Progressive Multifocal Leucoencephalopathy In A Patient With Chronic Lymphocytic Leukaemia

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Citation

R Abdulqawi, K Ashawesh, P Newman. *Progressive Multifocal Leucoencephalopathy In A Patient With Chronic Lymphocytic Leukaemia*. The Internet Journal of Neurology. 2006 Volume 7 Number 2.

Abstract

Progressive multifocal leucoencephalopathy (PML) is a demyelinating disease caused by the human polyomavirus JC (JCV). We describe a case of PML in a patient with chronic lymphocytic leukaemia.

Progressive multifocal leucoencephalopathy (PML) is a demyelinating disease of the central nervous system, which occurs in immunocompromisd patients. It is caused by a reactivation of the human polyomavirus JC.

CASE REPORT

We report a 67-year-old white female, right-handed, patient who was referred to our outpatient neurology clinic for gradually worsening three-month history of right arm jerky tremor. Her past medical history is significant for 7-year-history of Chronic Lymphocytic Leukaemia (CLL), for which she had received Chlorambucil therapy for 6 years achieving a favourable response until July 2005, when the disease progressed, and standard doses of fludarabine subsequently given. Evaluation of the patient after that showed a favorable response. On neurological examination the patient was alert and fully oriented. Cognitive functions and language were preserved. The optic fundi were normal, and no visual field defects or abnormalities of the cranial nerves were noted. There was remarkable myoclonus affecting the right arm which is worse with action. There were no sensory abnormalities. Abdominal examination was remarkable for splenomegaly.

Electroencephalogram (EEG) did not show any focus of epileptic activity. Cranial magnetic resonance imaging (MRI) revealed hyperintense T2-wighted signals within the motor strip but without mass effect (Figure 1). A lumbar puncture was performed, yielding cerebrospinal fluid (CSF) with glucose level of 2.7 mmol/L, a protein content of 320 mg/L (150-400). CSF cells were within normal limits. CSF polymerase chain reaction (PCR) was positive for JC virus. A diagnoses of PML was made. Treatment options, which are experimental, were discussed with patient who decided against it. Few months later, she deteriorated neurologically and developed hemiparesis of the right side. A repeat cranial MRI revealed a further marked increase in the size of the abnormality with some mild associated mass effect which was not there previously (Figure 2).

The immunosuppression caused by CLL, and possibly fludarabine[1] as well, in our patient predisposed to PML.

Figure 1

Figure 1: T2-weighted MRI brain revealing hyperintense signals (black arrow) within the motor strip.

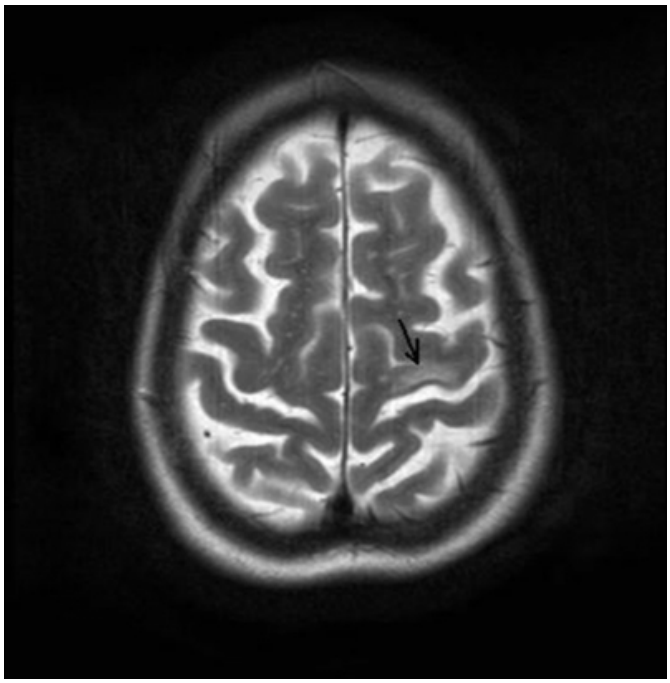
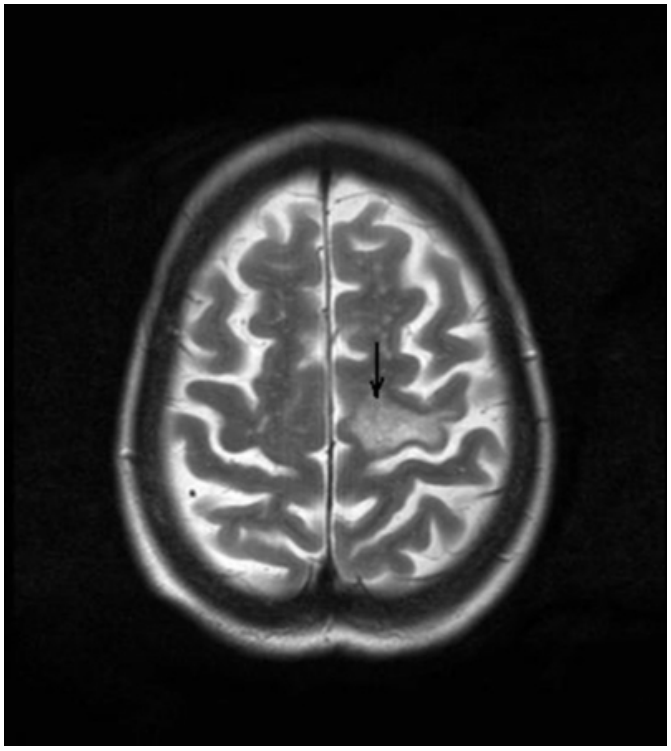


Figure 2

Figure 2: T2-weighted MRI brain revealing marked increase in the size of the hyperintense signals (black arrow) with some mild associated mass effect.



References

1. Kiewe P, Seyfert S, Korper S, Rieger K, Thiel E, Knauf W. Progressive multifocal leukoencephalopathy with detection of JC virus in a patient with chronic lymphocytic leukemia parallel to onset of fludarabine therapy. Leuk Lymphoma. 2003 Oct;44(10):1815-1818.

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