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The alien limb

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Background: Among the neurodegenerative diseases, the Sporadic Creutzfeldt-Jakob disease is one of the most complex to detect. It can start with no-specific symptoms, undergoing a daily rapid change, until death in few months.

Case presentation: We describe a case of Sporadic Creutzfeldt-Jakob disease presenting with a lateralized cortico-basal syndrome as first symptom. In particular, the patient came to our attention for an alien limb syndrome with apraxia/ataxia involving the right hand and a scanned speech. No lack of strength was noticed. At the beginning only some epileptic anomalies in the left parietal lobe were noticed, in absence of important alteration in neuroradiological studies. We saw a daily variation of the clinical picture, until a constellation of symptoms appeared, including bilateral apraxia-ataxia, myoclonus, axial rigidity, with difficulties in postural changes, and palilalia, with repetition of syllables and meaningless words. After three weeks we had an EEG revealing bilateral periodic triphasic waves and an MRI showing FLAIR hyperintensity in different cortical and subcortical regions. Moreover, the CFS analysis revealed a positive RT-QuIC test. According to the diagnostic criteria we were in front of a probable Creutzfeldt-Jakob disease [1]. After two months since the beginning of first clinical manifestations, our patient died.

Conclusions: First symptoms in Creutzfeldt-Jakob disease are variable, making the diagnosis insidious. Furthermore, neuroradiological and neurophysiological examens are not always suggestive of the disease at the beginning. Our case shows a particular start of the disease with a cortico-basal syndrome, and more specific an alien limb phenomenon as first symptom. Not so many cases have been described in literature. Indeed, around 23 cases of cortico-basal syndrome as initial Creutzfeldt-Jakob disease have been reported and only 6 of them with an alien limb syndrome [2]. A rapid clinical picture changing could help to direct the clinician focus towards such a disease.

References:

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