

# Reply to letter regarding the article entitled: ‘Rapidly progressive sporadic Creutzfeldt-Jakob disease: isolated Heidenhain variant or a combination with PRES?’

Resposta à carta referente ao artigo intitulado: ‘Doença de Creutzfeldt-Jakob esporádica rapidamente progressiva: variante Heidenhain isolada ou PRES combinada?’

Pedro Henrique Almeida FRAIMAN<sup>1</sup>, Carolina Militão TEIXEIRA<sup>1</sup>, Juliano César Dantas DE OLIVEIRA<sup>2</sup>, Thadeu Alexandre Paulino DE SOUSA<sup>2</sup>, Manuel MOREIRA-NETO<sup>3</sup>, Clecio de Oliveira GODEIRO-JUNIOR<sup>1</sup>

Dear Editor,

We thank Onder<sup>1</sup> for his valuable comments regarding our article<sup>2</sup> on the manifestation of probable Creutzfeldt-Jakob Disease (CJD) with Posterior Reversible Encephalopathy Syndrome (PRES).

First, we report a case of a patient with a pretty clear neurodegenerative course of the disease. Regarding the diagnosis of CJD, we believe that it is very likely because of clinical, imaging, electroencephalographic and cerebrospinal fluid findings – rapid-onset cognitive impairment with ataxia and myoclonus; diffusion-weighted images (DWI) revealing bilateral hyperintensities in parietal areas, cingulate gyrus, and basal ganglia; periodic sharp wave complex and positive 14-3-3 in cerebrospinal fluid<sup>3</sup>.

Second, we deeply agree with the author that the persistence of the early neuroimaging findings after the acute-onset

visual agnosia would support the diagnosis of CJD, as shown in the figures of our article and described in Figure 1 original legend<sup>2</sup>.

Then, we recognize that temporal evolution due to limitations of space could be more precise. Still, we believe that it is clear that neurodegeneration is not associated with PRES, but with CJD in this case, given that the cognitive dysfunction, ataxia, and myoclonus were present before acute-onset of visual loss as it can be seen in the article<sup>3,4</sup>.

Moreover, the association of DCJ and PRES has been previously described in the literature<sup>5</sup>, and the main focus of our article is a discussion of acute-onset visual loss is a manifestation of CJD Heidenhain variant - recognized by visual loss and occipital cortex hyperintensities in T2-FLAIR imaging - or a PRES manifestation associated with CJD<sup>5,6</sup>.







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<sup>1</sup>Universidade Federal do Rio Grande do Norte, Hospital Universitário Onofre Lopes, Divisão de Neurologia, Natal RN, Brazil.

<sup>2</sup>Casa de Saúde São Lucas, Divisão de Radiologia, Natal RN, Brazil.

<sup>3</sup>Universidade Federal do Rio Grande do Norte, Hospital Universitário Onofre Lopes, Divisão de Radiologia, Natal RN, Brazil.

PHAF  <https://orcid.org/0000-0002-5250-8602>; CMT  <https://orcid.org/0000-0003-0990-5044>; JCDO  <https://orcid.org/0000-0002-3079-6885>; TAPS  <https://orcid.org/0000-0001-9568-3411>; MMN  <https://orcid.org/0000-0002-3478-5323>; COGJ  <https://orcid.org/0000-0002-4312-1633>

**Correspondence:** Clecio Godeiro-Junior; Email: [cleciojunior@yahoo.com.br](mailto:cleciojunior@yahoo.com.br).

**Conflict of interest:** There is no conflict of interest to declare.

**Authors' contributions:** PF, CMT, JCDO, TAPS: writing and drafting the manuscript; MMN, COGJ: critical revision of manuscript for intellectual content.

Received on August 12, 2021; Accepted on August 13, 2021.