Author’s response to reviews

Title: “Neuromyelitis optica spectrum disorder with massive basal ganglia involvement: a case report”

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To the comments by Dr. Sara Mariotto (reviewer 1).

Abstract
・ The term antibodies to AQP4 should be used (here and through the main text)
Ans) “AQP4 antibody” is changed to “antibodies to AQP4” here and through the mail text accordingly.
・ “who” has to be avoided.
Ans) It is deleted in the revised text.

Background
・ The term “NMO” should be avoided, according to the recent criteria of NMOSD (Windgerchuk at al Neurology 2015)
Ans) We used the term “NMOSD” and deleted “NMO” throughout the revised text, accordingly.

・ AQP4-Abs are not always present, and this should also be clarified.
Ans) Although antibodies to AQP4 is a cardinal serum biomarker of NMOSD, it is known that approximately 20 to 25% of patients of NMOSD are AQP4Ab seronegative. Accordingly, we changed the 1st paragraph of Background section.
“Antoantibodies to the water channel aquaporin-4 (AQP4), which is predominantly expressed in astrocyte foot processes, is a serum biomarker and is expressed in a majority of the cases with this syndrome [1].”

Case presentation
· Treatment choices should be better explained (i.e. the choices to not to administer long-term immunosuppressants other than steroids after AQP4-Abs positive result).
Ans)
As far as we are aware, no completed randomized controlled trials of NMOSD is available for preventive treatment of NMOSD, although preventive treatment is strongly recommended. We believe low maintenance dose (15mg) of oral predonisone was an appropriate choice in this patient, which has been described in the text.
· The patient should have received a diagnosis of NMOSD also in the absence of AQP4-Abs so the sentence “she was diagnosed as having NMO based of the positive AQP4 antibody” is not completely correct.
Ans) We agree that the patient could have diagnosed as AQP4-Abs seronegative NMOSD based on current criteria (Windgerchuk at al Neurology 2015). Accordingly, we changed the sentence from “she was diagnosed as having NMO based of the positive AQP4 antibody” simply to “Serum antibody to AQP4 was positive, and she was diagnosed as having NMOSD”.
· Could aspergillosis of the lung have a role? This should be clarified.
Ans) We had performed Grocott methenamine silver staining for fungi and confirmed that the necrotic lesion in the basal ganglia was negative for the staining. This finding is added in the Text.
“ The Grocott methenamine silver stain was negative in the necrotic lesions.”

References
· A homogenous style has to be adopted.
Ans) We have made necessary changes and the total number of references is reduced from 16 to 14 in the revised text.