Case Report

Intravenous Immunoglobulin: A Good Choice for Acute Cerebellar Ataxia Associated with Varicella

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INTRODUCTION
Acute cerebellar ataxia (ACA) is characterized by rapid onset and progression of symptoms, typically developing over a few hours to 1–2 days. But in some cases, the symptoms can be seen a few weeks later. It usually resolves spontaneously. ACA is an important central nervous system disease that occurs most often in children under 6 years of age, often presenting as a postinfectious disorder.[1]

We describe a 5-year-old boy who developed gait disorder and garble speech 2 weeks after chicken pox and who was successfully treated with intravenous immunoglobulin (IVIG).

CASE REPORT
A 5-year-old boy was admitted to the emergency department with gait disorder and slurred speech, which had gradually increased over 2 weeks. Two weeks before, the child had an attack of chicken pox. Physical examination revealed temperature of 37°C, pulse of 86 beats/min, blood pressure level of 100/70 mm Hg, and respiratory rate of 24 breaths/min. No meningeal irritation findings were reported. The cranial nerves were intact, and the muscle tonus was normal. Deep tendon reflexes were normoactive, and no pathologic reflexes were observed. He was unable to sit and walk and had cerebellar dysarthria and ataxia.

The laboratory findings at admission revealed the following: white blood cell count, 7500/mm³ (neutrophil, 48.8%, hemoglobin, 10.8 g/dL, and C-reactive protein, 11.4 mg/L). The serum electrolytes and urine analysis were normal. A cerebrospinal fluid analysis showed normal protein (25 mg/dL) and glucose (86 mg/dL) and lymphocytes (3 cells/mm³). Polymerase chain reaction for varicella zoster virus was negative. Cranial and spinal magnetic resonance imaging (MRI) were initially normal. Electromyography (EMG) was within normal limits. Intravenous acyclovir (30 mg/kg/day) was given for 14 days. The symptoms of cerebellar ataxia did not resolve within 2 weeks. The parents were worried as he could not sit for a month and the patient’s symptoms did not regress during hospitalization. He could not sit and walk still, so we decided to give IVIG.

IVIG (0.4 g/kg) was initiated for 5 days. At follow-up, 5 days later, he was able to walk.

DISCUSSION
ACA is characterized by acute truncal and gait ataxia, variably with appendicular ataxia, nystagmus, dysarthria, and hypotonia. Postinfectious cerebellar ataxia is the most common cause of acute ataxia in childhood. The most frequently associated viral agent is varicella. Epstein–Barr virus, mumps, Legionella pneumophila, Mycoplasma pneumoniae, herpes simplex, influenza, enterovirus, parvovirus B19, rubeola, and

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hepatitis A are the other frequent infectious agents. Drug intoxication, opsoclonus–myoclonus–ataxia syndrome, episodic ataxia, acute cerebellitis, cerebellar stroke, acute disseminated encephalomyelitis, meningitis, cerebral vein thrombosis, Leigh’s disease, Miller–Fisher syndrome, and concussion are the other causes of ataxia.[1,2] Primary infection of varicella, which is characterized by vesicular rash in several stages of maturity, has a benign and self-limiting course. Severe complications include bacterial superinfections such as abscess, cellulitis, necrotizing fascitis, gangrene, and coagulopathies, and central nervous system manifestations can cause morbidity and mortality. ACA and meningocencephalitis are the most common acute neurological complications of chicken pox and are generally followed by complete recovery.[3]

The pathogenic mechanisms that underlie ACA have not been definitively established but are believed to be autoimmune in nature. Antiviral antibodies and autoantibodies against centrosomes, glutamate receptor delta 2, myelin-associated glycoprotein, neurons, and triosephosphate isomerase have been isolated from serum or cerebrospinal fluid of affected patients.[4–6] Radiologic studies of ACA generally are normal; if lesions are seen, they are usually limited to the cerebellum.[4] The diagnosis of ACA is made clinically after excluding other potential causes. Features that help establish the diagnosis are rapid onset of symptoms, history of a prodromal illness during the previous 2–3 weeks, absence of signs or symptoms that might suggest an alternative diagnosis such as fever, meningsmus, seizures, altered mental status, opsoclonus–myoclonus, acquired ocular misalignment, asymmetric neurologic deficits, weakness, sensory loss, abnormal reflexes, unremitting headaches, or recent history of trauma.[7] ACA is considered benign, and its symptoms usually diminish within several weeks. The symptoms remain unchanged in some cases. Oral prednisolone and intravenous methylprednisolone pulse therapy has shown a favorable outcome in some reports.[8] In our case, cerebrospinal fluid analysis, cranial and spinal MRI, and EMG were normal. A history of chicken pox, 2 weeks ago, was reported and he was diagnosed with ACA. His symptoms did not show regression within 2 weeks and he was unable to sit and walk. After the 5th day of IVIG treatment, he was able to walk. IVIG is a treatment option that is used in other causes of ACA and also used in complications of varicella infection.[9,10] In addition, IVIG therapy is worth considering in ACA that does not improve spontaneously.

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**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Nussinovitch M, Prais D, Volovitz B, Shapiro R, Amir J. Post-infectious acute cerebellar ataxia in children. Clin Pediatr (Phila) 2003;42:581-4.
2. Thakkar K, Maricich SM, Alper G. Acute ataxia in childhood: 11-year experience at a major pediatric neurology referral center. J Child Neurol 2016;31:1156-60.
3. Fogel BL. Childhood cerebellar ataxia. J Child Neurol 2012;27:1138-45.
4. Rudloe T, Prabhu SP, Gorman MP, Nigrovic LE, Harper MB, Landschaft A, et al. The yield of neuroimaging in children presenting to the emergency department with acute ataxia in the post-varicella vaccine era. J Child Neurol 2015;30:1333-9.
5. Cohen HA, Ashkenazi A, Nussinovitch M, Amir J, Hart J, Frydman M. Mumps-associated acute cerebellar ataxia. Am J Dis Child 1992;146:930-1.
6. Uchibori A, Sakuta M, Kusunoki S, Chiba A. Autoantibodies in postinfectious acute cerebellar ataxia. Neurology 2005;65:1114-6.
7. Suzuki Y, Kanno A, Minami M, Ogawa K, Oishi M, Kamei S. Case of acute cerebellitis as a result of varicella zoster virus infection without skin manifestations. Geriatr Gerontol Int 2012;12:756-7.
8. Go T. Intravenous immunoglobulin therapy for acute cerebellar ataxia. Acta Paediatr 2003;92:504-6.
9. Daaboul Y, Vern BA, Blend MJ. Brain SPECT imaging and treatment with IVIG in acute post-infectious cerebellar ataxia: case report. Neurol Res 1998;20:85-8.
10. Uysal F, Bostan OM, Cetin B, Uysal B, Guney B, Cil E. Complete atrioventricular block as a complication of varicella infection in a child: recovery with a single dose of intravenous immunoglobulin therapy. Clin Pediatr (Phila) 2016;55:677-9.