A Case of Huntington's Psychosis and Neurocognitive Disorder

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Learning Objectives:

1)Recognize psychotic symptoms in patients with Huntington's disease.

2) Offer suggestions for treatment of psychotic symptoms in Huntington's disease.

Case Summary:

A 77 yo female presented to the psychiatry consult-liaison service with altered mental status as the reason for consult. She had symptoms of worsening Huntington's psychosis for the past month, but she has had motor symptoms for the past 10 years, and psychotic symptoms for the past year. She presented with delusions of paranoia and grandiosity as well as auditory and visual hallucinations. Her other mood symptoms included anxiety, labile mood, and OCD symptoms. Her motor symptoms included unsteady gait, imbalance, lack of visual focus, jerking, poor dexterity, slurred speech, and forgetfulness. She was taking Lexapro, Xanax, and Valium from her PCP, in addition to vitamin D and levothyroxine, and did not have significant psychiatric history at the time. Benzodiazepines and the SSRI were discontinued in the hospital. Her family history was significant for Huntington's disease and bipolar disorder. She was smoking cigarettes for the past 10 years but does not have other alcohol or substance use history. She was married, living with her husband, no significant contributory social history. Her concentration and short-term memory recall were limited on interview, with evidence of executive function impairment. She was diagnosed with psychotic disorder due to medical condition, mood disorder secondary to medical condition, and major neurocognitive disorder. The plan was to titrate Risperdal to 0.5 mg BID (started with 0.25 mg initially), place patient with one to one observation, coordinate psychiatric and neurologic outpatient follow up, and patient was found to lack capacity to make her own medical decisions at the time. At 2 week follow-up, she was less anxious with improvement in delusions, including paranoia, and had no more hallucinations since her hospitalization. An in-home aide was coordinated for care.

Conclusions:

Psychotic symptoms, including hallucinations, delusions, and disorganization, may appear years after the onset of motor symptoms of Huntington's disease. In early disease, increased glutamate release onto direct pathway medium spiny neurons leads to increased dopamine tone, and often dopamine antagonists or medicines that decrease dopamine function can be used to treat motor symptoms. However, in later disease, decreased glutamate leads to decreased dopamine tone and hypoactivity, which may be responsible for psychotic symptoms. There are existing case reports of risperidone, a 5-HT2 and D2 antagonist as well as alpha adrenergic and histaminergic antagonist, successfully treating psychosis in Huntington's disease, also with improvement in motor functioning. This case report is likewise consistent. Other antipsychotic medications can be used, and selection can be tailored to specific motor symptoms including chorea and bradykinesia.