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CASE REPORT

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Successful treatment of psychosis in dentatorubral-pallidoluysian atrophy with quetiapine: A case report

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Abstract

Introduction: Patients with dentatorubral-pallidoluysian atrophy (DRPLA) sometimes elicit psychosis. First-generation antipsychotic drugs have been reported to be effective in treating psychotic symptoms associated with the disease. However, little information is available on the benefits of second-generation antipsychotic drugs (SGAs).

Case: We report on a 47-year-old man with DRPLA whose psychotic symptoms were effectively treated with quetiapine, one of the SGAs. He suffered from delusions, auditory hallucinations, and disorganized speech. Initially, other antipsychotic drugs were tried, but were withdrawn because of adverse effects before switching to quetiapine.

Conclusion: Our observations add to the notion that some of the SGAs are useful for ameliorating psychosis in DRPLA.

KEYWORDS

antipsychotics, progressive myoclonic epilepsies, psychosis, second generation, spinocerebellar degenerations

1 | INTRODUCTION

Dentatorubral-pallidoluysian atrophy (DRPLA) is one of the spinocerebellar degeneration diseases, which is caused by mutation of the atrophin-1 (ATN-1) gene on human chromosome 12p13.31.[1] In DRPLA, CAG repeats in the ATN-1 gene are expanded.[1] The morbidity of the illness in Japan is approximately 0.2-0.7/100 000.[2] One study reported that the occurrence of expanded CAG repeats are more frequent in Japan compared with other countries,[3] while non-Japanese population may also be vulnerable to DRPLA, with a morbidity higher than has been considered.[4] Patients with DRPLA typically show epileptic seizures, myoclonus, ataxia, and dementia.[5,6]

Patients with DRPLA occasionally elicit psychosis, and its treatment has yet to be established. Adachi et al[7] reported that first-generation antipsychotics drugs (FGAs), such as haloperidol and levomepromazine, were effective in treating psychosis of DRPLA. However, little information is available on whether second-generation antipsychotic drugs (SGAs) are useful.[8] Here, we report on a patient with DRPLA whose psychotic symptoms were successfully treated with quetiapine.

2 | CASE

Mr. A was a 47-year-old Japanese man. Ten years before consulting us, mild cerebellar ataxia and choreoathetosis emerged in him. Two years later, he experienced repeated epileptic seizures, which were controlled by co-administration of levetiracetam, clobazam, and clonazepam. Mild upper-limb dystonia and ataxic dysarthria developed next year, which encouraged him to visit the department of neurology of our hospital. These symptoms were gradually aggravating in him, but his physical condition was basically stable. Brain images with magnetic resonance imaging demonstrated cerebellar and

brainstem atrophy. Genetic examination revealed 59 CAG repeats in exon 5, which confirmed the diagnosis of DRPLA. His father and grandmother suffered from similar symptoms, although no genetic examinations were performed on them.

Three months before consulting us, cerebellar ataxia and choreoathetosis started to progress rapidly, producing gait disturbances. As a result, his daily living activity declined dramatically. Two months later, he was not able to move without a wheelchair and spent most of time lying on a bed. This progression was thought to represent deterioration of DRPLA. Several days before consulting us, he started to say aggressive words, such as "I will kill you." When we saw the patient for the first time, his speech was disorganized. For example, he uttered "One, two, three, go." and "Please delete all of my memory." In addition, the hallucinatory-paranoid state dominated. For example, he believed that his "child" was dead, although he actually did not have one. Moreover, he complained about hearing weird sounds, such as "You are a dead man." His memory was not disturbed, indicating the patient was unlikely to suffer from dementia. Electroencephalography recording was normal.

Psychotic disorder due to DRPLA was diagnosed in Mr. A. Choreoathetosis was treated with haloperidol at 0.75 mg/d for 2 months, and subsequently, fluphenazine at 0.5 mg/d for 1 week. However, both antipsychotics were withdrawn because tremor occurred. Then, 2.5 mg/d olanzapine was given for 2 months, but was discontinued because of concurrent nausea/vomiting. Thus, choreoathetosis was not ameliorated. For psychotic symptoms, we started giving quetiapine at 50 mg/d (before sleep) as initial dose. We titrated it to 300 mg/d (once a day) to effectively ameliorate his symptoms. Six weeks after the start of the medication, the hallucinatory-paranoid state was improved so that he no longer experienced auditory hallucination, delusion, and disorganized speech. Treatment with quetiapine was not accompanied with adverse effects.

The patient became observant and insightful enough to be adherent to quetiapine. Several weeks later, he moved to another hospital in order to undergo rehabilitation because cerebellar ataxia and ADL continued to deteriorate.

3 | DISCUSSION

To our knowledge, this is the first report of the ability of quetiapine, one of the SGAs, to improve psychotic symptoms associated with deterioration of DRPLA. Accordingly, Sato et al[9] reported that DRPLA sometimes progresses rapidly, followed by the development of psychosis. Schizophrenia is occasionally misdiagnosed in patients with psychosis due to DRPLA.[10,11] To identify DRPLA, it is necessary to carefully examine the history of present illness. In our patient, psychotic symptoms started on deterioration of DRPLA, consistent with the diagnosis of psychotic disorder due to DRPLA.[12] We started administrating antipsychotic drugs soon after our patient had complained about psychotic symptoms, indicating the absence of duration of untreated psychosis. This may have been a reason for the favorable prognosis in terms of psychotic symptoms.

The relative paucity of information on psychosis in DRPLA may be due to the low incidence of the disease. On the other hand, Adachi et al[7] reported approximately 10% of patients with DRPLA suffer from psychosis. Antipyschotic drugs are not always recommended for the treatment of organic mental disorders because of a greater incidence of adverse effects, such as sedation and extra-pyramidal symptoms (EPS).[13] The use of FGAs is generally associated with side effects, especially EPS, which are less common with SGAs.[14-16] Specifically, quetiapine elicits a more favorable profile regarding EPS compared with olanzapine and risperidone,[16,17] which encouraged us to use it.

We previously reported the ability of olanzapine, one of the SGAs, to improve psychotic symptoms associated with DRPLA.[8] In our patient, olanzapine was added to treat choreoathetosis, but was withdrawn because of adverse events. Therefore, we administered quetiapine, an antipsychotic drug known to have a favorable pharmacologic profile in terms of EPS, compared to other antipsychotics.[18] Both olanzapine and quetiapine possess a considerable affinity for serotonin-5-HT_{2A} (relative to dopamine-D_2) receptors, as well as others, including acetylcholine and noradrenaline receptors.[19] The observed effectiveness of the two SGAs may provide a promising strategy for treating psychosis associated with DRPLA.

A limitation of the current report is that we did not quantitatively evaluate severity and change of psychotic symptoms. In spite of this, the benefit of quetiapine to treat psychosis associated with DRPLA is encouraging, given the minimal chance of producing EPS by the drug. Further investigations with a larger number of patients are required to generalize the effectiveness of quetiapine.

CONFLICT OF INTEREST

The authors declare no conflict of interest for this article. [Correction added on 5 March 2018, after first online publication: The words, 'for this article', have been added to the back of the conflict of interest statement.]

INFORMED CONSENT

Informed consent was obtained from the patient.

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