Reviewer's report

**Title:** Sporadic Fatal Insomnia in Texas: A pre-mortem diagnostic challenge: Case report

**Version:** 1  **Date:** 29 July 2011

**Reviewer:** Stephane Haik

**Reviewer's report:**

The authors report in detail a case of sporadic fatal insomnia (sFI). sFI is a highly challenging diagnosis and the authors, in light of the clinical history of the case reported, underscore the importance of recording sleep alterations in the context of subacute dementia. Some of the diagnostic tests that are useful for diagnosing sFI are detailed. This informative report is a valuable addition to the literature. A few alterations may improve its clinical interest:

**Discretionary Revisions:**

The authors might want to add some EEG data as a figure and to discuss further the EEG findings.

The authors should detail the MRI scans that were performed. Were diffusion-weighted imaging and MR-spectroscopy performed? In addition, it is not clear whether a thalamic atrophy was observed or not at the second brain MRI.

Variant CJD is discussed among alternative diagnoses. For the non-specialist readers, it would be helpful to give in few words some basic information on vCJD and its link to the bovine spongiform encephalopathy. In addition, in countries where vCJD has occurred, it should be mentioned that, in the context of a young patient with a suspected diagnosis of CJD, psychiatric initial symptoms and no pulvinar sign on MRI, a tonsil biopsy might be useful.

It may be of interest to discuss the sensitivity of 14-3-3 protein detection in patients with this peculiar subtype of sCJD.

Page 5: “the hippocampal formation, basal ganglia and cerebellum were much less affected...”. This sentence is not clear to me and the authors may wish to reformulate.

Page 6: last sentence and page 7: first sentence. The genetic testing is mentioned in both sentences.

**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