



Short communication

Autonomic manifestations in acute sensory ataxic neuropathy: A case report



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ABSTRACT

Acute sensory ataxic neuropathy (ASAN) is known to occur with acute and monophasic sensory ataxia. Although autonomic dysfunctions have been reported, no detailed descriptions are currently available. We describe a case of ASAN in which the autonomic manifestations were systematically investigated. Although the patient did not complain of any autonomic symptoms, except for photophobia due to mydriasis, autonomic testing revealed widespread autonomic dysfunctions. Norepinephrine and dobutamine infusion test indicated the presence of sympathetic dysfunction. Additionally, the pupillary response to pilocarpine revealed the presence of parasympathetic dysfunction. In conclusion, widespread, subclinical autonomic dysfunctions may be present in ASAN patients.

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1. Introduction

Acute, monophasic neuropathy with predominant sensory impairment has been described using several names including acute sensory ataxic neuropathy (ASAN) (Dawson et al., 1988; Ito et al., 2011; Rojas-Garcia et al., 2012), acute sensory neuropathy (ASN) (Sterman et al., 1980; Windebank et al., 1990), and sensory variant of Guillain–Barré syndrome (sensory GBS) (Miralles et al., 1992; Oh et al., 2001). Although the definition of the specific clinical features is somewhat different in these neuropathies, they are considered to be in the same spectrum of pathophysiology. ASAN patients show acute sensory ataxia with no or minimal muscle weakness (Dawson et al., 1988; Ito et al., 2011; Rojas-Garcia et al., 2012). The majority of patients with ASN have been known to have sensory ataxia due to a loss of deep kinesthetic sensation without motor deficit (Sterman et al., 1980; Windebank et al., 1990), while a minority of patients manifest superficial sensory loss (Miyazaki et al., 2011). Sensory GBS is characterized by acute sensory ataxia accompanied by slightly abnormal motor nerve conduction velocities (Miralles et al., 1992; Oh et al., 2001). Although autonomic dysfunction has been reported to occur in these types of neuropathies, detailed information is lacking (Sobue et al., 1983; Taly et al., 1991; Ito et al., 2011).

In this report, we describe for the first time the features of autonomic dysfunction in a patient with ASAN.

2. Case report

A 58-year-old female patient experienced a drug eruption after a dental treatment and taking azithromycin. Her drug eruption became worse, and she was admitted to the Department of Dermatology in our hospital at 8 days after onset. On the day after admission, she felt numbness in her neck and face. Her numbness and dysesthesia expanded to her upper and lower extremities, and she subsequently noticed gait disturbance and dysarthria. Because her symptoms did not improve, she was referred to a neurologist 2 days after admission. Eleven years ago, she suffered from acute neuropathy with a monophasic clinical course. Although she was not admitted to our hospital at that time, a review of the medical records suggested that the neuropathy was characterized by predominant impairment of deep sensation and the reduction of sensory nerve action potentials in the median and sural nerves. She recovered from this first episode of neuropathy without disturbance of the activity of daily living.

At the time of the initial medical examination by neurologists in our hospital (second event of neuropathy), she had a height of 152 cm, body weight of 39.6 kg, and blood pressure of 89/64 (systolic/diastolic) mmHg in the supine position. Her resting heart rate was 71 beats per minute (bpm). She was alert and well oriented. A drug eruption was partially crusted on the skin of her extremities. Although her pupils, ocular motion, and facial movement were normal, she had dysarthria. Very mild muscular weakness existed in her hands. The finger-to-nose test and heel-to-knee test identified ataxia in her extremities. Deep tendon reflexes were moderately decreased in her upper and lower extremities. Sensations of pain and light touch were mildly decreased

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in her left hand, while sensations of vibration and position were diminished in her distal limbs. The Romberg test was positive.

Blood cell counts and a urine analysis showed no appreciable abnormalities. Her blood chemistry indicated slightly elevated levels of aspartate aminotransferase (AST) and alanine transaminase creatine (ALT) (AST 38 IU/L, normal 13–33 IU/L; ALT 49 IU/L, normal 6–27 IU/L). A serum autoantibody test showed slightly elevated anti-SS-A antibodies (anti-SS-A antibodies, 29.2, normal, <10.0; enzyme immunoassay). She was negative for anti-SS-B and anti-ganglioside antibodies including GM1, GM2, GM3, GD1a, GD1b, GD3, GT1b, GQ1b, Gal-C, and GT1a. Cranial and spinal magnetic resonance imaging revealed no abnormalities. Neoplastic lesions were not found by whole body fluorodeoxyglucose combined positron emission tomography and computed tomography scan. The cerebrospinal fluid examination performed at admission was normal, but at 1 week after admission, it revealed a protein elevation of 60 mg/dl (normal, <45 mg/dl) with no abnormality in cell count. Nerve conduction studies, performed using a previously described method (Koike et al., 2005), demonstrated normal motor conduction, while sensory nerve action potential was not elicited in the median or ulnar nerve (Table 1).

Intravenous immunoglobulin (IVIg) was administered following the diagnosis of ASAN on the third day of admission. Despite IVIg treatment, she showed a pupil dilation of 6 mm in diameter on both sides, the pupillary light reflex was diminished, and she had no deep tendon reflexes on the fifth day after admission. She became unable to maintain a sitting position because of sensory ataxia due to deep sensory impairment in the trunk. Although her serum sodium level was slightly decreased at admission (serum sodium, 135 mEq/l, normal, 138–146 mEq/l), the syndrome of inappropriate antidiuretic hormone secretion (SIADH), which presents as hyponatremia, low plasma osmolality, and normal renal function with secretion of antidiuretic hormone and cortisol (serum sodium, 108 mEq/l, normal, 138–146 mEq/l; plasma osmolality, 217 mOsm/l, normal, 273–287 mOsm/l; Cr, 0.29 mg/dl, normal, 0.40–0.70 mg/dl; ADH, 10.50 pg/ml, normal, 0.3–3.5 pg/ml; serum cortisol, 14.3 µg/dl, normal, 6.4–21.0 µg/dl), was observed on the sixth day after admission.

There was no obvious clinical symptom of autonomic failure except for photophobia due to dilated pupils. Digestive symptoms, urinary symptoms, orthostatic intolerance, and sicca symptoms were not observed. However, she showed extensive abnormalities in autonomic nerve function tests. The results of the autonomic nerve function tests are shown in Table 2. A head-up tilt test showed a drop in both systolic and diastolic pressure. Her blood pressure changed from 139/103 mmHg to 66/53 mmHg at the tilt angles of 0° and 60°, respectively, without a change of heart rate (72 bpm and 73 bpm, respectively). A norepinephrine infusion test and a dobutamine infusion test using the methods described previously (Nakamura et al., 2011) caused a blood pressure elevation (114/82 mmHg to 157/112 mmHg and 122/87 mmHg to 154/105 mmHg, respectively), indicating the

Table 2
Results of the autonomic nerve function tests.

	This patient	Normal value
Head-up tilt test		
0° sBP/dBP (mmHg)_HR (bpm)	139/103_72	<20 mmHg
60°	66/53_73	Reduction in sBP
NE infusion test		
Pre sBP/dBP_HR	114/82_71	>20 mmHg
Post	157/112_82	Increase in sBP
DOB infusion test		
Pre sBP/dBP_HR	122/87_70	>20 mmHg
Post	154/105_74	Increase in sBP
0.05% pilocarpine eye-drop test		
Pre Pupil diameter (mm)	5.6	−9.5 ± 8.2%
Post	3.8 (−32.1%)	
0.02% dipivefrine eye-drop test		
Pre Pupil diameter	6.3	+3.1 ± 5.8%
Post	5.7 (−9.5%)	
CV _{R-R} (%)	1.94	≥1.41%
123I-MIBG myocardial scintigraphy		
H/M Early	1.82	1.92–2.77
Delay	1.41	1.93–2.55
Washout rate (%)	48.8	–

sBP = systolic blood pressure; dBP = diastolic blood pressure; HR = heart rate; NE = norepinephrine; DOB = dobutamine; CV_{R-R} = coefficient of variation of the R-R intervals

Normal values were based on a previously published reports (Hamada et al., 2003; Nakamura et al., 2011; Yamashita et al., 2010).

presence of vasomotor denervation hypersensitivity due to sympathetic dysfunction (Nakamura et al., 2011). A miotic response was observed with 0.05% pilocarpine eye drops, indicating the presence of denervation hypersensitivity due to pupillary peripheral parasympathetic dysfunction (Yamashita et al., 2010). On the other hand, mydriasis did not result from 0.02% phenylephrine eye drops, indicating the absence of denervation hypersensitivity due to pupillary peripheral sympathetic dysfunction. In an electrocardiogram, her coefficient of variation of the R-R intervals (CVR-R) measured at rest was normal (1.94%; mean, 2.80%; lower limit, 1.41%). Her early and delayed heart-to-mediastinum (H/M) ratio was decreased in 123I-meta-iodobenzylguanidine (123I-MIBG) myocardial scintigraphy [early, 1.82, normal, 1.92–2.77 (Hamada et al., 2003); delay, 1.41, normal, 1.93–2.55 (Hamada et al., 2003)], indicating the presence of cardiac sympathetic dysfunction. A Valsalva test showed lack of heart rate elevation response in phase 2 and lack overshoot of blood pressure in phase 4, indicating the disturbance of parasympathetic nerve pathway in the baroreceptor reflex and sympathetic efferent activity.

As an improvement was not observed after IVIg treatment, pulsed methylprednisolone therapy was performed 1 week later. Her muscular weakness improved over the following 1 month, and her SIADH improved in approximately one and a half months after admission.

Table 1
Nerve conduction study.

		Median nerve					Ulnar nerve					Tibial nerve				Sural nerve	
		Motor			Sensory		Motor			Sensory		Motor			Sensory		
		MCV (m/s)	DL (ms)	CMAP (mV)	SCV (m/s)	SNAP (uV)	MCV (m/s)	DL (ms)	CMAP (mV)	SCV (m/s)	SNAP (uV)	MCV (m/s)	DL (ms)	CMAP (mV)	SCV (m/s)	SNAP (uV)	
Patient	Admission	52	3.1	8.6	NE	NE	53	2.2	7.8	NE	NE	40	4.0	7.2	48	3.9	
	1 month	56	3.0	8.6	NE	NE	59	2.4	7.0	NE	NE	41	4.4	7.5	NE	NE	
Controls		58 ±	3.4 ±	8.2 ±	56 ±	28.0 ±	58 ±	2.6 ±	7.4 ±	54 ±	23.8 ±	46 ±	4.0 ±	11.8 ±	49 ±	16.8 ±	
	(mean ± SD)	4	0.4	2.9	5	11.5	5	0.3	1.8	6	10.3	4	0.6	3.5	5	7.8	

MCV = motor nerve conduction velocity; DL = distal latency; CMAP = compound muscle action potential; SCV = sensory nerve conduction velocity; SNAP = sensory nerve action potential; NE = not elicited.

Control values were based on a previously published report (Koike et al., 2005).

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