CASE REPORT

Pitfall of Steroid Treatment in Acute Demyelinating Encephalomyelitis Coexistent with Chickenpox: A Pediatric Case Report

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ABSTRACT

Primary infection of varicella zoster virus (VZV) causes to chickenpox and easily diagnosed with typical vesicular rashes. It is epidemic in pre-school and school age children since acute infection is very contagious. Infection usually heals spontaneously in healthy children. While post-infectious cerebellar ataxia is the most common complication of the central nervous system (CNS), it leads less likely to encephalitis, encephalomyelitis, meningitis, seizures and demyelinating diseases. Steroid and immune modulator treatments such as intravenous immunoglobulin (IVIG) or plasma exchange (PE) may be administered in the treatment of chickenpox related post-infectious demyelinating diseases. However, there is no clear data whether steroid treatment of demyelinating disease coexistent with chickenpox is suitable or not. In this article, we present a pediatric patient who acute demyelinating encephalomyelitis (ADEM) coexistent with chickenpox and given steroid treatment following IVIG and PE treatments.

Keywords: chickenpox, acute demyelinating encephalomyelitis, children

Suçiçeği ile Birlikte Başlayan Akut Demiyelinizan Ansefalomiyelitte Steroid Tedavi Çelişkisi:Bir Pediatrik Vaka Örneği

ÖZET

Varicella zoster virus (VZV)'un primer enfeksiyonu suçiçeği hastalığına neden olmaktadır ve tipik veziküler döküntüler ile tanı konulmaktadır. Bulaşıcı bir enfeksiyon hastalığı olması dolaysıyla okul öncesi ve okul çağı çocuklarında epidemiktir. Sağlıklı çocuklarda enfeksiyon genellikle kendiliğinden iyileşmektedir. Postenfeksiyoz serebellar ataksi en sık gelişen nörolojik komplikasyondur. Daha az sıklıkta ansefalit, ansefalomiyelit, menenjit, nöbet ve demiyelinizan hastalıklar gibi ağır komplikasyonlar da görülmektedir. Suçiçeği ile ilişkili postenfeksiyoz demiyelinizan hastalıkların tevdisinde steroidler, intravenöz immunglobulin (IVIG) veya plasma değişimi (PE) gibi immun modülator tedaviler uygulanmaktadır. Ancak suçiçeği ile eş zamanlı demiyelinizan bir hastalık geliştiğinde mevcut viral hastalığı ağırlaştırma riski dolaysıyla steroid tedavisinin uygulanabilirliği konusunda tam bir netlik yoktur. Bu yazıda suçiçeği döküntüsü ile eşzamanlı olarak akut demiyelinizan ansefalomiyelit (ADEM) gelişen ve IVIG, PE ve steroid tedavilerine rağmen ağır nörolojik hasar kalan bir pediatrik vaka sunulmuştur.

Anahtar kelimeler: suçiçeği, akut demiyelinizan ansefalomiyelit, çocuk

INTRODUCTION

Primary infection of varicella zoster virus (VZV) causes chickenpox and easily diagnosed when vesicular rashes associated

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with symptoms such as fever and malaise in childhood. Infection in children usually heals spontaneously. It may rarely result in serious complications such as central or peripheral nervous system disorders (1).

Case Report

A six-year-old girl who hadn't any health problems formerly was admitted to the pediatric neurology with gait disturbance. On neurological examination she was conscious, cranial nerves were intact, muscle strength was 4+/5 in upper extremity, 3+/5 in lower extremity. Deep tendon reflexes were normoactive in the upper extremity and hypoactive in the lower extremity and plantar response was flexor. Lower extremity sensorial examination was normal, urinary and anal continence were present. No abnormalities were not detected on hemogram, blood biochemistry, c-reactive protein, serum creatine kinase and lumbar tomography. Sensorial and motor conduction rates were normal, but a slight extension was detected in the F responses. Totally, 2gr/kg intravenous immunoglobuline (IVIG) was given with the preliminary diagnosis of Guillain Barre syndrome (GBS). Very few numbers of itchy lesions appeared on her face and back after IVIG. These lesions turned into vesicular rashes and acyclovir was added to treatment. Central nervous system (CNS) magnetic resonance imaging (MRI) was performed due to the development of high fever, encephalopathy, right gaze palsy and respiratory distress 48 hours after the presence of rashes. The patient was diagnosed with acute demyelinating encephalomyelitis (ADEM) detection of MRI lesions (Figure 1). The lumbar puncture could not be performed because of the rashes in the lumbar region. Respiratory support was provided to the patient being transferred to the pediatric intensive care unit and plasmapheresis was done for 7 days. The patient improved after this treatment and was taken back to the neurology service. A total of 4 weeks of steroid treatment was given, one week of intravenous (IV) high dose. On the CNS MRI repeated one week after the first imaging, T2 hyperintense lesions in the brain and cervical were seen to be reduced, whereas the lesion in the lumbar region was reduced by sequelae change (Figure 2). Electroneuromyography (ENMG) was repeated 2 weeks after and the GBS diagnosis was excluded because of normal conduction rates. Despite these treatments, the patient who developed ambulatory loss and urinary incontinence was discharged after being admitted to the pediatric nephrology follow-up and physical therapy program. After getting permission from the parents, the article writing process was passed.

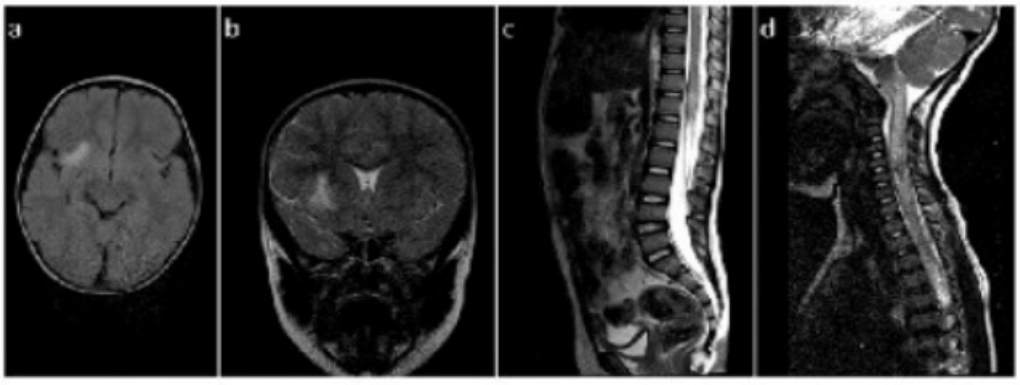

Figure 1. Initially MRIs, T2 hyperintens lesions in right hemisphere insular cortex, cervical and lumbar medulla spinalis.

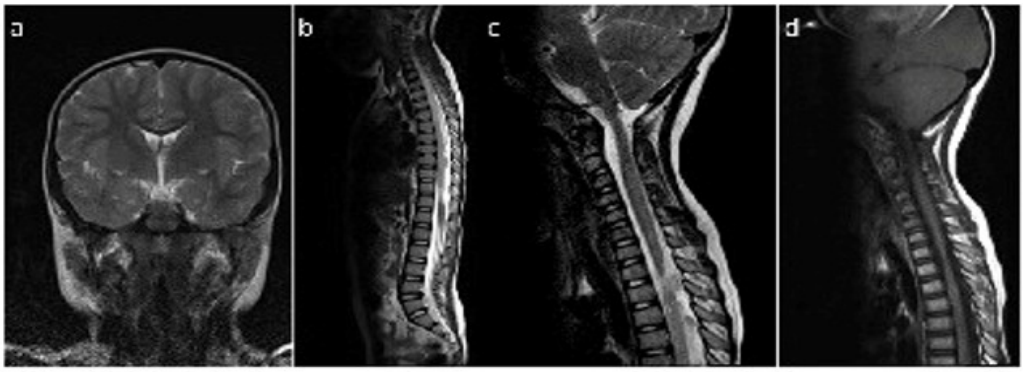

Figure 2. One week later MRIs, T2 hyperintens lesions disappeared in brain and cervical medulla spinalis whereas lumbar remained sequelae.

DISCUSSION

Chickenpox is epidemic in pre-school and school age children because it is highly contagious. Complications of the nervous system are the most serious ones and unfortunately patients have to live with permanent neurological sequelae for a lifelong (2). In this article we present a case who developed diagnostic chickenpox rashes co-occurring with ADEM.

The ratio of neurological complications has been reported to be 8-38% in chickenpox (3). It has been shown that these rates are lowered thanks to national varicella vaccination programs. CNS complications related to chickenpox can be divided into two main groups: stroke and nonstroke. The percentage of nonstroke among VZV-associated neurological complications is 88%. While the majority of nonstroke complications consists of acute cerebellar ataxia, encephalitis, isolated seizures, meningitis, and rarely demyelinating diseases have been reported (4). Demyelinating diseases manifest with clinical symptoms such as clouding of consciousness, seizures, paraparesis/plegia, sensory loss and urinary/anal sphincter dysfunction. Increased protein ratio or pleocytosis may be seen in cerebro spinal fluid analysis. Notably in CNS complications, the rate of VZV DNA detection by polymerase chain reaction is lower in demyelinating disorders compared to meningitis (5). MRI may be normal or T2 hyperintense lesions may be observed. It has been reported that while majority of the cases heal within 2 months, paraparesis or sphincter disorders may be permanent (6). Ours patient presented with acute flask paralysis (AFP) who has not urinary or anal sphincter defect and also conscious, was preliminarily diagnosed with GBS. Even if conduction rates were normal on ENMG, 2 gr/kg IVIG was given by considering the slight extension in ‘’F responses’’ was in favor of GBS acute phase. The patient was diagnosed with chickenpox due to occurrence of typical vesicular rashes on the 6-7th day of the AFP onset. ADEM was diagnosed based on CNS MRI as a result of developing encephalopathy, right gaze palsy and respiratory distress 48 hours after the occurrence of vesicular rash. In immune competent hosts demyelinating diseases following varicella occur either simultaneously or 1-2 weeks after the typical chickenpox rashes (7). A more recent study reported that the preclinical stage in which neurological symptoms occurred but any typical chickenpox rashes has not occurred yet, can extend to till six days before the typical rashes (8). The fact that chickenpox antibodies have not yet developed in this insidious preclinical stage and there is no history of chickenpox contact from the close environment of the patient unfortunately cause the clinician to be caught unawares and delay the initiation of antiviral treatment as in our case.

This patient, who developed ADEM could not be given steroid treatment risk of aggravation of possible CNS infection. In the ADEM treatment protocol high-dose IV steroids are usually administered as the first-

line therapy and reported respond well enough. However, there is no clear data on ADEM concomitant with chickenpox. If the steroid treatment is unsuccessful or is a simultaneous condition with the infection, IVIG or plasmapheresis is recommended as immune modulator treatment options (9). VZV-associated neurological complications have been reported to be more common in patients with few or no vesicular rashes (10). It is clear that this situation delays the initiation of antiviral therapie, thus it will lead to an increased risk of neurological complications. Indeed, our patient also had relatively few vesicular rashes and we did not give any antiviral therapy to during the preclinical stage.

CONCLUSION

We would like to remind that chickenpox should be included in the routine vaccination program particularly in developing countries since it is a preventable disease with a prophylactic treatment method such as vaccination.

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