Rapidly progressive dementia with positive 14-3-3 protein: description of a case with partial recovery.

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Objectives:

Rapidly progressive dementia are insidious conditions with a complex differential diagnosis. In last years, the literature has put more attention on neuroradiological findings and on sensibility and specificity of cerebrospinal fluids analysis. We describe a rapidly progressive dementia in a patient with deep brain lesions of gray and white matter and positive 14-3-3 protein and Tau in CSF, who presented partial neurological recovery at 12 months follow-up.

Materials and methods:

Male patient, 61 years old, was admitted in Psychiatric Unit for acute psycotic manifestations, then he was admitted in Clinical Surgery Department because of concomitant severe acute bowel occlusion. He was operated. During the hospitalization he developed rapidly progressive dementia complicated with visual allucinations, difficult in mentalization and confusion, vertical nystagmus, gait disorders and important weight loss. Based on clinical data, we have performed brain MRI, CSF examination with dosage of proteins of neuronal damage (14-3-3 and tau), serial EEG and consecutive follow-up..

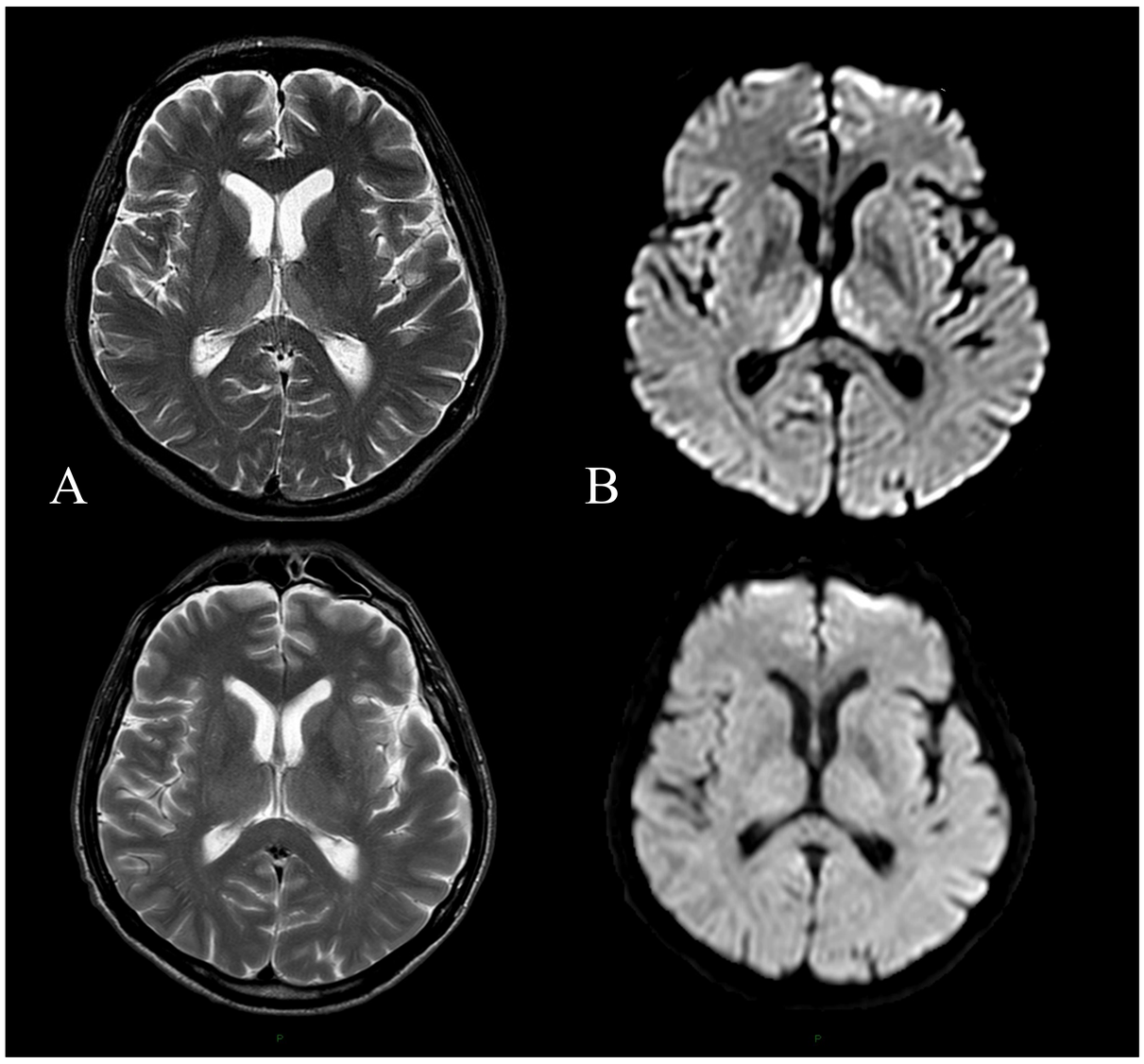
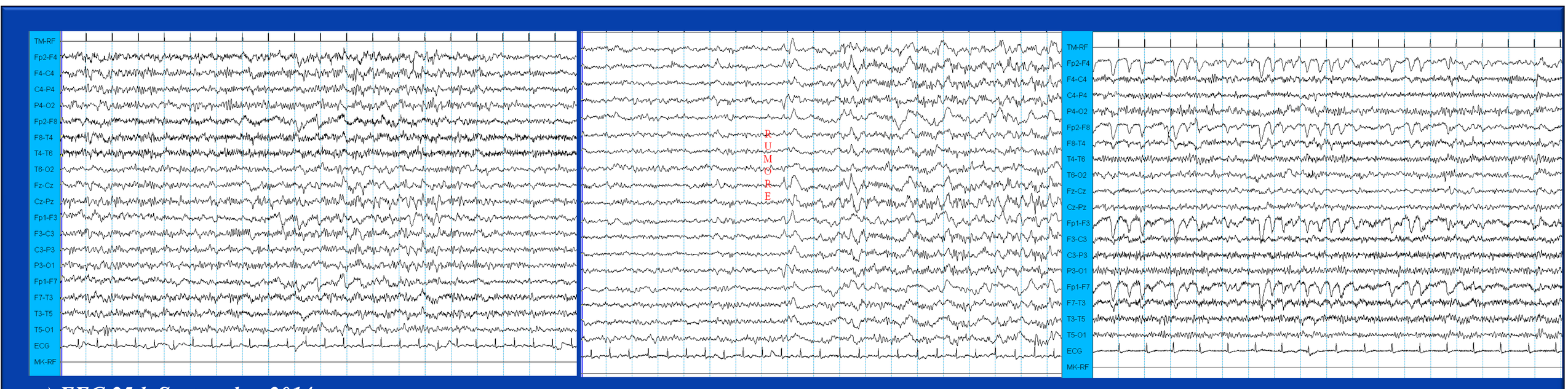


Fig.1 : Baseline and 3 months follow-up- An. axial T2; B. DWI

Results: Neuroradiological, neurophysiological and laboratoristic examinations excluded brain infection, neoplastic, vascular, iatrogenic or autoimmune etiology. Brain MRI T2 weighed images showed hyperintensity of pulvinar and ventral medial region of thalamus bilaterally and CSF analysis evidenced positive 14-3-3 protein and Tau protein. EEG showed not specific generalized slowing .Twelve months follow-up documented cognitive and behavioral disorders recovery associated to EEG and MRI findings normalization. Persisted severe cerebellar ataxia



a) EEG 25th Septermber 2014 ***b) EEG 25th September 2014*** ***c) EEG 10th September 2015***

Discussion: We describe a patient whose CSF 14-3-3 protein analysis and brain MRI suggested a Creutzfeldt-Jakob disease (CJD) diagnosis. At 12 months follow-up showed a good cognitive recovery and normalization of EEG and imaging findings. These results had put into question the diagnosis. CJD has a progressive and fatal course. The heterogeneous clinical features of CJD make diagnosis difficult and confirmation of the presence of CJD requires evaluation of brain tissue. Literature data underlie how 14-3-3 could be falsely positive and emphasize that the assay is an adjunctive but not confirmatory procedure in diagnosis of CJD.
Therefore, based on clinical data we hypothesized a Wernicke encephalopathy and the restoring nutrition also with administration of group of B-vitamines laid to significant clinical improvement.

Conclusion: This case emphasizes the importance of the clinical context of each patient and underlies caution respect to overreliance on putative biomarker and MRI findings in the CJD diagnosis. Differential diagnosis of rapidly progressive dementias is broad and includes primary neurodegenerative, inflammatory, metabolic, vascular, and malignant conditions. The clinical diagnostic accuracy is relevant to avoid missing treatable disorders for evident prognostic implications. .

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