Catatonia: A Rare Presenting Symptom of Wilson's Disease

To the Editor: Psychiatric manifestations may precede neurological signs in the early stages of Wilson's Disease (WD). Catatonia is rarely associated with WD. Here we discuss a 19-year-old adolescent patient who initially presented with catatonia and was later found to have WD. The differential diagnosis and management is discussed.

Wilson's disease (WD) is a rare, autosomal recessive genetic disorder of copper metabolism caused by mutations in the ATP7B gene.[1] The exact lifetime prevalence of psychiatric disorders in patients with WD is not known, but is estimated to range between 30% and 100%. The psychiatric manifestations of WD can be categorized into five groups of symptoms: personality changes, affective disorders, psychosis, cognitive impairment, and others.[2] Personality change, affective disorders including depression, and cognitive impairment are common, and schizophrenia-like psychoses are rare.[2,3] Catatonia is a syndrome that encompasses more than two dozen signs, some of which are relatively nonspecific. There is a single case report of catatonia as a presenting symptom of WD.[4]

Case Report

A 19- year-old adolescent patient from a rural background presented to the Department of Psychiatry with abnormal behavior. He did not suffer from any psychiatric illness in the past. There was also no history of neurological or psychiatric illness among the family members. The patient is the third of the four children born of a consanguinous marriage. Two months earlier, the patient had a high-grade fever for 2 days, not associated with chills and rigor, rash and vomiting. There was no history of seizures or myoclonic jerks. He was investigated by a physician, and no cause for fever could be found; he was treated symptomatically. After a week family members noticed that he was shouting without any reason, restless, had irrelevant speech, suspiciousness, odd behavior, frequent eye blinking, and abnormal body movements. After 2 days, he was found to be staring at one place, maintaining abnormal postures for long periods, with decreased speech, decreased food intake, and with episodes of excitement. For these complaints, he was brought to the department of psychiatry for further management. He was initially diagnosed with acute, schizophrenia-like psychosis. In view of catatonic symptoms, he was started on a trial of lorazepam 6 mg/day in divided doses for 2 days, but he failed to show any improvement. He was also started on olanzapine 5 mg, and the dose was gradually increased to 20 mg/day. In view of poor oral intake and catatonic symptoms not responding to trial of lorazepam, modified electroconvulsive therapy (ECT) was started. After five ECT treatments, the patient showed significant improvement and was discharged. After 1 week, catatonic symptoms worsened in spite of his being on medication. Tablet olanzapine was changed to trifluoperazine 15 mg/day, with trihexyphenidyl 6 mg/day in divided doses. After 1 week, he was found to be restless, with decreased arm swing, stooped posture, excessive salivation, and tremor of extremities. Patient was readmitted for evaluation of excessive sensitivity for extrapyramidal symptoms (EPS) with antipsychotics. Trifluoperazine was stopped, and EPS improved, but there was worsening of catatonic symptoms. On examination, patient was afebrile, and vital parameters were within normal limits. Patient had cogwheel-type rigidity, short stepping gait, mask-like face, and excessive salivation. On mental status examination, patient had echolalia, echoprexia, maintaining abnormal posture for long period, negativism, staring, mutism, perseveration, sudden excitement, and delusion of persecution. Bush-Francis catatonia rating scale score was 30 on admission.

As he was young boy and born of a second-degree consanguinous marriage and was sensitive for EPS, Wilson's disease was considered and investigated. On investigation, hemoglobin, WBC total and differential count, platelet count, SGOT, SGPT, blood urea, serum creatinine, fasting blood sugar, and urine microscopy were within normal limits. Serum calcium, phosphorus, and iron were also within normal limits; 24-hour urinary copper excretion was 2,165 μg/day (normal value: 32–64μg/day), serum ceruloplasmin of 45 U/lt (normal value: 62–140 U/lt) and serum copper 160 μgm/dl (normal value: 70–150 μgm/dl). MRI of brain did not reveal any hyperintensities. There was no Kayser-Fleischer (KF) ring on slit lamp examination of the eye. Neurophysician opinion was sought, and they diagnosed him to be suffering from Wilson's disease. He was started on oral zinc 200 mg/day. In absence of KF ring and copper deposition in the brain, chelating agents were not started.

The patient was started on quetiapine 100 mg/day, and gradually dose was

increased to 600 mg/day in divided doses. Simultaneously, he was given 6 more ECTs over a 2-week period. Patient was completely symptom-free at the end of 1 month. Patient is on regular follow-up for the last year; quetiapine was tapered and stopped in 6 months, and he was only on zinc supplements. There was no recurrence of catatonic symptoms. Patient is advised to have 6-month monitoring of urine copper excretion. His 24-hours copper excretion after 1 year was 431 μg/day (normal value: 32–64μg/day).

Discussion

Psychiatric manifestations may precede neurological signs in the early stages of WD. About 20% of them precede hepatic and neurological dysfunction.[5] An Indian study of a series of 350 patients with WD showed that in 15 patients (4.29%), psychiatric manifestations were the predominant symptom at the time of medical consultation.[6] Psychiatric manifestations in WD may depend on deposition of copper in various parts of the brain.

In the present case, catatonia and use of antipsychotic drugs in the patient has unmasked WD. Common differential diagnosis for young-onset extrapyramidal syndrome include Wilson's disease, young-onset Parkinson's disease, Hallervoden-Spartz disease, Huntington's disease, subacute sclerosing panencephalitis, and hypoparathyroidism. Care should be taken while treating WD patients with antipsychotic drugs. Quetiapine and clozapine, which have minimal extrapyramidal side effects, are preferred. ECTs can be safely used in patients with WD,[7] and, in the current case, there were also no complications during the course of ECT.

Conclusion

Catatonia is a rare presenting symptom of WD. The antipsychotic drug with low propensity for extrapyramidal side effects needs to be used for treatment of comorbid psychiatric symptoms with WD. ECT is the safe option for treatment of catatonia in patients with WD.

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