

Overlap between Creutzfeldt-Jakob disease and paraneoplastic limbic encephalitis

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Objectives: Creutzfeldt-Jakob disease (CJD) or Subacute Spongiform Encephalopathy is a very rare neurodegenerative disease of the brain that belongs to the prion diseases category. Limbic encephalitis (LE) is frequently associated with neoplasia, the most common incriminated being the pulmonary cancer (80%). An overlap of those two diseases is incredibly rare. Our patient exhibits signs and symptoms that suggest this rare occurrence.

Background: Keeping in mind the similar clinical characteristics (cognitive impairment, personality changes, depression, anxiety, hallucinations, convulsions), sometimes it is difficult to differentiate between CJD and LE, especially at the onset of the disease. The spinal fluid can be positive for the "14-3-3" protein in both pathologies but the presence of inflammatory cells and the Hu Antibodies are frequently present in LE. On the EEG, high voltage, slow (1-2 Hz) and sharp complexes on an increasingly slow and low-voltage background are suggestive for CJD.

Case description: Our patient is a 66 year old female, who in the absence of fever, presented a confusional state and in 6 months developed focal seizures. The IRM examination showed hyperintensity in the bitemporal area including the limbic zone. The EEG was suggestive for CJD. The spinal tap identified the Hu antibodies and the 14-3-3 protein. On the chest Rx examination we could see micronodules in both pulmonary fields. Our patients, evolution was a rapidly progressive one, she continued to have seizures which gradually became generalized (in 3 weeks) in spite of the maximal antiepileptic treatment. The patient became comatose and died due to infectious complications. The postmortem examination established the diagnosis of pulmonary neoplasia with small cells and the brain showed structural changes suggestive for CJD.

Conclusions: The particularity of this case consists in the overlap of those two rare neurological pathologies and the difficulty in establishing the accurate diagnosis.