Case Report

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Presence of Status Epilepticus with Ebstein Barr Virus Encephalitis

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ABSTRACT

Neurological involvement, mostly in the form of meningoencephalitis or encephalitis, represents the leading cause of death in patients with infectious mononucleosis (EF). Central nervous system involvement usually occurs after the first 1 to 3 weeks of disease, although patients presenting with neurological signs and symptoms have been rarely reported. In this case presentation, our aim was to examine the association between acute Ebstein barr virus (EBV) infection and refractory myoclonic convulsions in a patient presenting with myoclonic status.

Keywords: EBV, encephalitis, myoclonic status

Introduction

Ebstein Barr Virus (EBV) infection is a benign systemic viral infection that is usually observed in children. Central nervous system (CNS) involvement of EBV infection is seen with severe neurological symptoms. Meningoencephalitis, cerebrite, guillain barre (GBS), cranial nerve paralysis, transverse myelitis are some of them (1).

Epileptic seizures with EBV encephalitis are rare. These seizures have been described as focal, generalized tonic clonic, or status epilepticus (2).

In this study, a patient characterized with status epilepticus with myoclonic seizures resistant to antiepileptics is described.

Case Presentation

A 17-year-old female patient was admitted to the emergency department of our hospital with status epilepticus. In her history, it was learned that she had her first seizure in generalized tonic-clonic form about 1.5 years ago. At that time, her cranial magnetic resonance imaging (MRI) was normal, electroencephalography (EEG) was not performed, and follow-up was recommended.

One year ago, she was assessed at another center upon another similar seizure and EEG was performed. It was stated that she was started on valproic acid on a total of three seizures during EEG.

It was learned that she had been followed by pediatric neurology unit in Istanbul for the last six months and EEG and MRI had been performed. Cranial MRI revealed no additional findings other than asymmetric dilatation of the left temporal horn to the right.

It was determined that the EEG had been evaluated as compatible with the presence of parietooccipital generalized epileptiform anomaly and the control EEG performed at follow-up had been considered normal. Valproic acid use continued. However, upon the presence of hair loss due to valproic acid use, the subject drug was discontinued two months ago, and levetiracetam was initiated. No generalized seizure has been experienced after levetiracetam.

The patient was treated with intravenous epilepticus diazem in the emergency unit twice. However, since her seizures continued, she was taken to the intensive care unit. The patient with generalized myoclonic seizures was treated with valproic acid. When seizures continued, the seizures were tried to be controlled under general anesthesia. Antiepileptics

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were adjusted and she was given valproic acid, clonezapam, topiramate, levatiracetam respectively. However, treatment was continued with general anesthesia due to resistant seizures.

Since the patient had high fever and infectious diseases department was consulted. 2x2 g Ceftriaxone was started.

Echocardiography revealed 65% ejection fraction and no mass or structure indicating endocarditis in the valves was detected.

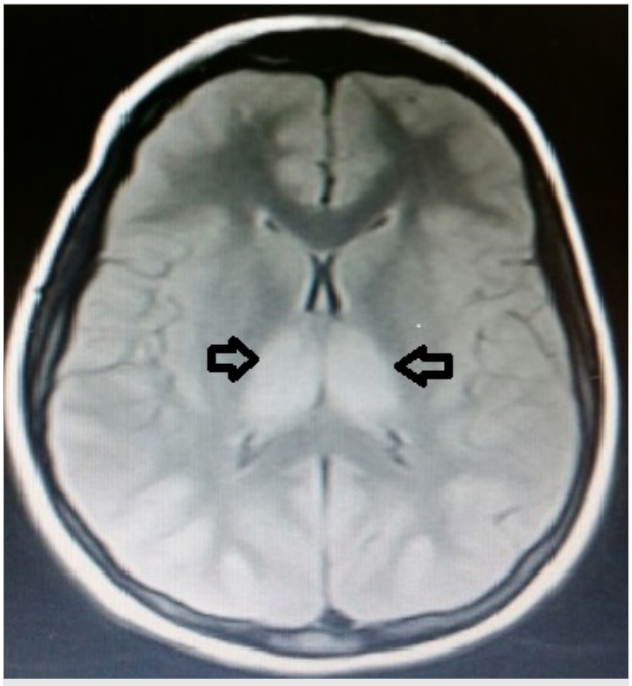

Figure 1. Signal increase in bilateral thalamic region in flair sequence is present during initial magnetic resonance imaging

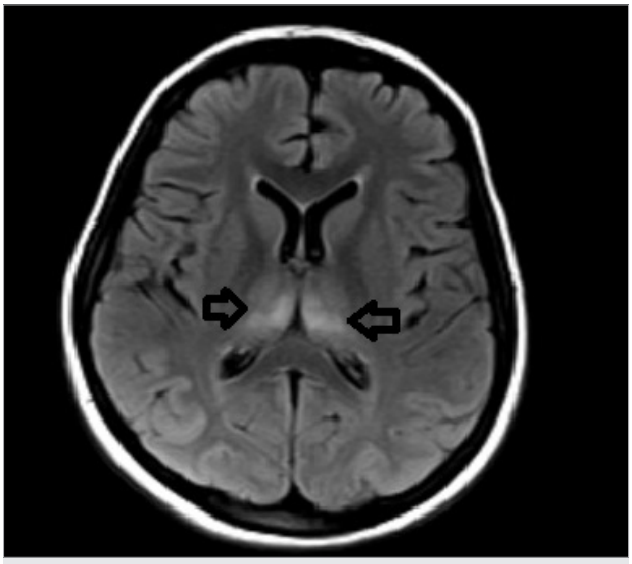

Figure 2. Magnetic resonance imaging showed a decrease in the bilateral thalamic region in flair sequence after 1 month

Cranial computed tomography (CT) was performed. Cranial MRI was performed when CT revealed no clinical explanatory lesion. Cranial MRI performed on the second day revealed hyperintensity in the axial section of the flair sequence in the bilateral thalamic and parietooccipital regions (Figure 1).

Due to leptomeningeal enhancement, meningoencephalitis was considered in the patient. Lumbar puncture (LP) was performed for this purpose. Ten cells were observed. No protein increase was detected. Cytomegolovirus (CMV) and herpes simplex virus (HSV) were sent in CSF for the etiology of encephalitis. The results were negative. It was added to acyclovir treatment.

Autoimmune limbic encephalitis was considered and the steroid was started because of the continuation of high fever, seizures and cognitive impairment in her history. Since the patient was non-responsive to a 10-day pulse steroid treatment, intravenous immunoglobulin (IVIG) was given for 5 days. Glutamate receptor antibody and vasculitis antibodies were negative.

In terms of status epilepticus due to autoimmune causes, and in order to decrease viremia, 5 sessions of plasmapheresis were applied. However, no change was observed in the patient's clinic after plasmapheresis.

Generalized spike and wave activity was observed in EEG. When the patient in the status table was under anesthesia, no suppression pattern was observed in her EEG. Due to cognitive disorder history and myoclonic seizures, subacute sclerosing panencephalitis (SSPE) was considered and LP was performed again. While the first LP had not shown increased protein (22,6), protein was detected as 57,5 in the second LP. Rubeola immunoglobulin g antibody specific index (IGG ASI Index) and oligoclonal band (OCD) were negative in CSF.

The results of hepatitis markers, Human Immunodeficiency Virus (HIV) and John Cunningham (JC) virus were negative.

EBV IGG, IGM; Toxoplasma IGG, IGM; Borrelia IGG, IGM; Brucella were checked in the blood. Several weeks after detecting EBV IGM positive (+), EBV IGG (+) was obtained. One month later the control EBV IGM was negative.

Abdominal ultrasonography and abdominal CT revealed hepatomegaly. Elevated fever supported hepatomegaly and EBV IG M (+) in the blood supported acute EBV infection.

In control cranial MRI, hyperintense involvement was observed to regress in the flair sequence starting from bilateral thalamic region and showing parietooccipital extension (Figure 2).

Discussion

Primary EBV infection often results in infectious mononucleosis. Clinical findings include high fever, sore throat, presence of lymphadenopathy and prodromal symptoms. Prodromal symptoms include sweating, loss of appetite, muscle and joint pain (3).

Neurological involvement during EBV infection is rare (1). The frequency of CNS involvement in EBV infection was reported between 0.37% and 7.3% in published studies (2).

The neurological manifestations seen in the EBV infection process may be meningoencephalitis, acute disseminated encephalomyelitis (ADEM), GBS, bell paralysis, cerebellar ataxia or transverse myelitis (2).

Although CNS involvement usually occurs after 1-3 weeks of the disease, cases that rarely present with neurological findings have been reported. Our patient is also one of the rare cases presenting with encephalitis (1).

Myoclonic epileptic seizures, which are seen as a sign of EBV encephalitis, are rare. Seizures in the encephalitis process are usually defined as focal, generalized tonic clonic, or status epilepticus (2).

The main neurological finding of our case was myoclonic seizures. Her previous EEG was normal, her seizures were generalized tonic-clonic type and under control. Furthermore, in her previous MRIs there was no finding of hyperintense involvement other than asymmetric dilatation of the left temporal horn to the right. Myoclonic seizures were observed with EBV infection, the status table developed and generalized spike wave activity was observed in EEG. In addition to serology positivity, seizure type, EEG findings, observation of MRI response to antiviral therapy, lack of metabolic disorder to provoke seizure showed that this seizure frequency can be associated with EBV.

Acute EBV infection presenting with resistant myoclonic epileptic seizures is limited to case reports in published data. Interestingly, our case was previously followed up with a diagnosis of epilepsy and her seizures were under control. Then she had cognitive deterioration and myoclonic seizures followed by status epilepticus. This suggests that acute EBV infection should be considered in the differential diagnosis of treatment-resistant generalized myoclonic status.

Looking at the laboratory findings, although EBV was not checked in the cerebrospinal fluid, after her hospitalization, her blood EBV was positive and IGG was negative. After about 30 days, when IGG was found to be positive and IGM was negative, it suggested that the patient had evidence of EBV infection.

Neuroimaging studies in EBV encephalitis are normal in most patients. In some publications, signal increases in the basal ganglia and thalamus have been reported in the acute process of the disease. In addition, lesions with corpus callosum splenium have been reported in EBV infection (2,4,5,6). In our case, bilateral thalamic involvement was observed that supports previous publications.

The lesions appear to be reversible in the control cranial MRI. These lesions may be secondary to seizures. As a result, increased cytotoxic edema may cause a signal increase in these areas.

In our case, regression in MRI findings was present, but there was no concurrent improvement in MRI. Although MRI findings were improved with antiviral therapy, seizure control under general anesthesia could not be achieved completely. Therefore, it seems impossible to establish a temporal relationship between MRI lesions and status epilepticus. Based on this; MRI changes that were responsive to antiviral therapy were thought to be associated with EBV infection.

In addition to anticonvulsant therapy; ceftriaxone and acyclovir were given for meningoencephalitis. However, on the continuation of seizures; IVIG and plasmapheresis were performed considering the diagnosis of autoimmune-induced status epilepticus. As a result of these treatments, no change was observed in the patient's clinic. In resistant epilepsies that do not respond to anticonvulsant therapies, autoimmune reasons and encephalitis are considered. In the treatment of status epilepticus due to autoimmune causes, immunomodulators are administered. Steroids, IVIG, plasmapheresis and other immunosuppressive agents are included in the immunomodulatory treatment (7).

Conclusion

This case suggests that acute EBV infection should be kept in mind in the differential diagnosis of patients presenting with myoclonic status epilepticus. In addition, it is stated that bilateral thalamic involvement may be seen in EBV encephalitis and that the prognosis may not always synchronize with reversible MRI lesions.

Informed Consent: Written informed consent was obtained from patients' parents who participated in this case.

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