LATE STRIATAL INVOLVEMENT AND PROLONGED DISEASE COURSE IN A CASE OF sCJD: A 14-MONTH LONGITUDINAL 3T-MRI STUDY

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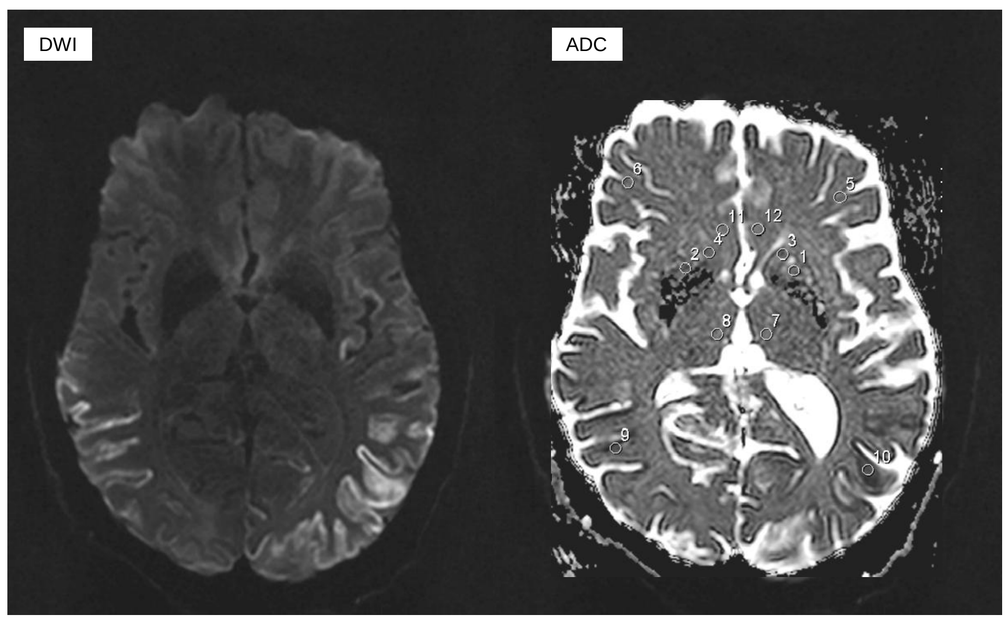
Background

Sporadic CJD (sCJD) is a transmissible neurodegenerative disorder with a fatal outcome, characterized by tissue deposition of a misfolded isoform of the cellular prion protein (PrPC), commonly referred to as PrPSc . In literature patients with long clinical course were reported (*Shiga 2007*) and, in these cases, absence of characteristic patterns on EEG and on cerebrospinal fluid test was observed (*Parchi 1999; Collins 2006; Sanchez-Juan 2006).* While in sCJD the pathognomonic MRI presentation is cortical ribboning and striatal hyperintensity with diffusion restriction, isolate cortical involvement was reported in about 30% of cases (*Vitali 2011*). Moreover, hyperintensity in DWI more than FLAIR, hypointensity in ADC in the same regions and not esclusive limbic involvement discern sCJD from the other non-prionic rapidly progressive dementias (npRPD).

To our knowledge, there is no published study on longitudinal MRI changes in sCJD. Here we report serial changes of diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) in a sCJD case with long disease's course.

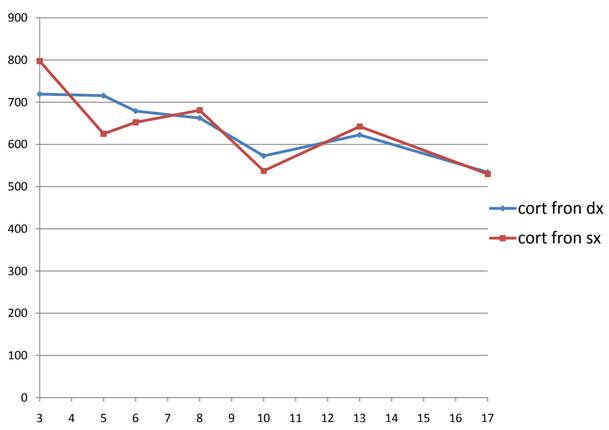
Materials and Methods

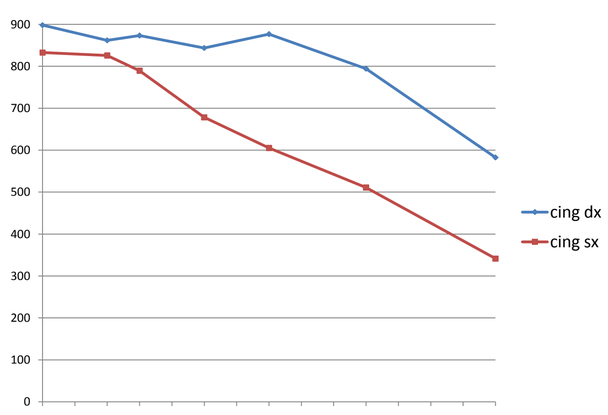
A 79 years-old woman presented with mild limb ataxia, postural instability and initial impairment of executive and memory functions. In the following 10 months, we observed a rapid and progressive cognitive involvement, with aphasia, apraxia, visuo-spatial impairment and severe amnesia. In the last 7 months, motor impairment became severe, with diffuse severe rigidity of limbs and trunk, myoclonic tremor and akinetic mutism. Seven MRI scans were acquired by a Siemens Skyra 3T scanner at 3, 5, 6, 8, 10, 13, and 17 months after symptoms onset. In every scan FLAIR and DWI were acquired and ADC was measured by circular ROI positioned over the same location in frontal, temporal, cingulate cortex and putamen, of both hemispheres.

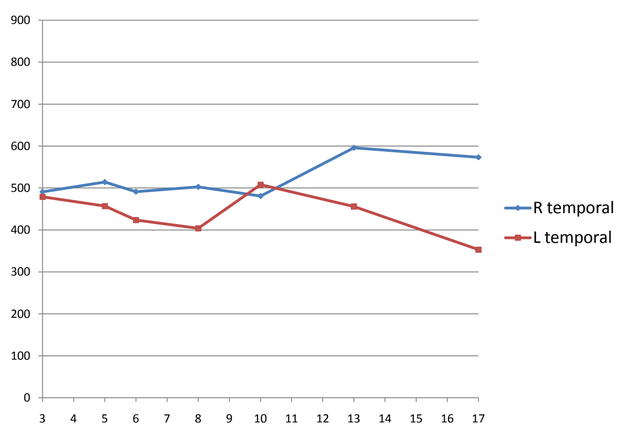


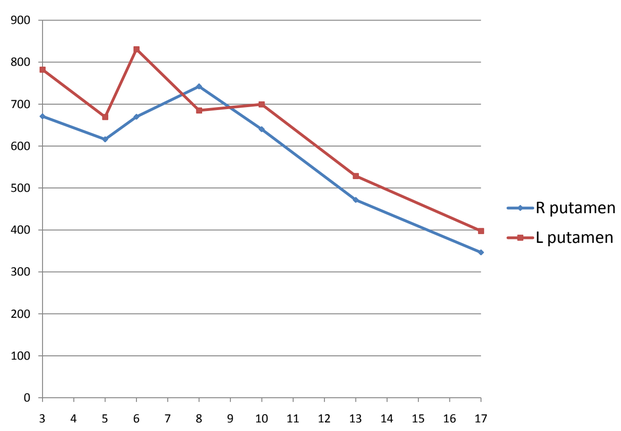
Results and Discussion

Visual inspection of DWI revealed diffuse cortical ribboning since the first scan, with posterior prevalence and left hemispheric dominance. This isolate cortical without striatal involvement remained stable until 10 months after symptoms onset, when a subtle hyperintensity of left external putamen was observed. A bilateral involvement of striatum (both putamen and caudate head) with typical anterior-posterior gradient appeared only in the last two scans, 13 and 17 months after symptom onset. The ADC measurement better evaluates the observed diffusion changes: substantial stability in the temporal cortex and progressive diffusion restriction in the frontal and cingulate cortex and in the putamina.









Serial MRI scans, with DWI and ADC, track the cortico-subcortical spreading of prion disease in sCJD. In our case, we have observed a progressive involvement of the frontal and cingulate cortex during the first ten months of disease. In this period, the clinical consisted of only a slow worsening of cognitive functions and ataxia. An acceleration of disease was instead observed after the 10th month, when it appeared diffuse rigidity and akinetic mutism. In MRI, this change corresponded to the involvement of putamina and the fall of the signal in ADC in basal ganglia.

On the contrary, the serial ADC scans showed a relative stability of the signal in the right temporal cortex, with only a minimum increase, as result of the process of “pseudo-normalization” (*Caverzasi 2014*).

From what we have reported, it can thus be assumed that, in cases with isolated cortical ribboning, striatal involvement can occur very late in the disease's course and it can correlate with a longer duration. This agrees with the data in the literature: the codon 129 genotypes that are associated with a late involvement of basal ganglia, have a longer survival *(Meissner 2009)*.

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