Reviewer's report

Title: Brain imaging in Kufs disease type B. Case report

Version: 3 Date: 1 June 2015

Reviewer: Alfried Kohlschuetter

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This is a short and clearly written description of neuroradiological findings in two siblings with a rare degenerative brain disease, manifesting itself in young adulthood. Knowledge of the results may be helpful in suspecting and diagnosing this genetic condition.

Minor Essential Revision

The findings described include abnormalities of cerebral white matter. In the abstract, as well as in line 104, the authors refer to these abnormalities as "demyelination", which is speculative and probably not correct. The T2 hyperintensity of cerebral brain white matter is probably caused by an increased water content of these brain areas due to the loss of neurons and not by the decay of myelin sheaths. This view is supported by the neurological signs described, which are not suggestive of damage to long myelinated tracts. So better avoid the term demyelination and stay with "white matter abnormalities" as in other parts of the manuscript.

Level of interest: An article of importance in its field

Quality of written English: Acceptable

Statistical review: No, the manuscript does not need to be seen by a statistician.

Declaration of competing interests:

I declare that I have no competing interests.