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Severe Pure Acute Motor Axonal Neuropathy

CASE REPORT

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ABSTRACT

Acute motor axonal neuropathy (AMAN) is a subtype of Guillain–Barré syndrome. Characteristic electrophysiological features of AMAN are reduced amplitude or absence of muscle action potentials. Our patient described progressive weakness in his leg and was not able to independently walk; he had a feeling weakness in his arms within 24 h after the onset of symptoms. He was diagnosed with AMAN according to the clinical and electrophysiological features. He could independently walk after two years following intensive physical therapy and monitoring. Neurologists and physiatrist should conduct long term monitoring and rehabilitation for patients with AMAN because neurological deficits may persist for a long time.

Keywords: Guillain–Barré syndrome, acute motor axonal neuropathy, rehabilitation

INTRODUCTION

Guillain–Barre syndrome (GBS) is an inflammatory demyelinating polyneuropathy characterized by flaccid paralysis, and it has various clinical and pathological features. It is a classical post-infectious autoimmune disease. GBS is shown to be the most frequent cause of flaccid paralysis following the eradication of poliomyelitis. It affects all age groups, including neonatals, and its incidence is 1.5–2.7 times higher in males. The term GBS is used for classical inflammatory demyelinating forms, which is most frequently seen in daily practice. However, it has been divided into the following two subgroups over time: "acute demyelinating polyneuropathy" and "acute motor axonal neuropathy (AMAN)" (1, 2).

AMAN was first reported to occur in patients having peripheral axonal dysfunction without demyelinating findings in the electroneuromyograph (ENMG) and is seen mostly during summers in the rural areas of China. Some cases were reported from some countries out of Asia, such as Mexico, in spite of the absence of seasonal predisposition. AMAN is characterized by acute motor paralysis, loss of reflexes or hyporeflexia, and peripheral axonal involvement without demyelinating findings in ENMG. In a study the median age of patients with AMAN in China was found to be 10 years (3).

In this study, it was aimed to present an 8-year-old patient with AMAN and who had muscle weakness in the lower extremity at the beginning and then developed tetraplegia as a result of pure motor involvement without sensory involvement in a short time. The patient was treated with intravenous immunoglobulin (IVIG) therapy.

CASE REPORT

An 8-year-old male patient presented to our outpatient clinic with complaints of incapability of walking and moving his hands and arms. The patient who had not experienced such a complaint before complained about loss of strength that had suddenly occurred in his right leg a week ago. He stated that the weakness spread to the other leg and arms within the first 24 h after the onset of the symptoms. He told that he did not have any pain at the onset of symptoms but that he began to feel pain as the weakness progressed. The investigation of his medical history revealed no previous infection, surgical intervention, or vaccination. Moreover, he did not have a history of fever, vomiting, and diarrhea before the illness; no serious pathological history was found in his family and relatives.

The patient was conscious, and he was completely oriented and cooperative. His systemic examination results were normal. The eyes were at the midline and moving to every direction. The patient could count fingers at 6 m. His pupils were isocoric; direct and indirect light reflexes were positive, and the examination of the fundus was normal. No pathological changes were detected on cranial nerve examination.

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While muscle strength was 0-1/5 in all muscles of the upper extremity, it was 0/5 in the lower extremity. Sensory examination was normal; abdominal skin reflex was negative; deep tendon reflex (DTR) was abolic; the plantar reflex was bilaterally responseless. There was no fecal and urinary incontinence.

The results of the patient's routine biochemistry and blood and urine analyses were within normal intervals. The first ENMG was performed one week after his hospitalization. In ENMG performed for four extremities, sensorial conduction velocity was found to be normal, but motor conduction velocity measures and compound muscle action potentials could not be found. Moreover, F latencies could not be recorded; denervation potentials at rest were not detected, and the patient could not voluntarily contract his muscles.

The findings of cervical and cranial magnetic imaging techniques and also of lumbar puncture were normal. No pathology was found in the analysis of cerebrospinal fluid (CSF). No microorganism growth was detected in the CSF culture.

Salmonella, brucella, hepatitis panel, and autoantibodies, which were ordered for investigating possible etiological factors, were negative. Anti-GM1 ganglioside antibodies were not studied for *Campylobacter jejuni*.

Based on protected sensorial conduction, impaired motor conduction, and CSF changes in ENMG, the diagnosis of AMAN was established. The following day after ENMG, IVIG therapy was initiated at the dosage of 400 mg/kg for 5 days. Gabapentin at the dose of 30 mg/kg was given for the patient's pain. Six days after the beginning of the treatment, there was no change in the muscle strength of the lower extremity, but the strength of proximal muscles in the upper extremities progressed to the value of 2-3/5. On the other hand, motor strength in the distal muscle groups was 0-1/5. Deep tendon reflexes were hypoactive in the upper and lower extremities. However, no pathological reflex was detected.

Because the result of the examination conducted two weeks later did not reveal any change, he was discharged from the hospital by recommending outpatient clinic examination and EMG control 25 days after the admission. Gabapentin therapy was continued for the first three months. This treatment was discontinued when his pain stopped. Vitamin B support was recommended to the patient. He continued physical therapy and the rehabilitation program.

The patient's clinical examination and EMG results after six months were as follows: he still could not walk, and the muscle strength progressed to 4/5 in the upper extremities. The motor strength in the lower extremities was 3/5. Atrophy was observed in the intrinsic muscles of the hand and distal muscles of the lower extremities (Figure 1). Although the sensorial examination findings were normal, DTRs were hypoactive in the upper extremities and abolic in the lower extremities. No pathological reflex was found. In his ENMG, the sensorial conduction studies of four extremities were found to be normal, but the motor conduction velocities could not be recorded. In needle ENMG, denervation potentials in the muscles at rest, complex repetitive discharges, thin high-amplitude long-term polyphasic motor unit potentials (MUP) in the proximal muscles, and reinnervation potentials were observed. The patient continued physical therapy and the rehabilitation program regularly.

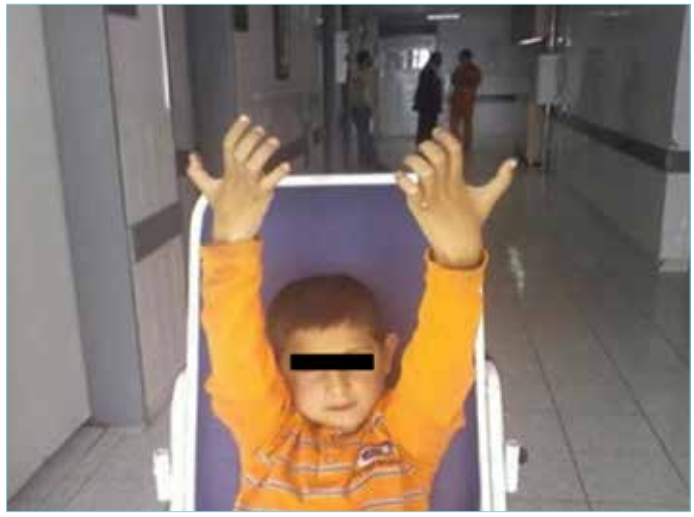

Figure 1. Six months after the treatment of the patient, the muscle strength was 4/5 in the upper extremities but was 1/5 in the distal muscle group of the fingers. Minimal atrophy was observed in the intrinsic muscles. The muscle strength was 3/5 in the lower extremities, and he still could not walk

In the neurological and electrophysiological evaluation performed two years later, the patient could walk independently. The muscle strength was 4+/5 in the proximal and 4/5 in the distal of the upper extremities. It was 4+/5 in the proximal, 4+/5 in the plantar flexion, and 2/5 in the dorsiflexion of the lower extremities (Figure 2, 3). Deep tendon reflexes were normal, and there was no pathological reflex. In ENMG of four extremities, tibial and peroneal nerve motor responses could not be obtained in the left lower extremity. On the other hand, the tibial and peroneal nerve motor amplitudes were low, distal latencies were long, and conduction velocities were normal in the right lower extremity. In the left upper extremity, median, ulnar, and right median nerve motor amplitudes were found to be low, conduction velocities were slow, and distal latencies were normal. In the right upper extremity, ulnar nerve motor amplitude was low; distal latencies and conduction velocities were observed to be normal. The results of the sensorial conduction analyses of the lower and upper extremities were found to be within the normal interval. The needle ENMG revealed long-term thinned polyphasic MUPs with a high amplitude in the muscles. The patient progressively recovered owing to IVIG therapy and regularly performed physical therapy and exercised. Written informed consent was obtained from the patient before preparing the study.

DISCUSSION

AMAN, which is characterized by only the motor axonal involvement of GBS, was first described in China, and then, other cases from all over the world were reported (2, 3).

AMAN is a pathological disorder. Its diagnosis is established through electrophysiological tests. The prevention of conduction in AMAN generally occurs in the first three weeks of the disease, and it is mostly reversible. It was reported that performing serial electrophysiological testing is useful for understanding the pathophysiology of AMAN (4). While the findings of laboratory and CSF evaluations were normal and ENMG examination revealed normal

Figure 2. At the end of two years the muscle strengths were 4+/5 in the proximal and distal extremities and 2/5 in the dorsiflexion of the ankle. The patient could walk independently

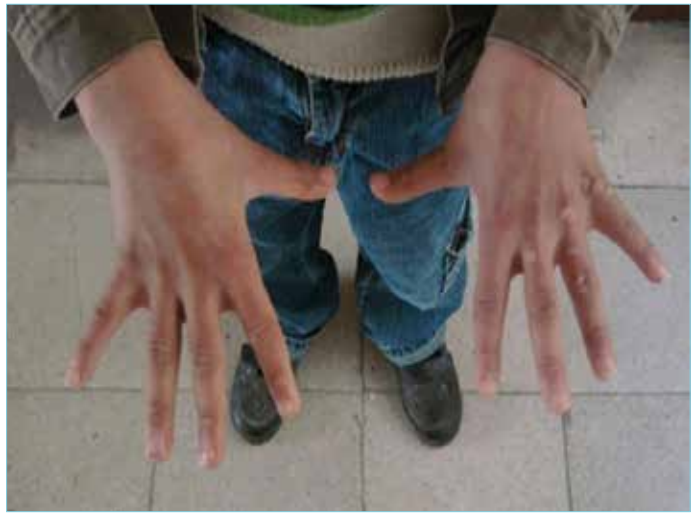

Figure 3. At the end of two years, the muscle strength was 4+/5 in the proximal and 4/5 in the distal of the upper extremities.

sensorial conduction, motor conduction was affected in our case. This condition reminded us AMAN syndrome.

It was detected that there are IgG antibodies against gangliosides GM1, GM1b, and GD1a in the serum of patients with AMAN patients and that these antibodies are generated against membrane liposaccharides of *C. jejuni*. Antibodies bind to nodal axolemma and cause complement activation and macrophage infiltration, thus leading to impaired nerve conductions (2). In our case, we could not examine anti-GM1 ganglioside antibodies for *C. jejuni*. However, other possible infectious causes were ruled out.

Among GBS cases, before the beginning of the symptoms, upper respiratory tract infection comes first and lower gastrointestinal infection follows (5, 6). It was interesting that our patient did not have signs of infection before the symptoms, and the examinations did not reveal a history of previous infection.

In a meta-analysis conducted, it was found that IVIG therapy given for GBS is more effective than plasmapheresis, but this difference is not statistically significant (7). Van der Meche also reported that IVIG therapy is more efficient than plasmapheresis (8). In agreement with literature, IVIG therapy was preferred in this study, and a positive response was obtained from the treatment.

AMAN usually heals without leaving any permanent disability, and the time for recovery can extend up to two years. Therefore, it is important to follow-up patients regularly (9). Nagasawa et al. (9) reported in their study that 80% of patients with AMAN could walk within six months from the beginning of the symptoms, but this period might sometimes be two years. Moreover, Hiraga et al. (3) stated that AMAN patients with a severe clinical picture could not regain their ability to walk in the first six months, and a complete healing could last a longer time. Devos et al. (10) conducted a study on 19 children and reported that 17 of them were able to completely recover after more than a year. In our patient with a severe clinical picture, early healing was not observed. In the first six months, healing was seen in the proximal muscles, but this healing was not apparent in the distal muscles as in the proximal muscles. The patient could not regain his ability to walk independently at the end of the first six months. However, with healing in the distal muscles two years later, the patient started to walk independently.

The ambulation of AMAN patients with a severe clinical picture takes a long time, and the rate of remaining sequel is high. Therefore, physical therapy and rehabilitation programs are as important as efficient medical treatment for the prognosis of the disease. In our patient, the recovery period developed more positively with rehabilitation programs implemented after medical treatment.

CONCLUSION

In patients developing progressive loss of muscle strength, medical treatment should be initiated during the early period considering the diagnoses of GBS and AMAN. Based on the fact that healing can take a long time, long-term practices of physical therapy and rehabilitation programs should be taken into consideration because they can yield satisfying results as in our patient and rehabilitation programs should be continued.

Informed Consent: Written informed consent was not obtained due to retrospective nature of the study.

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Authors' contributions: Conceived and designed the experiments or case: MK. Performed the experiments or case: NT. Analyzed the data: PO, KN. Wrote the paper: PO, KN. All authors have read and approved the final manuscript

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