Late-onset oligosymptomatic myotonic dystrophy type 1 mimicking prodromal dementia with Lewy bodies

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Letters to the Editor

Late-onset oligosymptomatic myotonic dystrophy type 1 mimicking prodromal dementia with Lewy bodies

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Myotonic dystrophy type 1 (DM1) is caused by a CTG expansion in the DM protein kinase (DMPK) gene; furthermore, there is a correlation between CTG repeat size and age of onset.[1] There have been few reports on the psychiatric features of DM1 in older patients because patients with adult-onset DM1 have a shorter life expectancy,[1] and late-onset oligosymptomatic DM1 often remains undiagnosed. Here, we describe a case of late-onset oligosymptomatic DM1 mimicking prodromal dementia with Lewy bodies (DLB) accompanied by severe psychosis.

A 71-year-old woman had been suffering from visual hallucinations and mild cognitive impairment (MCI). She had a history of cataracts in her 40s. When she was 69 years old, her two grandchildren were diagnosed with childhood-onset DM1; both had CTG expansions in the DMPK gene of around 850 repeats. Subsequently, she was diagnosed as having late-onset oligosymptomatic DM1, based on detection of 60-90 CTG repeats. At 71 years old, she presented with detailed visual hallucinations and MCI. Thereafter, she suffered from auditory hallucinations and persecutory delusions for 3 months. She had become aggressive and incoherent, and had bitten off part of her little finger after being persuaded by her auditory verbal hallucinations. Therefore, she was admitted to our hospital. Upon admission, routine laboratory investigations were within normal ranges, except for mild elevation in C-reactive protein (4.85 mg/dL) and creatine phosphokinase (221 U/L). **Blonanserin administration was started to alleviate her severe psychotic symptoms and**

psychomotor excitement. However, she showed intense drowsiness when the dose of blonanserin was increased up to 16 mg/day. Therefore, we had to reduce it to 8 mg/day. Electroencephalogram revealed no abnormalities. Brain magnetic resonance imaging revealed diffuse and marked bilateral occipital cortical atrophy, and T2-weighted diffuse high-intensity signaling in white matter, but no parahippocampal gyrus atrophy. Single-photon emission computed tomography demonstrated hypoperfusion in the exterior occipital lobe and a decrease of cardiac ^{123}I-meta-iodobenzylguanidine uptake, but no reduction of striatal ^{123}I-dopamine transporter uptake. Neurological examinations revealed mild parkinsonism, but no myotonic symptoms. Her Mini-Mental State Examination score was 23, indicating mild impairments in short-term memory and calculation. Therefore, she was diagnosed as having probable DLB according to the criteria for clinical diagnosis.[2] **After diagnosis of probable DLB, blonanserin was replaced with memantine.** Her psychotic symptoms were improved under memantine 10 mg/day, and she was discharged.

This older case with DM1 exhibited the unique psychiatric manifestations including schizophrenia-like psychosis. Previous studies have suggested that adult DM1 has been associated with particularly paranoid and aggressive personality traits.[3] In addition, aberrant default mode network functional connectivity has been suggested to underlie schizotypal-paranoid personality traits in adult DM1.[4] Our case suggests that the DM1-associated neurobiological and psychological alterations may also cause psychotic

symptoms in older patients. Alternatively, her psychotic symptoms might have been due to psychiatric-onset DLB, which is prodromal DLB characterized by primary psychiatric disorders.[5] The unique clinical presentations of this case might suggest that the psychiatric manifestation reflects an interaction between DM1 and DLB rather than a single brain pathology. However, we could not confirm a definite clinical interaction between DM1 and DLB, because the confirmation of Lewy bodies by autopsy has not been conducted. Further studies are needed to elucidate the association of psychotic symptoms with DM1 in older patients, as well as the clinical interaction between DM1 and DLB.

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AUTHOR CONTRIBUTION

All authors contributed to the study conception and design. The acquisition of data and interpretation of the clinical data were performed by Tomoyasu Wakuda, Riho Yoshida, Asuka Sakurai, and Hidenori Yamasue. The first draft of the manuscript was written by Tomoyasu Wakuda, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

DISCLOSURE

The authors declare that they have no conflict of interest.

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