Acute encephalopathy with biphasic seizures and late reduced diffusion(AESD);2 different clinical outcome

Hale Atalay Celik[1], Zeynelabidin Ozturk[2], Yasemin Tascı Yıldız[3], Rahsan Gocmen[4], Deniz Yüksel[1]

[1] University of Health Sciences Turkey, Dr Sami Ulus Maternity and Child Health and Diseases Research and Training Hospital, Department of Pediatric Neurology

[2] University of Health Sciences Turkey, Dr Sami Ulus Maternity and Child Health and Diseases Research and Training Hospital, Department of Pediatric Intensive Care Unit

[3] University of Health Sciences Turkey, Dr Sami Ulus Maternity and Child Health and Diseases Research and Training Hospital, Department of Radiology

[4] Hacettepe University Faculty of Medicine, Department of Radiology

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Introduction

AESD was described as biphasic seizures on day 1 and days 4-6 accompanied by radiological findings showing no/mild acute abnormality on days 1-2, followed by magnetic resonance imaging (MRI) findings showing reduced diffusion in subcortical white matter on days 3-9. AESD develops in association with systemic as well as central nervous system (CNS) viral or bacterial infections. AESD is most often noted with influenza or human herpesvirus 6 infection as well as bacterial infections in previously healthy infants and in children with developmental retardation. In this study, 2 cases of AESD associated with influenza A virus are presented.

Case report

Case 1: 3 years old girl with unremarkable history, admitted to the hospital with febrile seizure; after 3 days, stupor/coma and generalised tonic seizures were added. The patient was treated for refractory status epilepticus. All laboratory tests including cerebrospinal fluid were normal except influenza A rapid antigen test which was positive. On the 3rd day, cranial computed tomography was normal. On the 4th day there were diffuse cerebral cortical edema, diffusion restriction in cerebral white matter, partial involvement in deep gray matter, but perirolandic region was preserved on the cranial MRI(figure 1). Oseltamivir was started. On the 5th day she was seizure free and on the 15th day she was discharged. In the first month, improvement was observed in the neuroimaging. Now, she is seizure free with one antiseizure medication and developmentally normal.

Case 2: 6 years old boy with language delay and epilepsy admitted to the hospital with febrile status that lasted 30 minutes. The patient was treated for refractory status epilepticus. On the 3rd day MRI was normal, but on 6th day he was still unconscious, so the MRI was repeated. The images(figure 2) was the same with the 1st patient, so we added oseltamivir treatment and performed Influenza A PCR test which was positive. IVIG treatment was given for 5 days. Steroid treatment wasn't given because of the nosocomial infections. He had refractory seizures during hospitalization period and didn't have conscious state anymore The sequela lesions were observed in the follow-up MRIs. Now he has 2 seizures/month with three antiseizure medications and he is at minimally conscious state.

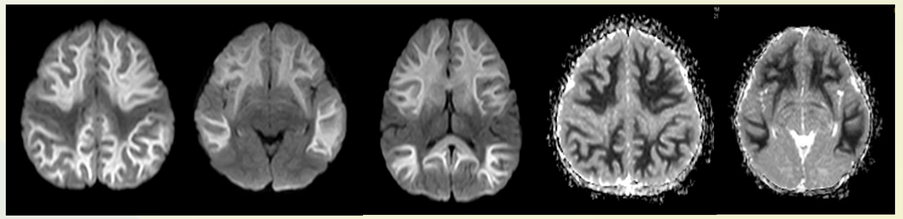

FIGURE 1: There is diffusion restriction in the cerebral white matter. In addition, corpus callosum is involved, but perirolandic and perisylvian regions are spared.

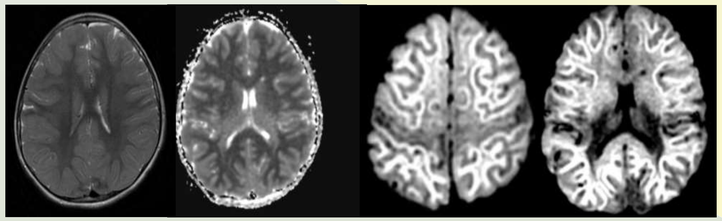

FIGURE 2: There is diffusion restriction in the cerebral white matter extensively, but the perirolandic and the perisylvian regions are spared.

Discussion

Although AESD is usually observed after seizures longer than 30 minutes, it can also be seen after simple febrile seizures. As the same of our cases, Yamaguchi et al. revealed that, patients with AESD who had shorter seizures had better prognosis than those with prolonged seizures.

Diffusion MRI of AESD has diffuse diffusion restiriction at cerebral white matter or centrally spared lesions(bilateral sylvian fissures and perirolandic areas are preserved).

Yadav et al. reported that, patients with diffuse lesions were characterized by coma, and poor neurologic outcome, whereas those with central-sparing lesions were characterized by a biphasic clinical course and relatively mild neurologic sequelae. In contrast, imaging features of our both cases were consistent with central sparing lesions, the clinical outcomes of them were different. First case had a better clinical course than the latter one.

Reduced diffusion in the bilateral hemispheres can be seen with other causes, such as hypoxic-ischemic encephalopathy and shaken infant syndrome. To distinguish between acute encephalopathy and brain injuries due to other causes, not only neuroimaging, but also clinical symptoms, examination and laboratory findings should be considered.

Fukuyi et al. suggested that early administration(within first 24 hours of onset) of vitamin B1, vitamin B6 and L-carnitin would improve the clinical course of acute encephalopathy. We prescribed these vitamins to the patients, but not at the early stages, therefore we couldn't evaluate the effect of vitamin administration, exactly.

Conclusion

AESD should be kept in mind in pediatric patients presenting with signs of acute encephalopathy during an influenza epidemic.

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