

Successful Treatment with Olanzapine of Psychosis in Dentatorubral-pallidoluysian Atrophy: A Case Report

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Patients with dentatorubral-pallidoluysian atrophy occasionally elicit psychosis. So far, one study reported first generation antipsychotics drugs may provide an effective treatment; however, there is no literature on the benefits of second generation antipsychotics. We report on a 44-year-old man with dentatorubral-pallidoluysian atrophy whose psychotic symptoms were effectively treated with olanzapine. Our observation suggests some second generation antipsychotics provide a therapeutic option for ameliorating psychosis in dentatorubral-pallidoluysian atrophy.

KEY WORDS: Progressive myoclonic epilepsies; Spinocerebellar degenerations; Psychosis; Antipsychotics.

INTRODUCTION

Dentatorubral-pallidoluysian atrophy (DRPLA), one of the spinocerebellar degeneration diseases, is a genetic autosomal dominant condition caused by an expansion of CAG repeats.^{1,2} The morbidity of the illness in Japan is estimated approximately 0.2-0.7/100,000.³ Although Burke *et al.*⁴ reported that expanded CAG repeats occur more frequently in Japan compared to other countries, Warner *et al.*⁵ argues that non-Japanese population may also be vulnerable to DRPLA, which would be more common than generally considered.

Patients with DRPLA typically show symptoms, such as epileptic seizures, myoclonus, ataxia, and dementia.^{6,7} Also, these patients sometimes experience psychosis, but its treatment has yet to be established. For example, Adachi *et al.*⁸ reported first generation antipsychotics drugs (FGAs), such as haloperidol and levomepromazine, ameliorated psychosis in DRPLA. However, there is no in-

formation on the efficacy of second generation antipsychotics (SGAs), based on the search of MEDLINE and PsycINFO.

Here, we report on a patient with DRPLA whose psychotic symptoms were effectively treated with olanzapine.

CASE

The patient was a 44-year-old Japanese man. One year before consulting our hospital, cerebellar ataxia and choreoathetosis had developed. Also, he had started hearing weird voices and showing hyperactivity. These symptoms had not been sufficiently improved with sodium valproate at 1,200 mg/day, which rendered him to consult us. Brain imaging with computed tomography demonstrated cerebellar and brainstem atrophy, and genetic examination confirmed the diagnosis of DRPLA. His mother also received the diagnosis of DRPLA, and has been hospitalized for many years because of severe physical conditions.

When the patient was admitted to our hospital, the hallucinatory-paranoid state dominated. For example, he believed that his “younger sister” married a 65-year-old man, although he actually does not have a sister. Also, he complained about auditory hallucinations, for example, hearing sounds as if a surgeon was performing an operation.

Psychotic disorder due to DRPLA was diagnosed in the

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patient, and we started treatment with olanzapine, at 2.5 mg/day (before sleep) as initial dose. We titrated it to 7.5 mg once a day to effectively ameliorate his symptoms. Four weeks after the start of medication, the hallucinatory-paranoid state was improved, so that he no longer heard weird sounds or elicited hyperactivity. Throughout the treatment, he did not experience adverse effects, such as extrapyramidal syndromes (EPS), and follow-up blood test results were normal.

When the patient consulted us for the first time, he did not understand the necessity of treatment for psychotic symptoms. At the time of discharge, he became more observant and insightful enough to be adherent to medications. He is seeing us regularly and keeping treatment with olanzapine at 7.5 mg/day.

DISCUSSION

To our knowledge, this is the first report of the ability of olanzapine, one of the SGAs, to improve psychotic symptoms associated with DRPLA. When the patient showed the hallucinatory-paranoid state, he was fully conscious, not delirious, a condition similar to the patient with schizophrenia. In fact, schizophrenia has been sometimes misdiagnosed in patients with psychosis due to DRPLA.^{9,10} To identify DRPLA, it is necessary to carefully examine the history of present illness. In our patient, neurological and psychotic symptoms started simultaneously, consistent with the diagnosis of psychotic disorder due to DRPLA.¹¹ He had been treated with sodium valproate in previous hospitals soon after complaining about these symptoms, indicating the absence of duration of untreated psychosis.

The relative paucity of information on psychosis in DRPLA may be due to the low incidence of the disease itself. On the other hand, Adachi *et al.*⁸ reported approximately 10% of patients with DRPLA complain psychosis, suggesting the importance of information on its treatments. For patients with organic mental disorders, antipsychotics are not always recommended because adverse effects tend to occur frequently.¹² The use of FGAs is generally associated with side effects, such as EPS, which are less frequent with SGAs.^{13,14} Therefore, we administered olanzapine to the patient, as it has favorable profile in terms of the incidence of EPS compared to other antipsychotics.¹⁵ Hence, the effectiveness of olanzapine,

not associated with adverse effects, observed here, may provide a promising strategy for treating psychosis in DRPLA.

A limitation of this study is that we did not use quantitative measures, such as the Positive and Negative Syndrome Scale and the Brief Psychiatric Rating Scale, to evaluate severity of psychosis. This issue warrants further investigations for the benefit of SGAs to treat psychosis with DRPLA.

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