〈Case Report〉

Clinical value of abnormal median-normal sural sensory response pattern for the early diagnosis of acute oropharyngeal palsy: a comparison of recent and previous cases

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ABSTRACT To evaluate whether nerve conduction study (NCS) findings are useful to make an early diagnosis acute oropharyngeal palsy (AOP).

We evaluated 2 AOP patients. Patient 1: An 18-year-old male who developed a nasal voice two weeks after common-cold symptoms with no weakness in his extremities. Patient 2: A 16-year-old female who developed a nasal voice two days after diarrhea with no weakness in her extremities. We conducted a routine NCS on these 2 patients and followed up their changes. Both patients had an abnormal median-normal sural (AMNS) sensory response pattern, which improved over time. Both patients were diagnosed with AOP and treated with intravenous immunoglobulin (400 mg/kg/day) with a good prognosis. Anti-GQ1b and/or anti-GT1a IgG antibodies were identified in the serum of both patients a few weeks after their initial diagnoses.

The AMNS sensory response pattern can be useful for the early diagnosis of AOP.

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Key words：Acute oropharyngeal palsy, Guillain-Barré syndrome, Nerve conduction study, Abnormal median normal sural sensory responses pattern

INTRODUCTION

Acute oropharyngeal palsy (AOP) is a variant form of Guillain-Barré syndrome which was first reported on by O'Leary[1] in 1996. In AOP, the main symptom is localized oropharyngeal weakness without limb weakness or areflexia. Unfortunately, some cases may be misdiagnosed as a brainstem stroke[2], and may not be properly treated.

In this study, we evaluated whether nerve conduction study (NCS) findings might be useful for the early diagnosis of AOP.

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CASE REPORTS

Recently diagnosed case

An 18-year-old male developed a nasal voice two weeks after suffering from common-cold symptoms. He had a slight amount of liquid reflux to his nose upon swallowing. He noticed a tingling sensation in his palms and feet the next day and was admitted to our hospital. Neurologically, he had rhinolalia without hoarseness. His soft palate movement was bilaterally decreased and palatal reflex was absent. He had no weakness in his extremities and his deep tendon reflexes (DTRs) were normal. The cerebrospinal fluid analysis was normal (no pleocytosis, protein content was 33 mg/dL).

Summary of a previous case[3)]

A 16-year-old female developed a tingling sensation in her palms and a nasal voice two days after having diarrhea. She had no weakness in her extremities and her DTRs were normal. The cerebrospinal fluid showed an elevated protein level (43 mg/dl) without pleocytosis.

NCS

Both patients received a routine NCS and were followed up with for any changes. We evaluated the median, ulnar and tibial nerves with a motor conduction study (MCS) and the median, ulnar and sural nerves with a sensory conduction study (SCS). The orthodromic technique was used for the median nerve SCS and the lower normal limit of sensory nerve action potential (SNAP) amplitude was 8 μV. The antidromic technique was used for the sural nerve SCS and the lower normal limit of SNAP amplitude was 5 μV.

Results

The MCS results were normal in both patients. In Patient 1, the median nerve SNAP amplitude was 8.4 μV and the sural nerve SNAP amplitude was 18.3 μV on admission. Both amplitudes were within the normal range, but the ratio of median to sural amplitudes suggested an abnormal median-normal sural sensory (AMNS) response pattern. The median nerve SNAP amplitude improved to 25.9 μV on Day 110 (Fig. 1).

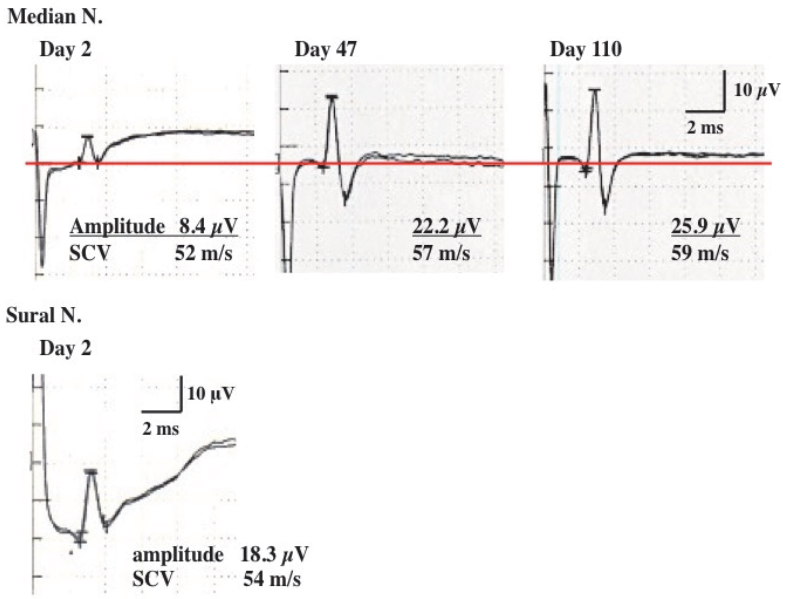

Fig. 1. Sensory nerve conduction study findings in Patient 1. (Upper) SCS in the median nerve on Days 2, 47 and 110. (Lower) SCS in the sural nerve on Day 2.

In Patient 2, the median nerve SNAP amplitude was 10 μV and the sural nerve SNAP amplitude was 22 μV on admission. Both amplitudes were within normal range, but again the median to sural amplitude ratio showed an AMNS sensory response pattern. The median nerve SNAP amplitude improved to 16 μV two months later.

Both patients were diagnosed as having AOP and treated with intravenous immunoglobulin (400 mg/kg/day) on their respective dates of hospital admission, with a good prognosis. Anti-GQ1b and/or anti-GT1a IgG antibodies were identified in the serum for both patients a few weeks after treatment.

DISCUSSION

We showed an AMNS sensory response pattern in two patients with AOP. AMNS pattern has been reported in acute inflammatory demyelinating polyneuropathy and chronic inflammatory demyelinating polyradiculoneuropathy. To our knowledge, this is the first report of AMNS pattern in AOP.

There are previous reports of a total of 14 patients with AOP that received some type of NCS[1, 2, 4–13]. Table 1 showed clinical features of those patients. Eight of those patients (patient 9～16) showed normal findings and six patients (patient 3～8) showed abnormal SCS findings (Table 2). O'Leary[1] mentioned three patients that showed both motor and sensory conduction abnormalities but the exact details were not published. Kamakura[4] reported a case in which sensory nerve conduction velocity (SCV) was decreased by 10% in the sural nerve, and that SNAP of the median and sural nerves was 50-80% lower than the average value based on age. Whether the median or sural nerve was more affected was not mentioned in the report. Ikuta[5] reported that SNAP was not evoked in the sural nerve, while SNAP in the median nerve was normal in an AOP patient with an isolated anti-GM1b IgG

Table 1. Clinical features of patients with AOP

#	Report	Age/sex	Preceding infection	Symptoms at onset	Areflexia	Limb weakness	Ataxia
1	This study	18/M	URTI	Nasal voice	none	−	−
2	This study	16/F	Diarrhea (*C. jenuni*)	Nasal voice	none	−	+
3	O'Leary 1996 [1]	39/F	URTI	Dysarthria, dysphagia	partial	−	−
4	O'Leary 1996 [1]	49/M	Diarrhea (*C. jenuni*)	Dysarthria, dysphagia	complete	−	+
5	O'Leary 1996 [1]	52/M	URTI	Dysarthria, dysphagia	complete	+	−
6	Kamakura 2000 [4]	23/F	Diarrhea, sneezing	Nasal voice, dysphasia	none	−	+
7	Ikuta 2003 [5]	47/F	absent	Nasal voice,	none	−	+
8	Yamaji 2005 [6]	13/M	Diarrhea (*C. jenuni*)	Dysarthria, dysphagia	complete	−	+
9	Onodera 2002 [10]	29/M	Diarrhea (*C. jenuni*)	Nasal voice, dysphasia	none	−	N.M.
10	Makaki 2005 [11]	26/F	URTI	Nasal voice, dysphasia	complete	−	−
11	Kamitani 2006 [12]	46/M	Diarrhea (*C. jenuni*)	Nasal voice,	partial	−	−
12	Wada 2006 [7]	19/F	Diarrhea (*C. jenuni*)	Nasal voice,	partial	−	−
13	Hamidon 2006 [8]	19/F	absent	Nasal voice, dysphasia	complete	+	−
14	Okazaki 2013 [2]	73/M	absent	Nasal voice, dysphasia	partial	−	+
15	Okazaki 2013 [2]	19/M	Diarrhea (*C. jenuni*)	Nasal voice, dysphasia	partial	−	+
16	Nakajima 2013 [13]	40/F	URTI	Numbness, dysarthria	none	−	−
17	Okazaki 2013 [2]	18/M	absent	Nasal voice	partial	−	−
18	Jimura 2017 [9]	16/M	Diarrhea	Nasal voice	None?	−	−

URTI: upper respiratory tract infection, NM: not mentioned

Table 2. IgG anti-ganglioside antibodies, NCS findings and treatment for AOP

#	IgG anti-ganglioside antibodies	SCS findings	Admission Day	Treatment
1	GQ1b, GT1b, GD1a	AMNS pattern	Day 2	IVIg (start on Day 2)
2	GT1a, GQ1b	AMNS pattern	Day 6	IVIg (start on Day 6)
3	GT1a, GQ1b	abnormal		Dietary modification
4	GT1a, GQ1b	abnormal		Dietary modification
5	GT1a, GQ1b	abnormal		Tube feeding, IVIg
6	GT1a, GQ1b, GT1b	MN: amp↓ SN:SCV↓, amp↓	Day 4	PP
7	GM1b	MN:N, SN:not evoked	Day 5	IVIg (start on Day 7)
8	GT1a, GQ1b	SN: SCV↓	Day 5	IVIg (start on Day 10)
9	GT1a, GQ1b	N	Day 15	PP (start on Day 20)
10	GT1a,	N	Day 4	IVIg (start on Day 5)
11	GT1a, GQ1b, GD1a, GD1b, GM1b	N	Day 5	IVIg (start on Day 6)
12	GT1a, GQ1b	N	Day 7	None
13	Not done	N	Day 7	Tube feeding
14	GT1a, GQ1b	N	Day 1	Treated as stroke, IVIg (start on Day 5)
15	GQ1b	N	Outpatient	None
16	GT1a, GQ1b, GT1b, GD1a	N	Day 5	IVIg (start on Day 6?9?)
17	GT1a, GQ1b	N.M		None
18	GT1a, GQ1b	N.D.		None [diagnosed on Day 18]

MN: median nerve, SN: sural nerve, amp: amplitude, IVIg: intravenous immunogloblin, PP: plasma pheresis
N.M.: not mentioned, N.D.: not done

antibody. This patient had no IgG antibodies against GQ1b or GT1a, which may be atypical among AOP patients. Yamaji[6] reported on a patient in which the SCV and amplitude were decreased in the sural nerve.

Among the eight AOP patients with normal NCS findings[2, 7–13], one[2] was first treated as having a brainstem infarction, and some patients[2, 7–9] were not treated with immunotherapy or were treated late[10]. AOP may not be a critical disease but some patients were subjected to tube feeding or needed dietary modification[1], so an early diagnosis is important to start appropriate treatment.

In our study, the absolute values of the median nerve SNAP amplitude were within the normal range and we may have interpreted the NCS as normal if we had overlooked the AMNS pattern. There may be some cases whose NCS showed AMNS pattern in the eight AOP patients with normal NCS findings (patient 9～16). We believe that the use of AMNS pattern may be beneficial in making an early diagnosis of AOP, which we believe is important for effective treatment.

Clinical subtypes of GBS are related to the antigenic specificities of the antibodies[2, 14, 15]. Anti-GQ1b and anti-GT1a have appeared together in AOP, Fisher syndrome, GBS with ophthalmoplegia, acute ophthalmoparesis, the pharyngeal-cervical-brachial variant of GBS, and Bickerstaff's brainstem encephalitis[5]. Several case reports suggest that anti-GT1a antibody, with or without cross-reactivity with GQ1b, is involved in the pathogenesis of oropharyngeal palsy in GBS[2, 5, 14, 16]. Although measurement of serum antiganglioside antibodies should provide useful for supporting the diagnosis of GBS, the assay may take several days to perform. We believe that the use of AMNS pattern may be beneficial in making an early diagnosis of AOP, which we believe is important for effective treatment.

Lastly, reports of GBS in patients with COVID-19 are emerging[17, 18]. Cases of AIDP, AMAN or Fisher syndrome were reported[17]. So, NCS should be considered to perform in patients with COVID-19 who showed acute neurological findings.

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