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FMRI findings in the final stage of Creutzfeldt-Jakob-disease

C. Florea¹, S.M. Golaszewski², A.F. Unterrainer³, M. Kronbichler⁴, M. Seidl², R. Nardone², S. Weis⁵, F. Gerstenbrand⁶, E. Trinka⁷

¹Salzburg, Austria, ²Department of Neurology, Paracelsus Medical University, Salzburg, Austria, ³Department of Neuroanesthesiology, Paracelsus Medical University, Salzburg, Austria, ⁴Neuroscience Institute, Paris Lodron University and Christian Doppler Clinic Salzburg, Austria, Salzburg, Austria, ⁵Linz, Austria, ⁶Vienna, Austria, ⁷Department of Neurology, Paracelsus Medical University Salzburg, Salzburg, Austria

Background and aims: In the final stages of CJD, the cortical functions are believed to be absent and the patient is in an apallic syndrome (vegetative state, unresponsive wakefulness state-UWS). There were no reports of fMRI performed in patients suffering from CJD.

We report on a 62-year-old male patient with a blunt neurological history who presented the clinical picture of a vegetative state/apallic syndrome in a rapidly progressive disease.

Methods: CCT scan showed enlarged cortical sulci without any further abnormalities. T2w-MRI one week later showed minimal hyperintensive signals in the right thalamus and in the left caudate nucleus. Hypointensities, as a sign of restricted diffusion, of all lobes were detected in DWI MRI. MRS showed a decreased NAA/CHO ratio. 3 EEGs revealed general slowing, a left temporal focus, Rademaker's complexes, and an encephalopathic pattern. Tau-protein and 14-3-3-protein were elevated in CSF. Cortical malperfusion was detected by SPECT in the parietal lobes, but not in temporal lobes. HMPAO- and Ioflupan-SPECT could not proof Parkinson's or Alzheimer's.

Results: To further assess consciousness in the patient, a functional magnetic resonance imaging (fMRI) investigation was performed that demonstrated a clear response within the sensorimotor cortex. Three weeks later the patient died. Pathology confirmed the diagnosis of sporadic Creutzfeldt-Jacob-Disease (sCJD).

Conclusion: In conclusion, in spite of the believed apallic end-stage of CJD and in spite of the induced anaesthesia, our patient showed a BOLD response to vibratory somatosensory stimuli to the hand. Thus, fMRI contributed to precise the patient's diagnosis as not apallic, that would have ethical and clinical implications.

Disclosure: Nothing to disclose

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Prevalence of venous sinus stenosis in pseudotumor cerebri (PTC) using digital subtraction angiography (DSA)

M. Hamdy

Neurology, Ain Shams University, Cairo, Egypt

Objectives: To study the prevalence of intracranial venous stenosis in pseudotumor cerebri patients.

Methods: 30 patients were diagnosed having PTC according to Dandy criteria. All underwent general and neurological assessment. Radiological assessment included CT scan brain ±MRI brain without contrast, MRV. All underwent digital subtraction angiography (DSA) (venous phase) to confirm the validity of filling gaps seen at the level of MRV.

Results: MRV brain showed that 24 patients (80%) showed filling gaps. Digital subtraction cerebral angiography (venous phase) showed 9 patients (30%) had stenosis in their dural sinuses. MRV showed to be a good screening tool since it had 100% sensitivity and negative predictive value. However, since it has a moderate specificity (62%) with a positive predictive value (PPV) of only 35%, then lesions detected should be confirmed with digital subtraction cerebral angiography (venous phase) particularly those involving the transverse and sigmoid sinus.

Conclusion: Studying the intracranial venous system in patients with PTC is an important step in understanding the pathophysiology of the disease. Detection of venous sinus stenosis opens the way to a novel therapeutic option for refractory patients like venous sinus stenting.

Disclosure: Nothing to disclose

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HEAD OFFICE: Breite Gasse 4/7
1070 Vienna, Austria

PHONE: +43 1 889 05 03
FAX: +43 1 889 05 03 13
E-MAIL: headoffice@eaneurology.org
WEB: www.eaneurology.org

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