Elmer Press

Case Report J Neurol Res. 2017;7(1-2):16-18

Nascent Potential: May It Be a Marker in Prediction of Malignant Course in Acute Motor Axonal Neuropathy?

Halil Onder[a, b]

Abstract

Acute motor axonal neuropathy (AMAN) is a subtype of Guillain-Barre syndrome (GBS) that is associated with a longer duration of illness and worse prognosis. Nonetheless, AMAN recovers well according to acute motor-sensory axonal neuropathy (AMSAN) that has been attributed to the underlying mechanisms such as terminal axonal damage as well as ganglioside antibody related injuries at nodes of Ranvier, rather than a primary axonopathy. On the other hand, in malignant coursed AMAN, ventral root degeneration has been shown to be the responsible mechanism for slow and incomplete recovery in these patients. In this report, I present a patient with malignant coursed AMAN who developed a rapidly, severe course of illness and no significant response to therapies (intravenous immunoglobulin and plasmapheresis) could be achieved even on the second month of follow-up. Via the presentation of this case, I draw attention to nascent potentials on electromyography as a potential paraclinical marker for malignant course in AMAN. Future studies investigating the utilizability of nascent potentials in determination of the prognosis of AMAN may provide vulnerable perspectives. In my opinion, these studies may also add crucial data for clarification of the unsolved pathophysiological mechanisms of AMAN.

Keywords: AMAN; Nascent potentials; Electrophysiology; Pathophysiology

Introduction

Acute motor axonal neuropathy (AMAN) is a subtype of Guillain-Barre syndrome (GBS) that is associated with a good prognosis in a considerable number of patients [1]. However, in some AMAN patients, the prognosis can be poorer with incomplete recovery, which has been suggested to be caused by involvement of the nerve roots in the pathophysiological aspect [2]. In our patient presenting with a malignant coursed AMAN, needle electromyography (EMG) study revealed widespread, small and polyphasic motor unit action potentials (MUAPs) compatible with nascent potentials. Via the presentation of this rare patient, we will discuss the vulnerability of the sign of nascent potentials in EMG for prediction of the prognosis of the patients. Furthermore, considering the knowledge that nascent potentials represent true axonal regeneration [3], I will address its importance in enlightening the underlying pathomechanisms in distinct subtypes of AMAN.

Case Report

An 18-year-old female had presented to another medical center with rapid onset respiratory failure and quadriparesis developing in several days following an upper respiratory tract infection (URTI). She had been intubated due to respiratory failure and laboratory investigations (cranial magnetic resonance imaging (MRI) and lumbar puncture (LP)) had not yielded any abnormal findings. Due to the clinical findings and normal imaging results, the pre-diagnosis of GBS was considered. The patient was referred to our center for further investigations on the 15th day of the event. At admission, she was intubated and connected to a mechanical ventilator. On neurological examination, she was conscious and cooperative; cranial nerve examinations were within normal limits. Nonetheless, she was severely quadriparetic (upper extremity proximal muscles grade (MRC) 1/5, distal muscles 2/5, lower extremity proximal muscles grade 2/5, and distal muscles 3-/5) and deep tendon reflexes were absent. Sensorial evaluations were found to be normal. For proper diagnosis, spinal MRI and LP second time were performed which were within normal limits (LP protein: 13 mg/dL, glucose: 81 mg/dL, cytology: non-significant). Electrophysiological studies (EPS) performed on the 20th day of the disease revealed reduction in compound muscle action potentials (CMAPs) in all motor nerve studies, whereas motor nerve velocities and sensorial nerve examinations were found to be normal. Needle EMG investigations showed widespread denervation and reduced interference pattern in all muscles, but prominently in distal extremity muscles (biceps brachii, first dorsal interosseous, vastus medialis, tibialis anterior, and gastrocnemius muscles). Hence, the diagnosis of AMAN was made and intravenous immunoglobulin (IVIG, 0.4 mg/kg/daily for 5 days) therapy was started immediately. Follow-up EPS on the 27th day of the disease showed slight improvements in CMAPs and needle EMG revealed small and polyphasic MUAPs (4 ms) which were compatible with nascent potentials representing the pri-

Manuscript accepted for publication February 17, 2017

[a]Department of Neurology, Hacettepe University Hospital, Ankara, Turkey
[b]Corresponding Author: Halil Onder, Department of Neurology, Yozgat State Hospital, Yozgat, Turkey. Email: halilnder@yahoo.com

doi: https://doi.org/10.14740/jnr418w

mary axonal regeneration. Following 5 days course of daily IVIG treatment, only very slight improvements in muscle strengths could be achieved. Therefore, IVIG was switched to plasmapheresis therapy for every other day and third EMG was planned on follow-up. On 44th day of disease, a mild recovery in muscle strengths was achieved and concurrent EPS showed mild increase in CMAPs and needle EMG revealed widespread polyphasic and small MUAPs (nascent potentials), but decreased interference pattern. On the third month evaluation twice a week, only a further slight improvement was achieved in motor examinations.

Discussion

AMAN is a subtype of GBS that is associated with a longer duration of illness and a worse prognosis. It is characterized by electrophysiological evidence of primary axonal degeneration and decreased CMAPs are reported to be as the early findings of AMAN in EPS which suggests a mechanism of primary axonal pathology [4]. However, a significant number of patients recover well which is incompatible with the clinical course of primary axonopathy [5, 6]. Furthermore, following studies focusing on the pathophysiology of AMAN suggested other mechanisms such as terminal axonal damage [7] as well as ganglioside antibody related injuries at nodes of Ranvier and axolemma [8]. Remarkably, some subgroup of patients with AMAN recover rapidly as patients with acute inflammatory demyelinating polyneuropathy [1], which conflicts with some autopsy studies showing severe, extensive Wallerian-like degeneration in the specific localizations of ventral roots in AMAN [5]. On the other hand, in a distinct report by Ho et al, denervated neuromuscular junctions and reduced fiber numbers in intramuscular nerves in a patient with AMAN were illustrated. In conclusion, the authors suggested that beside antibody related injury at Ranvier nodes, the distal-most motor nerve damage where the blood-brain barrier is deficient, might constitute another key component among the underlying mechanisms of AMAN [7]. In addition, based on the previous autopsy reports, they suggested that previously defined mechanism of ventral root degeneration might be processing as the main responsible factor only in most severe patients, portending a slow and incomplete recovery [7]. In accordance with these hypotheses, Tamura et al (as a result of their study of wide case series) supported the hypothesis of Ho et al [7] that rapid recovery occurred in circumstances of distal terminal axonal damage, whereas axonal loss in the nerve roots should be responsible for poorer and incomplete recovery [2].

In our patient, EMG performed on the 27th day showed small, polyphasic MUAPs which were corresponding to the nascent potentials reflecting the regeneration of proximal axonal bud of damaged region [9]. This suggests an underlying mechanism of prominently primary axonal regeneration rather than a process of terminal collateral sprouting which has been associated with rapid recovery in the better coursed AMAN subtypes particularly in the early phase [2, 7]. Taken together the literature knowledge and presentation of our case, I draw attention to nascent potentials as a possible, utilizable finding in prediction of poor prognosis in AMAN.

Collateral reinnervation has been regarded as a crucial mechanism in the early recovery phase of AMAN and it is responsible for increase of MUAP amplitudes in the recovery period [2] which was absent in our patient. In my opinion, nascent potentials indirectly representing the underlying pathomechanisms can be an underestimated marker which needs to be focused on in future studies addressing pathogenesis of several disorders of peripheral nervous system. On the clinical course, concurrent neurological examinations with EMG studies have not revealed any significant improvement which is in accordance with the knowledge that immature nascent potentials are small and may be incapable of generating a significant force [3] and on the third month follow-up, only very little improvements in muscle strengths were achieved.

In conclusion, I present a malignant coursed AMAN patient who developed a severe and rapidly progressing illness course and no significant response could be achieved in the immunotherapies (IVIG and plasmapheresis) even on the third month of follow-up. Via this case, I suggest nascent potentials as a potential, alternative marker for malignant course in AMAN. This report may also draw attention to the importance of needle EMG as a useful tool for localizing the underlying pathomechanisms in these diseases. However, future EPSs on wider case series should be performed to assess the utilizability of nascent potentials as well as to clarify the underlying pathogeneses of AMAN.

Conflicts of Interest

There are not any conflicts of interest.

Funding

Not any funding has been received.

References

1. Ho TW, Li CY, Cornblath DR, Gao CY, Asbury AK, Griffin JW, McKhann GM. Patterns of recovery in the Guillain-Barre syndromes. Neurology. 1997;48(3):695-700.
2. Tamura N, Kuwabara S, Misawa S, Kanai K, Nakata M, Sawai S, Mori M, et al. Time course of axonal regeneration in acute motor axonal neuropathy. Muscle Nerve. 2007;35(6):793-795.
3. Feinberg J. EMG: myths and facts. HSS J. 2006;2(1):19-21.
4. Ye Y, Zhu D, Wang K, Wu J, Feng J, Ma D, Xing Y, et al. Clinical and electrophysiological features of the 2007 Guillain-Barre syndrome epidemic in northeast China. Muscle Nerve. 2010;42(3):311-314.
5. Griffin JW, Li CY, Ho TW, Xue P, Macko C, Gao CY, Yang C, et al. Guillain-Barre syndrome in northern China. The spectrum of neuropathological changes in clinically defined cases. Brain. 1995;118 (Pt 3):577-595.

6. Hiraga A, Mori M, Ogawara K, Kojima S, Kanesaka T, Misawa S, Hattori T, et al. Recovery patterns and long term prognosis for axonal Guillain-Barre syndrome. J Neurol Neurosurg Psychiatry. 2005;76(5):719-722.
7. Ho TW, Hsieh ST, Nachamkin I, Willison HJ, Sheikh K, Kiehlbauch J, Flanigan K, et al. Motor nerve terminal degeneration provides a potential mechanism for rapid recovery in acute motor axonal neuropathy after Campylobacter infection. Neurology. 1997;48(3):717-724.
8. Kieseier BC, Kiefer R, Gold R, Hemmer B, Willison HJ, Hartung HP. Advances in understanding and treatment of immune-mediated disorders of the peripheral nervous system. Muscle Nerve. 2004;30(2):131-156.
9. GJ H. Neuropathic Needle Examination. AAEM Course A: Fundamentals of EMG, American Association of Electrodiagnostic Medicine. 1984.