

Response to reviewers' comments:

1. Since Cockayne is an inherited disease, family history, especially the situations of her parents should be illustrated.

Parents of this patient had no symptoms. No family history was noted as well. We explain the details at the paragraph of “history of past illness” (p.6, line 146-147).

2. Line 152, please explain the evidence or basis of the dose of 210 units of the drug.

We add the dosage in 15 units/kg at the first paragraph of treatment (p.7, line 153-154). We also explained the recommended dose based on European consensus in 2009 at the paragraph of “discussion” (p.10, line 244-246).

3. (1) Try to elucidate the possible mechanism of Botulinum neurotoxin type A the disease, and why its efficacy could only be maintained a short period. (2) If it possible that intermittent administration of this drug could get sustained improvement? Try to demonstrate.

We addressed mechanism of botulinum toxin type A (BoNT-A) at the paragraph of “discussion” (p.9, line 216-220). BoNT blocks the acetylcholine release, which results in temporary and reversible chemical denervation. The nerve terminals will reestablish the new neuromuscular junctions. That is the reason that the efficacy of botulinum toxin type A is only maintained for short period.

Intermittent administration of this drug could provide sustained improvement. Safety and effectiveness of repeated BoNT-A injection every 3 months have been proved. We add the contents and references at paragraph of “discussion” (p.9, line 221-223).