

Do Severe Acute Respiratory Syndrome Coronavirus 2 Vaccines Change Creutzfeldt-Jakob Disease Prognosis?

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To the Editor,

Creutzfeldt-Jakob disease (CJD) is a rare and fatal neurodegenerative disease. It is caused due to misfolding of prion proteins, which cannot be proteolyzed and continue to accumulate in the neurons, leading to degeneration and inflammation. Genetic, sporadic, and acquired forms of this disease have been described. The sporadic form accounts for 85-95% of cases, and 1-2 cases per million individual are reported annually.¹ We report the case of a 59-year-old man suspected of having CJD with an expected course of the disease, who developed symptoms following multiple severe acute respiratory syndrome coronavirus-2 (SARS-CoV-2) vaccinations. Written informed consent was obtained from the patient's wife. A 59-year-old man with subacute gait problems for 2 months was suggested neurosurgery. He was diagnosed with lumbar spinal stenosis and underwent surgery. However, after the surgery, his gait worsened, he became ataxic, and began experiencing difficulty in finding words. He had two episodes of alien hand syndrome and was therefore hospitalized. He was never infected with SARS-CoV-2. He received two doses of the SARS-CoV-2 vaccine (Sinovac®) voluntarily. After approval of the same vaccine in Turkey, he was vaccinated twice. After the approval of mRNA-based vaccine BNT162b2 (Pfizer-BioNTech®), he was vaccinated twice again. His gait complaints started 2 weeks later. The diffusion-weighted magnetic resonance imaging (DWI-MRI) performed at a neurology clinic revealed suspicious caudate, putaminal, and cortical hyperintensities. Moreover, we observed periodic slow waves in his electroencephalogram (EEG). After the observation of myoclonic jerks, levetiracetam was prescribed. He was evaluated for differential diagnosis of CJD, autoimmune,

and paraneoplastic limbic encephalitis. No signs of malignancy or autoimmunity were observed during the investigation. Lumbar puncture revealed slightly elevated protein levels. Therefore, 1 g pulse methylprednisolone was administered for 5 days prior to cerebrospinal fluid (CSF) markers results. He insisted for an mRNA-based vaccine BNT162b2 vaccination repeat and received his seventh vaccine for SARS-CoV-2.

Unfortunately, we did not investigate anti-SARS-CoV-2 antibody response or neurodegenerative biomarkers like tau and others as they were not available at our institution. His cognitive decline progressed, and he was unable to walk because of ataxia and myoclonic jerks. CSF examination for 14-3-3 protein was negative. His consciousness worsened, and he was transferred to intensive care for follow-up. Control DWI-MRI revealed progression of hyperintensities and brain atrophy. EEG showed 1 Hz periodic sharp waves and probable sporadic CJD was suggested as the final diagnosis (Figure 1). After six months of symptom onset, he was still alive.

Present literature about SARS-CoV-2 infection, SARS-CoV-2 vaccines, and CJD incidence and progression remains debated. There are a few case report series proposing increased progression of neurodegenerative diseases, such as CJD, amyotrophic lateral sclerosis, and Alzheimer's disease, due to SARS-CoV2 infection.² The proposed mechanisms are systemic inflammation and aggregation of proteins caused by a spike protein over the virus.^{2,3} Both the vector and mRNA vaccines express the spike protein. A CJD case was reported with rapid progression to death after vaccination,⁴ but epidemiologic studies showed no increase in CJD incidence or rapid progression of the disease severity during the



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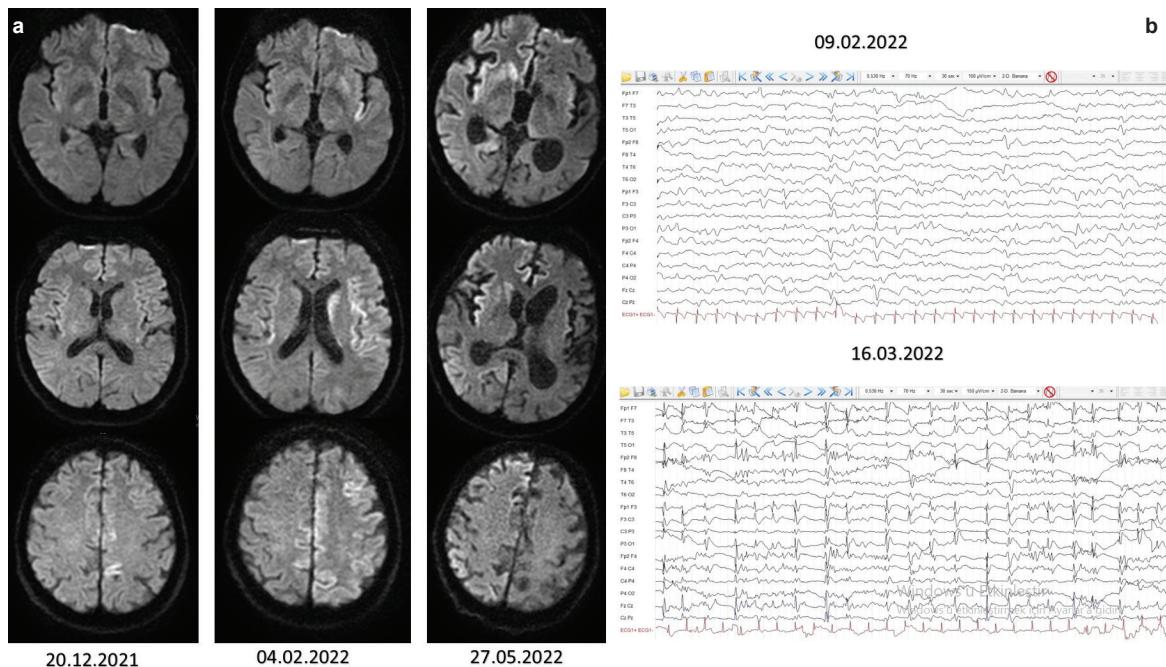


FIG. 1. (a) Progression of cortical and basal ganglionic hyperintensities on diffusion-weighted magnetic resonance imaging over time. (b) Progression of sharp waves to periodic 1 Hz discharges on electroencephalography over time.

coronavirus disease 2019 pandemic.⁵ The overall disease duration after the symptoms was reported to be 5.5 months. Our patient is still alive sixth month after the diagnosis. It is debatable whether this a positive effect for survival after the disease onset caused by the vaccines. CJD should be considered for the differential diagnosis of cognitive decline after SARS-CoV-2 infection or vaccination.

Informed Consent: Written informed consent was obtained from the patient's wife.

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