoms in HD are thought to be associated with greater cognitive decline, functional deficits, increased incidence of behavioral disturbances, and increased risk for suicide[3]. This case supports that additional consideration should be given to the use of ECT as an adjunct in treatment resistant cases as well as in cases with wider psychiatric manifestations such as mood and psychotic symptoms. ECT may control these symptoms more effectively and reduce overall polypharmacy[4], as was evident in this case.

Conclusion

There is limited evidence with regards to treatment of psychotic and depressive symptoms in HD. Antipsychotics and antidepressants can be useful but other than what is reported in a few case series or case reports, there are no randomized controlled trials to guide treatment choices. ECT can simplify polypharmacy and allow better symptom control in patients with debilitating psychiatric manifestations of HD.

References

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