CASE REPORT

Psychosis with Huntington's disease: role of antipsychotic medications

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SUMMARY

This is a case of a 60-year-old man who presented with a 6-month history of increasing agitation and emotional volatility. His family brought him to the emergency room as they were concerned about his threatening and aggressive behaviour. The patient was initially incoherent and uncooperative. During the interview, the patient's family revealed that he had a previous diagnosis of Huntington's disease; there was also a family history of personality changes preceding Huntington's chorea. The patient was admitted to the psychiatric inpatient unit and started on low-dose risperidone. Consequently, there was marked improvement in his symptoms. Subsequent cognitive tests revealed deficits in multiple domains. After a month, the patient was discharged to a community home in stable state.

BACKGROUND

Huntington's disease affects 2.71/100 000 individuals globally, and with a prevalence of about 5.7/100 000 in Europe, North America and Australia combined.[1] Of all patients, an estimated 3–11% have psychotic symptoms, which can precede motor symptoms.[2–4]

Psychiatric components to Huntington's disease have long been known.[5] However, clinical recognition of the disease can be difficult. Patients may present with acute psychosis, and chorea may be a side effect of psychotropic medications.[6] We found the following case to be very interesting and would like to share with medical colleagues working in emergency settings and mental health.

CASE PRESENTATION

The patient is an unemployed 60-year-old man who is divorced and living with his former wife, her current partner, his daughter and her partner. He presented at the emergency room (ER) with his family who was concerned about his behaviour.

On questioning, his family revealed increasing agitation and emotional volatility over the past 6 months. The patient had at least one physical altercation with a family member during this period. He had also developed a preoccupation with finances, and became fixated on the idea that his former wife was 'taking his money'. One week prior to presenting at ER, he became agitated and demanded money from his family, which he received. He then left home and was robbed of his money and medications. Over the subsequent days, he visited friends during the day, and slept on a porch at night. On returning home, his family drove him to a local shelter and obtained a restraining order against him. He returned home once more after. This time, he was rambling and verbally abusive. Under the advice of a local organisation responsible for community care, he was brought into the ER.

The patient had no previous episodes of psychosis or psychiatric difficulties. He had seen a psychiatrist once, about 5 months prior, but had refused further involvement. His medical history was significant for hepatitis C and a hernia repair 6 years ago. He had not been on any medications for at least a week prior to admission. He smoked marijuana and cigarettes, with a 60-pack-year history; he also had a history of intravenous drug use at his youth.

He was genetically tested and confirmed for a diagnosis of Huntington's disease about 9 years ago, for which he is followed up by a neurologist. His brother and father had the same illness.

Socially, he was unemployed. He separated from his wife 12 years ago. Together, they had a daughter and a son. He lived with his former wife and daughter, as well as their partners. The family was supportive, but unable to cope with the changes in the patient's mental status over the past 6 months. He previously worked as a welder. His highest level of education was secondary school. He had no known legal history since his youth. Developmentally, he remembered struggling in school with mathematics, but otherwise had done well. He had never been sexually, physically or mentally abused.

He was independent in his activities of daily living , but his ex-wife assumed responsibility for some instrumental activities of daily living, including food preparation and banking.

On examination, the patient was angry, volatile and irritable. He was in fetal position. He turned his face away from the interviewers, and refused to make eye contact. He spoke in short, incoherent sentences. No chorea was noted. No further examination was performed due to the uncooperative nature of the patient.

INVESTIGATIONS

The patient received a complete blood count, serum chemistry, thyroid-stimulating hormone and a toxicology screen on presentation. Alcohol and tetrahyrocannabinon were noted in his urine; there were no other significant abnormalities.

A CT scan revealed no significant intracranial abnormalities.

The patient participated in cognitive testing 1 week after admission. In the clock-drawing test, the patient was able to complete the contour but

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could not place the numbers correctly; Mini-Mental State Examination (MMSE) score was 20/30, with points lost on visuospatial/executive function (−4), attention (−3), language (−1), delayed recall (−1) and orientation (−2). Delayed recall performance was improved by multiple choice cueing.

DIFFERENTIAL DIAGNOSIS

- Traumatic brain injury—ruled out by CT
- Substance abuse—unlikely, given toxicology screen
- Schizophrenia—late-onset schizophrenia could not be ruled out, but an underlying medical condition was present
- Psychosis secondary to Huntington's disease—most likely diagnosis

TREATMENT

Risperidone 0.5 mg orally twice a day was initiated in hospital.

The patient had previously been on citalopram 20 mg orally once daily and amantadine 200 mg orally once daily. Citalopram was continued in hospital. Given amantadine's dopaminergic effects, it was tapered to 100 mg orally daily for 5 days and then discontinued.

Medical staff at the hospital also prescribed tiotropium 18 µg inhaled once daily. For a possible pneumonia, he was prescribed cephalexin 500 mg orally four times a day for the first 7 days of his hospital stay.

OUTCOME AND FOLLOW-UP

The patient's symptoms improved in hospital. He was more cooperative with a psychiatric interview after 2 days; however, he remained agitated and incoherent at this point. One week postadmission, a psychiatric reassessment found his behaviour to be normal. His mood volatility and periodic agitation resolved more slowly and returned to baseline in 2 weeks. His preoccupation with finances subsided. His relationship with his family recovered. Cognitive tests revealed moderate cognitive impairment, with difficulties in reading, abstract thinking, judgement, attention, memory and mathematical skills.

Later, the patient indicated that personality changes were the predominant feature of Huntington's disease in his family. Both his father and brother's first Huntington manifestation were argumentativeness, irritability and perseveration of ideas, followed by motor symptoms. The patient himself had noticed small involuntary motor movements over the past year, which was manageable so far.

After a month as an inpatient, he was discharged to a community home in stable state.

DISCUSSION

Huntington's disease is a neurodegenerative disorder characterised by motor disturbances, progressive dementia and psychiatric illness. Research and clinical emphases have been on motor and cognitive aspects.[7] Among the psychiatric components, depression, irritability, apathy and anxiety are the most common presentations, with prevalences of 33–76%.[2 8] Psychosis is less common. In acute psychosis, recognising an underlying Huntington's disease without the stereotypic choreiform movements presents a diagnostic challenge. This case presents such a dilemma that was resolved with collateral information from a supportive family.

The treatment of psychosis in Huntington's disease is a topic of some discussion. A retrospective study demonstrated that risperidone was effective over the period of 1 year in reducing psychiatric symptoms.[9] We are aware of only three case reports in which risperidone was used to treat psychosis successfully with a prior diagnosis of Huntington's disease: one of a 55-year-old woman treated with risperidone 1 mg twice daily; the second of a 40-year-old woman treated with risperidone 1 mg twice daily; and the third of a 32-year-old-woman treated with risperidone 4 mg daily.[10–12] Our patient received a lower dose of risperidone than the previous cases. Other case reports have also highlighted atypical antipsychotics in patients with Huntington's disease, including aripiprazole and quetiapine.[13 14]

In summary, a patient presented with an episode of psychosis. His family provided collateral information which revealed a previous diagnosis of Huntington's disease. A medical cause should therefore be considered in any differential for psychotic patients. Furthermore, a low-dose regimen of risperidone was successfully used to treat the psychosis. This case adds to the literature supporting the use of atypical antipsychotics in Huntington's disease.

Learning points

- Huntington's disease should be on the differential for late-onset psychosis.
- If confirmed, cognitive testing should be administered to determine the extent of neurological deficits.
- Psychosis associated with Huntington's disease can be treated with low-dose risperidone.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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