Dear Editor-in-Chief,

Manuscript Number: 75638

Title: Creutzfeldt-Jakob disease presenting with bilateral hearing loss: A case report

We sincerely appreciate your kind consideration. The authors would also like to thank the reviewers for their insightful comments on our manuscript. We have revised the manuscript according to the reviewers' comments. We hope that the revised manuscript is suitable for publication in *World Journal of Clinical Cases*. Below we provide a point-to-point response to the reviewers' comments. Reviewers' comments are in bold and the revised texts are highlighted. We have highlighted the changes in the revised manuscript as well.

Reviewers' comments:

Reviewer #1: In this paper, a case of sporadic Creutzfeldt-Jakob disease (sCJD) with bilateral hearing loss at onset was reported. Combined with literature review, the authors suggest that for patients with abrupt bilateral hearing impairment, especially in the elderly, sCJD should be considered one of various differential diagnoses, and repeat brain imaging including diffusion-weighted imaging and CSF analysis would be helpful for diagnosis. The article needs minor revisions, see the red text of the manuscript.

➔ We appreciate your detailed comments. We corrected the typos and revised the manuscript as you pointed out.

Reviewer #2:

1. **I didn't find any figures in the submitting system. Did the authors forget to upload these figures?**

➔ We apologize for the mistake. We uploaded the figures through the editor's help.

2. Since Sporadic Creutzfeldt-Jakob disease (sCJD) is a prion disease, the authors did not mentioned the possibility of how to get the prion.

➔ Thank you for your comments. He was diagnosed with sporadic CJD according to the diagnostic criteria of sCJD and he showed the typical feature of sCJD in the brain imaging study. And we excluded the possibility of iatrogenic CJD because he had never undertaken neurosurgery dealing with a dura meter or a lumbar puncture procedure until the visit. He also conducted the genetic analysis and there was no mutation or polymorphism of the prion gene. It means that familial CJD was excluded. Thus it would be difficult to clarify the origin of prion in this patient, since it was sporadic case. We revised the manuscript as below:

➔ "(Line 112) Considering all the data, we diagnosed probable sCJD. The amended diagnostic criteria added that the combination of cognitive decline, positive CSF RT-QuIC, and one or more typical CJD symptoms can draw the diagnosis of probable sCJD [3]. Because he had never undertaken neurosurgery dealing with a dura mater and the genetic analysis revealed that there was no mutation or polymorphism of the prion gene, we excluded the possibility of familial CJD or iatrogenic CJD. Moreover, our patient revealed a positive RT-QuIC test and met the criteria of probable sCJD."

Reviewer #3(Science editor)

This manuscript describes a case of Creutzfeldt-Jakob disease who showed only bilateral hearing impairment initially but eventually progressed to dementia. The author need to discuss the possible origin of the prion. Please add more references if possible and highlight the new information this case may provide.

➔ We appreciate for your comments. We revised the discussion section of manuscript and added a reference. The highlight of the new information is written in the ‘core tip’ and ‘conclusion’ section. We revised the manuscript as below:

➔ “The possibilities of familial or iatrogenic CJD were excluded because there was no mutation or polymorphism of the prion gene and he had no history of epidemiological evidence. Although the etiology of sCJD has been unknown, many researchers assumed that the prion disease might be initiated by the stochastic misfolded cellular prion protein or mutations in the prion protein gene at ongoing neurogenesis areas [4].”