

Modified electroconvulsive therapy for the treatment of refractory schizophrenia-like psychosis associated with Huntington's disease

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Dear Sirs,

Huntington's disease (HD) is a progressive neurodegenerative disorder characterized by involuntary movements, dementia and psychiatric symptoms [1]. Psychiatric symptoms in HD resemble those of schizophrenia; therefore HD is sometimes clinically misdiagnosed as schizophrenia [1–3].

The efficacious effect of modified electroconvulsive therapy (mECT) has been reported in both psychiatric disorders and in movement disorders including dyskinesia and Parkinson's disease [4, 5]. However, there have been few reports of mECT for the treatment of schizophrenia-like psychosis in HD. Here, we describe a case of a patient with HD who had psychiatric symptoms refractory to neuroleptics, and whose symptoms were dramatically improved by mECT.

A 59-year-old man was admitted to our hospital because he had auditory hallucinations and delusions of persecutions. His mother had been diagnosed with dementia resulting in long-term hospitalization. His clinical characteristics were mostly unremarkable, except for imperceptible involuntary movements of the lip. Initial treatment using risperidone (to 6 mg/day) was ineffective and caused extrapyramidal side effects; subsequently, his medication was changed to olanzapine (20 mg/day). However, his psychiatric symptoms showed no improvement.

Based on treatment-refractory psychosis, we then introduced mECT. After the fourth treatment with mECT, his delusions and hallucinations were dramatically improved and he regained fluent speech, although his involuntary lip

movements were still present. During the patient's serial mECT treatment, his elder brother had visited the department of neurology in our hospital complaining of involuntary movement and was subsequently diagnosed with HD by a genetic test. This finding led us to surmise that the patient's psychiatric symptoms were associated with HD. The genetic test for HD revealed that the patient had 44 CAG repeats. Thus, we recognized the patient's clinical symptoms as psychiatric features of HD.

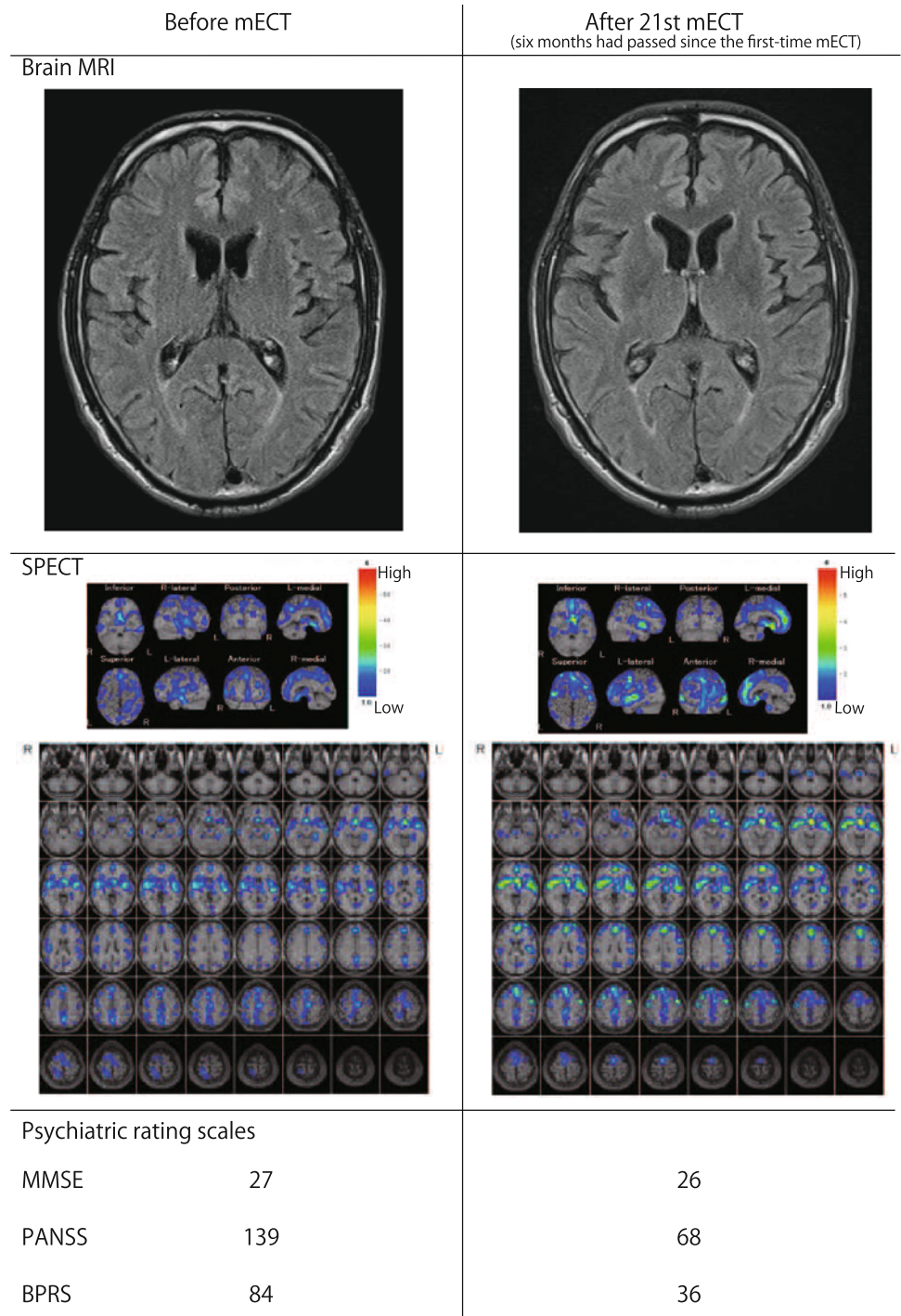
Subsequently, we have continuously performed mECT and the patient has shown no psychotic exacerbation. Single-photon emission computed tomography (SPECT) scans, brain magnetic resonance imaging (MRI) images, mini-mental state examination (MMSE) scores, positive and negative symptom scale (PANSS) and brief psychiatric rating scale (BPRS) scores before and after mECT are summarized in Fig. 1.

Several studies have reported that patients with HD frequently have accompanying psychiatric symptoms [1–3]. The frequency of schizophrenia-like psychosis ranges from 3 to 12 % of patients with HD [6]. Before the identification of the *HTT* gene, dementia or schizophrenia-like psychosis associated with HD that displayed little involuntary movement would have been misdiagnosed as Alzheimer's disease or schizophrenia, and would have led to long-term hospitalization as in the case of the patient's mother. Our case suggests that a renewed focus on taking a detailed family history into account should be applied when considering the possibility of HD, especially if early-onset dementia and/or late-onset psychosis exists in other family members.

To date, there is just one report of mECT for the treatment of schizophrenia-like psychosis associated with HD [7]. Neuroleptics are a standard treatment of psychosis in HD; however, the effect of pharmacotherapy on psychosis in HD remains unclear. Furthermore, neuroleptics cause abnormal movements due to extrapyramidal side effects

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Fig. 1 Comparison of brain MRI, SPECT and psychiatric rating scales before and after mECT. All data from the SPECT images were analyzed using an easy Z-score imaging system. The brain MRI before mECT showed slight atrophy of the subcallosal gyrus, similar to after mECT treatment. The SPECT after mECT treatment showed a level of 99mTc uptake in the basal ganglia, cingulate gyrus and thalamus that was greatly decreased compared with the analysis before mECT



[8]. In this particular case, mECT dramatically improved psychosis associated with HD without worsening involuntary movements. The fact that mECT does not cause extrapyramidal side effects raises the possibility that mECT could be a first-line treatment of schizophrenia-like psychosis in HD (Table 1).

Recently, Mughal et al. [9] demonstrated that ECT could protect neurons against mutant huntingtin protein, resulting in improved functional outcome and leading to slow

disease progression. In agreement with Mughal et al., the results of brain MRI and MMSE scores of before and after treatment showed that the cognitive function of our patient remained clinically unchanged. Several studies have indicated that the changes in SPECT reflect progression of the disease [10]. Furthermore, van den Bogaard et al. [11] suggested that it is difficult to detect the visible structural changes of HD patients at 2-year follow-up. Unchanged MMSE scores and MRI images may not be due to the

Table 1 The pros and cons of mECT treatment versus neuroleptics treatment for psychosis in Huntington's disease

	Merit	Demerit
mECT	More potent effect	Cumbersome introduction
	Fewer side effect	Need particular equipment
	No absolute contraindications	High cost Anterograde/retrograde amnesia and transient cardiovascular complications as side effects
Neuroleptics	Easy introduction (easy to take)	Lethal side effects, including malignant syndrome and cardiovascular complications Worsening abnormal movement due to extrapyramidal side effect

mECT treatment, but be a natural course of the disease. Together with the SPECT results, we conclude that the patient's pathological state may progress, which may indicate that mECT does not arrest the progression of HD.

Conflicts of interest The authors report no disclosures related to the current case presentation.

Ethical standard This study has been approved by the appropriate ethics committee and has therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

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