Abstract

Plasma Coenzyme Q10 Levels in Patients With Multiple System Atrophy.

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IMPORTANCE: Multiple system atrophy (MSA) is an intractable neurodegenerative disease characterized by autonomic failure in addition to various combinations of parkinsonism, cerebellar ataxia, and pyramidal dysfunction. It has recently been reported that functionally impaired variants of COQ2, which encodes an essential enzyme in the biosynthetic pathway of coenzyme Q10 (CoQ10), are associated with MSA. However, little is known about the role of CoQ10 in the pathogenesis of MSA.

OBJECTIVE: To compare the levels of plasma CoQ10 in patients with MSA with those in age-, sex-, and COQ2 genotype-matched controls.

DESIGN, SETTING, AND PARTICIPANTS: We enrolled 44 Japanese patients with MSA and 39 Japanese controls from September 1, 2012, to December 31, 2015. Patients with MSA were diagnosed on the basis of the second consensus criteria by at least 2 neurologists. Plasma CoQ10 levels were measured by high-performance liquid chromatography with electrochemical detection. Sanger sequencing of COQ2 was performed to determine the COQ2 genotypes. Multiple logistic regression analysis was performed to determine the association between MSA and the plasma CoQ10 level.

MAIN OUTCOMES AND MEASURES: Plasma CoQ10 levels in patients with MSA were compared with those in controls after adjusting for age, sex, and COQ2 genotype.

RESULTS: Among 44 patients with MSA (mean [SD] age, 63.7 [8.3] years) and 39 controls (mean [SD] age, 60.3 [13.0] years), the mean (SD) plasma level of CoQ10 in patients with MSA was lower than that in controls (0.51 [0.22] vs 0.72 [0.42] µg/mL; P = .01) (difference between medians: -0.14; 95% CI, -0.25 to -0.03). The mean (SD) plasma levels of CoQ10 in patients with the cerebellar variant of MSA and those with the parkinsonian variant of MSA were 0.58 (0.19) and 0.49 (0.26) µg/mL, respectively. After adjusting for age, sex, and COQ2 genotype, the levels of plasma CoQ10 were significantly associated with MSA (95% CI, 0.10; range, 0.02 to 0.66) (P = .02).

CONCLUSIONS AND RELEVANCE: Our data showed decreased levels of plasma CoQ10 in patients with MSA regardless of the COQ2 genotype, supporting a hypothesis that supplementation with CoQ10 is beneficial for patients with MSA.

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