Reviewer’s report

Title: Amyotrophy in Creutzfeldt-Jakob disease

Version: 1 Date: 1 May 2013

Reviewer: Tibor Kovács

Which of the following best describes what type of case report this is?: Unexpected or unusual presentations of a disease

Has the case been reported coherently?: Yes

Is the case report authentic?: Yes

Is the case report ethical?: Yes

Is there any missing information that you think must be added before publication?: Yes

Is this case worth reporting?: Yes

Is the case report persuasive?: Yes

Does the case report have explanatory value?: Yes

Does the case report have diagnostic value?: Yes

Will the case report make a difference to clinical practice?: No

Is the anonymity of the patient protected?: Yes

Comments to authors:

This is a very interesting case and the paper is well written in general, although I have some minor comments and notes related to it.

1. Firstly, there are several disturbing typing mistakes:
   Abstract, first line: "Creutzfeld-Jakob" Creutzfeldt
   Keywords: "Creutzfeld-Jakob" Creutzfeldt
   There is an upper case 4 in line 6 of the introduction and 5 in line 8 of the Discussion.
   Case presentation: "paraspinals muscles" paraspinal muscles
   Reference 6: "CSD" CSF, "creutzfeldt-jakob"
   Reference 10: Lantos PL (last author) is missing
Reference 11: "creutzfeldt-jakob"
Reference 12: "-Jackob"

2. Citations 1 and 2 cited in the third line of the introduction are too specific; a general review paper about prion diseases might be more suitable.

3. Normal cognition is mentioned at the time of initial presentation of the patient, however the score on clock drawing test was 4 and MMSE was 25/30. These should raise some concern about the normal cognitive function.

4. The authors state that the patient had no mutation in the PRNP gene; the codon 129 polymorphism should be mentioned (this might influence the clinical phenotype). In addition, prion type (Collinge or Parchi nomenclature) (if known) should be mentioned.

5. Amyloid pathology was found but the authors did not mention neurofibrillary degeneration (present or not, if yes, Braaks' stage?).

6. What about the prion immunohistochemistry of the brainstem motoneurons?

Tibor Kovács MD, PhD

**Quality of written English:** Acceptable