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Case Report

Meningoencephalitis Due to Epstein Barr Virus in An Immunocompetent Man with Atypical Images — Case Report

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Abstract

Epstein-Barr virus (EBV) is a common, asymptomatic, widespread human herpesvirus that affects more than 90% of the population. Severe manifestations are rare and, in immunocompetent adults, are most often self-limited. We report a 79 -year-old immunocompetent, independent man who presented with irregular fever, acute confusional syndrome, generalized tonic-clonic seizures, and decreased caliber and signal intensity in the distal segments of the left middle cerebral artery. Etiologic diagnosis was confirmed based on the positive PCR test for EBV DNA. The patient developed severe meningoencephalitis and multiple organ failure, and eventually passed away after 30 days in the intensive care unit.

Keywords: *Epstein Barr; Meningoencephalitis; Multiorgan failure; Immunocompetent; MRI*

Introduction

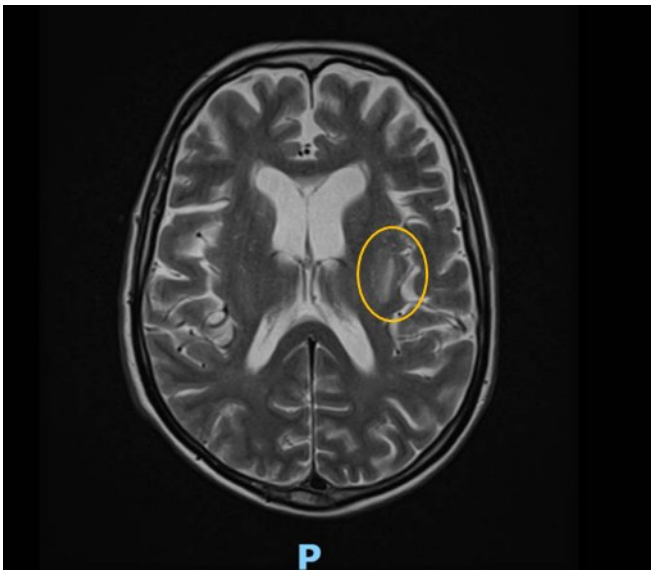
Epstein Barr virus (EBV), a herpesvirus type 4 (HHV-4), is a B-lymphotropic double-stranded DNA virus identified by Epstein Barr in 1964. In the Central Nervous System (CNS), Epstein-Barr virus (EBV) can cause acute encephalitis, cerebellar ataxia, acute disseminated encephalomyelitis (ADEM), myelitis, meningitis, radiculopathy and psychosis among others. [1,2] Humans are the only host and primary infection occurs commonly during childhood (being children more susceptible) with mild symptoms; in adults it often presents with infectious mononucleosis. Dispersed cases of severe EBV complications have been reported in the literature; yet, these cases are extremely rare and mainly affect immunocompromised individuals. Since most infected patients stay asymptomatic throughout their lives, clinical dubitation of EBV in the first place as a causative agent for the severe manifesting picture is of great significance. [3,4] Adults Epstein-Barr virus meningoencephalitis is a serious infectious disorder of the central nervous system, relatively rare in this age range and clinically characterized by fever, headache, and meningeal irritation, and it lacks specificity when compared with other viral encephalitis. There is no current standardized treatment. Images obtained, such as MRI or angio-resonance, during a patient's hospitalization are invaluable tools for diagnosis, treatment, and monitoring. [5,6]

We presented a rare case of Epstein-Barr virus (EBV) meningoencephalitis in an immunocompetent man with decreased caliber and signal intensity of the distal segments of the left middle cerebral artery.

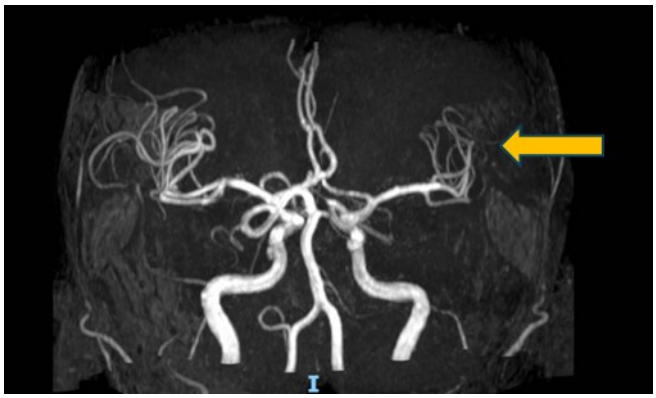
Case Presentation

A 79-year-old male patient was admitted to our institution with a 24-hour history of acute altered language, mental disorder, low-grade fever. As a personal background, he has a coronary stent. According to family was functional and independent, but it started during dinner with hallucinations, eating utensils confusion and strange generalized movements.

Physical examination revealed somnolence and poor response to simple commands; vital signs were: BP 130/80mmHg; heart rate 90 beats/ min; body temperature 37.8°C; respiratory rate 23 breaths/min. Complementary studies: complete blood count, liver and kidney function, electrolyte, blood glucose was normal, HIV/VDRL negative, cpk 9000 U/L. Lumbar puncture showed normal pressure, CSF protein was 21mg/L associated with pleocytosis (30 cells/ mL-lymphocytes dominance) without the appearance of atypical cells and cryptococcus neoformans, with glucose and chlorides tests normal. CSF culture was negative. Whole-body CT scan did not detect any specific abnormalities. CT brain scan not showed abnormal images, MRI 48 hours after informed a sequelae ischemic area observed in the region of the left basal ganglia, to be linked to a history of a long-standing ischemic event and decrease in caliber and signal intensity of the distal segments of the left middle cerebral artery (M3-M4 distally), which have no translation in the DWI and ADC sequences. Because suspected diagnosis of meningoencephalitis patient was treated with acyclovir intravenous (800mg, q 8h for 14 days) and empiric antibiotic therapy. Despite the treatment, his clinical symptoms did not improve and he had to be referred to intensive care unit. Methylprednisolone (1000mg, 3 days) was initiated and needed mechanical respiratory assistance. He suffered several seizures with poor respond to anticonvulsants. Polymerase chain reaction (PCR) multiplex of CSF were all negative except for the EBV-PCR. EBV meningoencephalitis was finally diagnosed. Thirty days after admission, the patient passed away.



A. *Sequelae ischemic area observed in the region of the left basal ganglia.*



B. *Decrease in caliber and signal intensity of the distal segments of the left middle cerebral artery (M3-M4 distally).*

Discussion

Neurological manifestations of Epstein-Barr (EBV) may include meningoencephalitis, brain stem encephalitis, GBS, polyradiculomyelitis, myelitis, nerve palsies, cerebellar ataxia and psychosis. Particularly rare in immunocompetent adults and usually self-limited; the pathogenesis of EBV encephalitis is not clear, may be due to either direct central nervous system (CNS) EBV invasion or a postinfectious inflammatory response mediated by antineuronal antibodies [1,2]. EBV encephalitis is seen in up to 5% of all causes of viral meningoencephalitis [3]. We initially diagnosed the meningoencephalitis based on the clinical findings and confirmed by detecting EBV PCR in the CSF, although some cases could be negative. Approximately 60% of EBV meningoencephalitis contain abnormal CT or MRI findings, with include cortical-subcortical involvement, white matter changes, basal ganglia, and thalamic involvement [4]. In this presentation, CT and MRI were normal, but we found alterations on angio-resonance of middle cerebral left artery. We could not rule out vasculitis due to the impossibility of performing a biopsy because patient's general status and the express request of the family. Most of EBV-specific antiviral therapy is unnecessary in common conditions; however, treatments have not yet been established for EBV-related complications in the nervous system. In this case, steroids and anti-herpes virus agents did not appear to have any significant effects on EBV infection. Ganciclovir and valganciclovir inhibit the replication of EBV in vitro, but have limited efficacy in treating acute infection. Both drugs have been shown to reduce the incidence of EBV-encephalitis or prevent EBV infection; however, these findings have not been consistently replicated. Furthermore, no studies have directly compared these drugs [5].

The main highlights of this rare case are not typical imaging manifestations, an adult patient with normal immune system but not good response to treatment and fatal outcome. Establishing a diagnosis of EBV encephalitis could be difficult due to unusual presenting faces of EBV infections. The prognosis of EBV encephalitis can vary from complete recovery to death. Most patients with EBV encephalitis with normal immune function have good prognosis [6].

Conclusion

We report a very rare case of EBV infection with severe neurological complications causing multiorgan dysfunction in an immunocompetent patient. Due to the presence of normal brain images like those in our case, we suggest performing angio-resonance. This particularly imaging presentation may provide new clinical value allow for the diagnosis of EBV encephalitis. At present, there is no specific treatment and most of patients have a full recovery, but it is crucial to recognize that more high-quality studies are needed to offer an appropriate option.

Conflicts of Interest

The author has no conflict of interest.

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