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A fulminant case of JC virus encephalopathy supporting a novel syndrome associated with JC virus infection of cortical neurons

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Objective

The JC virus (JCV) is well known for causing progressive multifocal leukoencephalopathy (PML), a potentially fatal, demyelinating disease of the brain. PML almost exclusively affects immunosuppressed patients, mainly as a complication in haematological malignancies, lymphoproliferative disorders, HIV-positive patients and patients treated with various immunosuppressive and immunomodulatory drugs for autoimmune disorders. Recently, a new clinical entity, named JCV encephalopathy (JCVE), has been observed. In particular, JCVE mainly affects grey matter, conditioning an encephalitis–like clinical picture.

Methods

We present the case of a 62-year-old male who had a six months history of visual hallucinations and odd-behaviour with personality change, persecutory delusion and anxiety. Past medical history was positive for recurrent prostatitis and a thymoma. On admission, he developed aphasia and a progressive reduction in consciousness. Then, he developed bilateral palpebral and right arm myoclonic jerks. He passed away 3 weeks after onset of symptoms. On admission, magnetic resonance imaging (MRI) of the brain demonstrated a prominent non-enhancing T2, FLAIR and DWI signal abnormality bilaterally involving the grey matter. Laboratory tests showed a slightly increased white blood cell and neutrophil counts. Serological tests were all negative. CSF analysis revealed a normal cell count and normal protein and glucose levels. PCR for neurotropic viruses, Chlamydia and Borrelia were negative. A whole-body CT scan was negative. Several electroencephalograms showed a diffuse anterior theta activity with bilateral parietal epileptic periodic discharges. A second MRI imaging showed a more prominent non-enhancing grey and white matter involvement. Finally, CSF- PCR for JCV was performed and resulted positive.

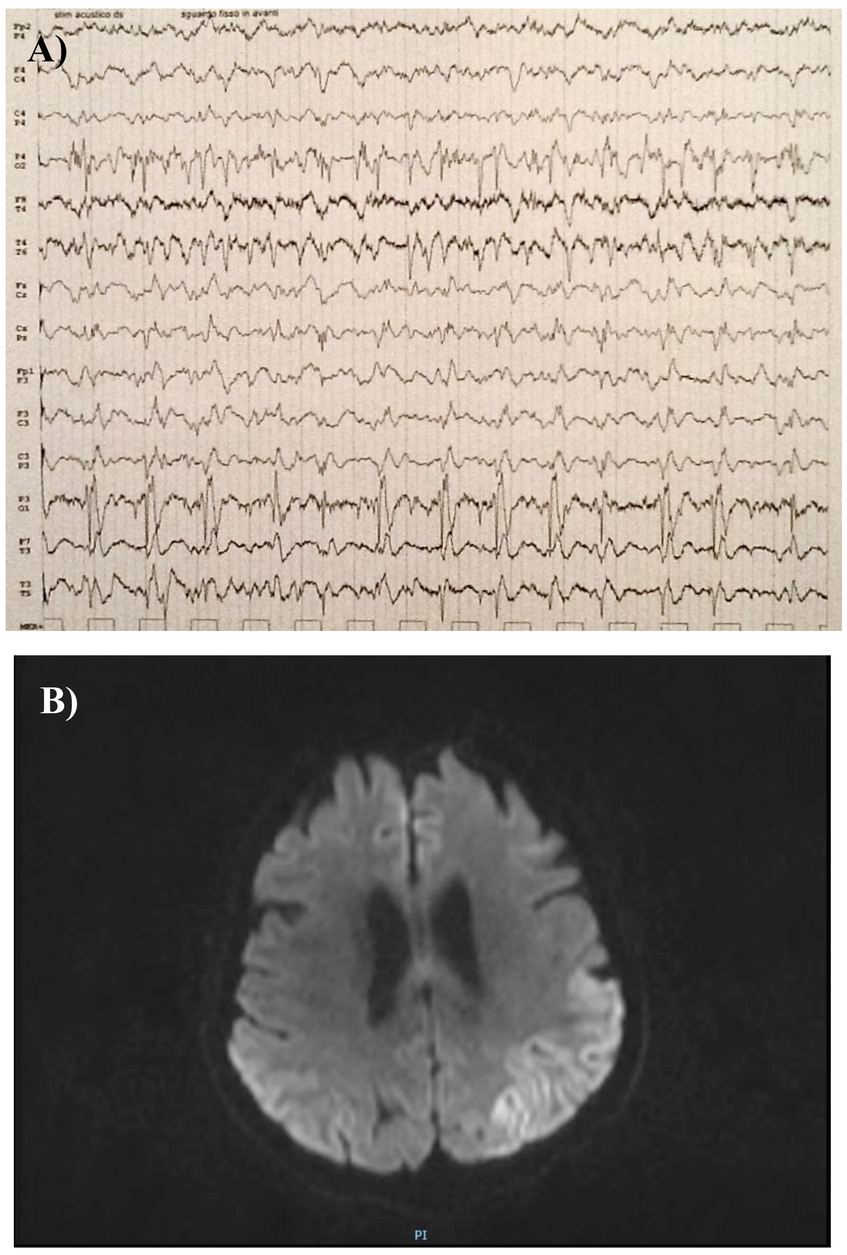

Figure 1.EEG and Brain MRI on admission. In A), EEG showing prominent bilateral parietal epileptic discharges. In B) DWI signal abnormalities bilaterally involving the parietal cortex.

Results

Our patient presented with a subacute onset of psychiatric symptoms, cognitive decline and aphasia. Initial clinical, radiological and neurophysiological findings were consistent with an encephalopathy. Although prion disease was initially suspected, we ruled out the presence of other possible subacute encephalopathies. As the patient did not have clinical and radiological signs compatible with PML, he was not initially tested for JC virus. CSF analysis and post-mortem findings excluded prion disease and confirmed the diagnosis of JCV infection.

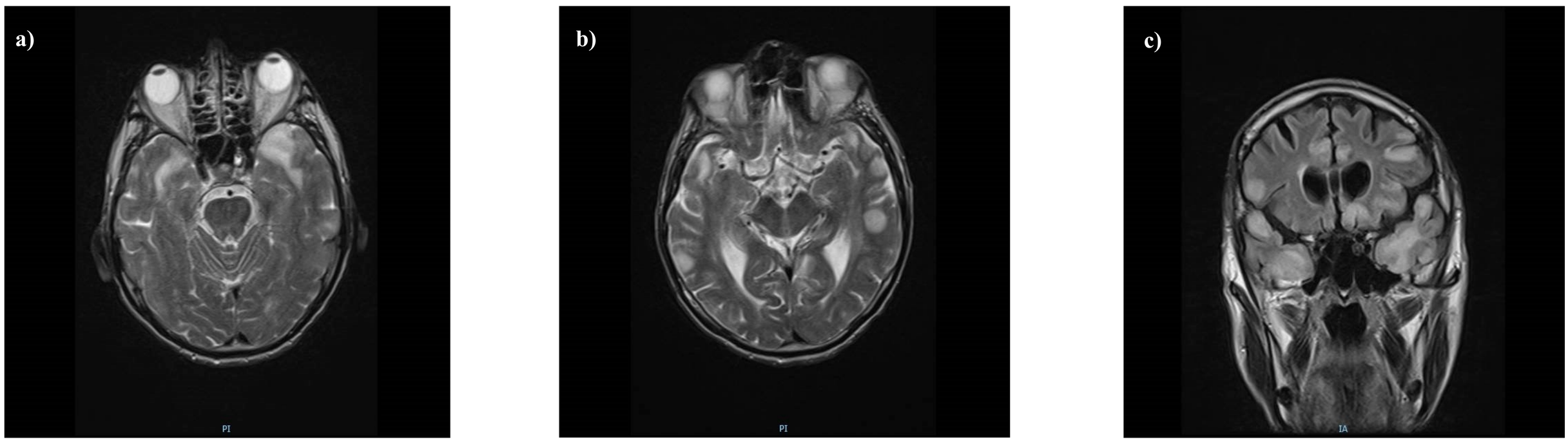

Figure 2. Progression in brain MRI abnormalities. T2 (a,b) and FLAIR (c) MRI imaging showed a more prominent non-enhancing grey and white matter involvement.

Discussion

JCVE is a relatively new and very rare condition, with only one case described so far. Similarly to our patient, the previous case developed symptoms consistent with an encephalopathy. MRI abnormalities were initially restricted to the hemispheric grey matter and only later extended to the subcortical regions. These data suggest that JCV should be considered in evaluation of immunosuppressed patients presenting with unexplained cortical lesions and encephalopathy.

Reference

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