Autoimmune encephalitis mimicking Creutzfeldt-Jakob disease

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Background: Differential diagnosis of severe progressive dementia includes a wide spectrum of inflammatory and neurodegenerative diseases. Particularly challenging is the differentiation of potentially treatable autoimmune encephalitis and Creutzfeldt-Jakob disease. Such a coincidence may indeed complicate the correct diagnosis and influence subsequent treatment.Case presentation: A 75-year-old woman was admitted due to rapid progressive cognitive impairment. Her husband observed a temporal disorientation and confusion. The initial neurological examination and an extensive neuropsychological evaluation showed significant impairments in almost all tested cognitive domains. All other neurological functions including motor, sensory and coordinative function were intact. Initial diagnostics included EEG, MRI and lumbar puncture with unspecific results. Complementary blood testing revealed a positive result for antineural antibodies to Contactin-associated protein 2 (CASPR2) and the patient received treatment for CASPR2 autoimmune encephalitis. Further symptoms and results, including 14-3-3 proteins, led to suspected Creutzfeldt-Jakob disease. The postmortem examination supported the diagnosis of a definitive Creutzfeldt-Jakob disease.Conclusion: One could argue that global screening for antineural antibodies may lead to a false diagnosis triggering intense and potentially dangerous procedures. We believe, however, that potentially treatable causes of dementia should aggressively sought out and subsequently treated in an attempt to curtail the course of disease and ultimately reduce the rate of mortality.
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