Author's response to reviews

Title: A patient with ALS and atypical clinical and electrodiagnostic features: a case report

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I appreciate the reviewers’ comments.

In response to the first reviewer’s comments, I have added to the manuscript amplifying on the uniqueness of the case. I have amplified my presentation of the relevant literature in the Discussion. I believe that I have discussed all such material and do not feel an additional table would add to this.

The second reviewer raises several issues:

• There is no known family history of similar problems in a large well, known family. As such, there is no reason to think this was a familial type of ALS. SOD1 studies were not performed on this patient and can obviously not be performed at this time. Given the patient’s presentation and family history, SOD1 studies at the time of the patient’s evaluation did not seem indicated.

• There was no evidence for conduction block or temporal dispersion. This is now stated in the text.

• Lewy body-like and skein-like inclusions were seen in the spinal cord and are so indicated in the text. A photograph showing Lewy body-like inclusions has been added. As best as I can determine, vacuolar degeneration in the descending motor pathways is not unusual in ALS.

• Although TDP-43 studies may have been of interest, these were not commonly being performed at the time this patient was evaluated. I am also not certain these would necessarily have added more definitive information about this patient. As I am certain the reviewer is aware, TDP-43 is not necessarily specific for ALS nor, if absent, rule it out. (e.g., Rothstein JD. TDP-43 in amyotrophic lateral sclerosis: pathophysiology or pathobabel. Ann Neurol 51:382-384, 2007).

There have been some changes in the references.

I would be glad to address any additional issues if that would be helpful.