Clinical Image

PET-CT for the Diagnosis of Creutzfeld-Jakob Disease

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CLINICAL IMAGE

A 68 year-old man with past medical history of right ventricular dysplasia and automatic implantable cardioverter defibrillator (AICD) placement presented with three weeks of cognitive and functional decline. His only prior medications were aspirin, rosuvastatin and tamsulosin. His initial symptoms were intermittent word-finding difficulty, situational confusion, and impaired social interactions. Serum laboratory results, head CT, and CSF studies were unremarkable; unfortunately, an MRI could not be obtained due to his specific AICD model. His initial EEG demonstrated left greater than right slowing, but no epileptic form discharges. He was discharged to home but continued to neurologically decline and was readmitted within one week. Repeat head CT and EEG were unchanged. His affect flattened and he progressed to being unable to recognize family members.

His family requested transfer to our institution. Upon arrival, repeat EEG demonstrated persistent bilateral slowing (left > right) and generalized triphasic waves. Serum and CSF were sent to rule out infectious causes or paraneoplastic processes. The clinical suspicion for Creutzfeld-Jakob Disease (CJD) remained high, although his multiple EEGs were non-confirmatory, MRI was not available, 14-3-3 and T-Tau were pending at this time. The diagnosis of CJD during life can be challenging. Normally, MRI with diffusion-weighted imaging (DWI) and fluid-attenuated inversion recovery (FLAIR) are highly sensitive, specific, and highly indicative of CJD prior the return of laboratory results, characteristic EEG abnormalities, and/or myoclonus [1].

Figure 1 [18F] PET-CT images of hypo metabolism. Head CT demonstrates no gross abnormalities (A, B, C), PET-CT fusion images demonstrate left temporo-parietal (D and E) and subtle left frontal hypo metabolism (F).
Given his AICD, we opted for a fluorodeoxyglucose [\(^{18}\)F] PET-CT, which has demonstrated hypo metabolism of primarily the cortex, with fewer basal ganglia and thalamic changes in the 63 published cases of CJD [2]. His PET-CT demonstrated left frontal and parietal hypo metabolism consistent with CJD (Figure 1) in conjunction with his clinical presentation. In the 24 published patients with both MRI and [\(^{18}\)F] PET-CT, cortical hypo metabolism was noted prior to changes on DWI [3,4]. He rapidly became more agitated and difficult to direct so his family opted for hospice and he expired 3 weeks later. An autopsy was performed and demonstrated spongiform change (Figure 2) and PrP-immuno reactivity within the cortex and cerebellum consistent with spongiform encephalopathy. There were multiple areas of tissue sampled, which were normal (not pictured). The T-Tau level was 888 pg/mL and the 14-3-3 protein was positive on western blot. Western blot analysis of frozen autopsy tissue performed at the National Prion Disease Pathology Surveillance Center in Cleveland, OH, revealed the presence of abnormal protease resistant prion protein (PrPSc), confirming the diagnosis of prion disease.

REFERENCES