Acute Transverse Myelitis Responsive to Steroid Pulse Therapy in a Two-Year-Eight-Month-Old Boy

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(Received for Publication: November 7, 2018)

Abstract
Acute transverse myelitis develops with sudden spinal mobility impairment, symmetric sensory impairment, and rectal or bladder impairment. Although it is commonly treated with steroids, there remains no consistent opinion regarding their effectiveness. We present the case of a two-year, eight-month-old male child in whom steroid pulse therapy was effective. This innately healthy child was brought to hospital with the main complaint of disappearance of spontaneous movements of the bilateral upper extremities. He had developed fever and symptoms of a common cold one month prior to initial examination, at which spontaneous movements of the bilateral upper extremities were absent, and deep tendon reflexes were weak. Spontaneous movements of the lower extremities disappeared the following day, and he developed abasia. Magnetic resonance imaging of the spine showed swelling of the spinal cord from C2 through Th4 levels with high signals on T2-intensified images. Based on the clinical and radiological findings, the child was diagnosed as having transverse myelitis. Steroid pulse therapy was immediately initiated that promptly alleviated the decrease in muscle strength in the lower extremities. After the third course, the condition remitted without sequelae. Early diagnosis and steroid pulse therapy were effective in this young child with acute transverse myelitis.

Key words
Transverse myelitis, steroids, paralysis, spinal cord disease

Introduction
Acute transverse myelitis (ATM) is a disease that causes sudden bilateral spinal dysfunctions in one or more myelomeres. Although some aspects of its mechanism of onset are unknown, many cases have occurred after viral infections or vaccination, and some cases are reported to accompany vascular lesions and autoimmune diseases. Although the onset of ATM may range from infancy to senescence, it rarely occurs in children aged five years or less. Steroids are frequently used in its treatment. However, there is no consistence opinion on their effectiveness. We report the case of a two-year-old male child with ATM in whom steroid pulse therapy was effective, and remission was achieved without any sequelae.

Case
A two-year, eight-month-old male child, with a history of normal growth and development, and no past medical or significant family history, presented with the inability to move both of his upper extremities.

History of current illness: About one month prior to the initial examination at our hospital, the child developed fever and symptoms of a common cold that continued for several days. On the day of the initial examination, he was noticed not to have spontaneous movements in his bilateral upper extremities. By that evening, he could not move either his upper arms or other regions at all. That night, he was brought to our department during an emergency visit.
Physical findings at the visit: His general physical examination findings were as follows. Height: 90 cm; weight: 11.7 kg; body temperature: 37.7°C; blood pressure: 118/68 mmHg; heart rate: 128/min; and SpO₂: 98%. His consciousness was clear, and the color of his face was not pale. No abnormalities were detected on auscultation of the chest or by abdominal manipulation. The examination of his cerebral nervous system was normal, and the child could lift up his left and right shoulders normally.

The child’s posture was normal, and he could stand up. Anti-gravity movements in both the upper extremities were absent, however, and the manual muscle test (MMT) score was 2/6. Only minimal muscle contraction was seen in the hands, and the grasp strength bilaterally was only 1/6. The deep tendon reflex was weak in both upper extremities and increased in both lower extremities. Pathologic reflexes were absent. Abnormal phonation was absent during the collection of blood from both the upper extremities, indicating sensory impairment.

Examination findings at admission: Table 1 shows the results of the examination at admission. The blood tests revealed mild leukocytosis and elevated alkaline phosphatase levels. The common cerebrospinal fluid tests showed no abnormalities. The examination of blood viral antibody titer for Coxsackie B5 by complement fixation showed an increase of 64 times. The spinal myelin basic protein level was elevated at (1260 pg/mL).

Imaging findings: Magnetic resonance imaging (MRI) of the brain done on hospital day 2 showed no abnormal findings.

Peripheral nerve conduction velocity was within the normal range.

Course after admission (Figure 1): On hospital day 3, flaccid paralysis of the lower extremities also appeared. The spinal MRI on this day showed spinal swelling from the C2 through Th4 levels, with high signals on the T2-intensified images (Figure 2). Contrast-enhanced MRI showed no abnormal contrast effect at the same site. Symptoms and radiological findings in this patient met the diagnostic criteria for ATM¹, and steroid pulse therapy (intravenous methylprednisolone 30mg/kg/day for 3 days and, oral prednisone 1 mg/kg/day for 4 days as post-treatment therapy) was administered from day 3 onwards. After the first course of steroid pulse therapy, the paralysis in the lower extremities began to gradually alleviate. After the third course, all abnormal neurological findings had disappeared. The spinal MRI on day 21 revealed improvement in the spinal swelling and high signal intensities on T2 imaging (Figure 3).

The ATM remitted without any sequelae, and the symptoms did not recur after the steroid pulse therapy was terminated.

Discussion

Etiologies of ATM include viral infections and immunoreactions after vaccine inoculation, bacterial and parasitic infections, and inflammatory demyelinating diseases such as disseminated sclerosis and, collagen disease. Most reported cases are idiopathic². However, it cannot be denied that the ATM resulted from viral infection in our patient, because the blood viral antibody titer for Coxsackie B5 examined by complement fixation had increased, and he had a common cold before suffering ATM. We consider the ATM in this patient to be caused by idiopathic or viral infection.

| Table 1. Laboratory Data at admission |
|-------------------------------------|
| **Peripheral Blood:**               |
| White blood cell 11,600 /μl         |
| Differentiation                     |
| Stab 1.5%                           |
| Seg 44.5%                           |
| Lymph 43.0%                         |
| Mono 8.5%                           |
| Eosino 1.5%                         |
| Baso 1.0%                           |
| Red blood cell 541×10⁴/μl           |
| Hemoglobin 13.7 g/dl                |
| Hematocrit 41.1%                    |
| MCV 76.0 fl                         |
| MCH 25.4 pg                         |
| MCHC 33.4%                          |
| Platelet 35.6×10⁴/μl                |
| **Chemistry:**                      |
| TP 7.7 g/dl                         |
| AST 34 IU/l                         |
| ALT 15 IU/l                         |
| LDH 312 IU/l                        |
| ALP 1002 IU/l                       |
| CK 129 IU/l                         |
| Cr 0.29 mg/dl                       |
| BUN 11.7 mg/dl                      |
| Na 143 mEq/l                        |
| K 4.2 mEq/l                         |
| CI 110 mEq/l                        |
| Mg 2.6 mEq/l                        |
| CRP <0.03 mg/dl                     |
| **Virus antibody titer:**           |
| Coxsackie A6 <8 times               |
| Coxsackie A7 <8 times               |
| Coxsackie A9 <8 times               |
| Coxsackie B3 <8 times               |
| Coxsackie B4 <8 times               |
| Coxsackie B5 64 times               |
| **Cerebrospinal fluid:**            |
| Cell count 4/μl                     |
| Lymphocyte 100%                     |
| Neutrophil 0%                       |
| Protein 47 mg/dl                    |
| Glucose 76 mg/dl                    |
| MBP 1260 pg/ml                      |
**Steroid pulse therapy** (10mg/kg/dose 3day 3series)

Prednisone (1mg/kg/day)

acyclovir

Body Temperature

Muscle Strength (MMT)

Figure 1. Clinical course

Figure 2. Spinal MRI before therapy (Day 3)

Figure legend: Spinal MRI showed swelling spinal from C2 through Th4, with high signal on T2WI (Marked by triangles)

Figure 3. Spinal MRI after therapy (Day21)

Figure legend: Spinal MRI revealed improvement in the spinal swelling and high signal on T2WI, compared with day3 imaging.
The presence of symmetrical motor paralysis and sensory impairment in the upper extremities and their absence in the lower extremities on admission was an atypical symptom of ATM in our patient. Therefore, we considered spinal MRI to be necessary for diagnosis. The spinal MRI on hospital day 3 showed spinal swelling and high signal intensities from C2 through Th4 on the T2-intensified images, even though the range of flaccid paralysis was limited to the upper extremities. This suggested that the inflammation existed only in the cervical spinal cord initially, and that it gradually progressed to the lower extremities area of the lateral corticospinal tract on the thoracic spinal cord, which resulted in flaccid paralysis of the lower extremities. Steroids are frequently used in the treatment of ATM. However, there is no consistence opinion on their effectiveness. There is one report of plasma exchange being effective in a case where steroids were ineffective\(^3\). According to the report by Berman et al., which included 62 ATM cases, the occurrence of ATM is rare, with an incidence rate of 1.34 per one million persons, and the age of onset ranged from one and a half to 80 years old\(^4\). Patients between one and nine years old constituted four of the 62 cases. Dunne et al. reported that among 21 ATM patients aged 15 years or younger, only three patients were five years old or less\(^5\). The onset of ATM in our patients was at the age of two years and eight months.

The prognosis of ATM varies greatly between studies. Dunne et al. reported that the predictors of poor prognosis included progression of symptoms from appearance to completion within one day, severe sensory impairment, and the disappearance of deep reflexes\(^5\). In contrast, Miyazawa et al., who investigated 50 patients of ATM in Japan, reported that the cases with poor prognosis had younger onset (30% of whom had sequelae), increased deep tendon reflexes, and positive Babinski reflexes\(^6\). Moreover, Banwell et al. listed early onset as a factor for poor prognosis\(^7\). There were no pathologic reflexes or the disappearance of deep tendon reflex as predictors of poor prognosis in our patient. In contrast, he had several other predictors of poor prognosis, such as increased deep tendon reflex in both lower extremities, younger onset, and progression of symptoms from appearance to completion within one day. Nevertheless, our patient suffered no sequelae.

Although ATM occurred at a young age of onset in our patient, the disease remitted without sequelae. Harisdangkul et al. administered steroid pulse therapy within one week of the appearance of symptoms in cases of ATM complicated with systemic lupus erythematosus and reported a and that favorable prognosis\(^8\). We started steroid pulse therapy on hospital day 3. We believe that this early treatment may have contributed to the improved prognosis. There are no specific findings on blood test for ATM. In contrast, spinal MRI findings can be helpful in the diagnostics evaluation of ATM. For early diagnosis, it is important to perform a spinal MRI at an early stage. Among pediatric patients with paralysis, it is important to formulate an examination plan that also takes the potential of ATM into consideration.

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