A case of autoimmune autonomic ganglionopathy with peripheral neuronopathy

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Introduction

Autoimmune autonomic ganglionopathy (AAG) is an autoimmune disease that involves both sympathetic nerves and parasympathetic nerves [1]. An autoantibody for nerve type Ach receptor (ganglionic Ach receptor: gAChR) which is present in autonomic ganglions is detected in about 50% of cases [2]. Because AAG is a relatively rare disease, the clinical type including the neuropathic merger except autonomic nervous system disorders has many questions. We experienced one case of AAG where the right peroneal nerve palsy appeared during treatment. Because it seemed to be an interesting case to suggest the colorfulness of the clinical type of AAG, it is reported here.

Case report

The patient was a 36-year-old man. In August, 2013, rotatory vertigo at rising, and constipation developed. In the autumn of the same year, photophobia of the right eye, dysuria and impotence developed. Ophthalmology was received in January, 2014. The ophthalmologist found that the pupils were dilated and the opposition areflexia of both eyes was noted. The patient was sent to Tokushima National Hospital for a closer inspection. The skin of the four extremities was dry. A general physical examination showed no abnormalities. The patient was lucid neurologically. In the cranial nerve system, the pupil dilated all right and left of them (diameter 7/7mm). As for the light reflex, the right eye disappeared, and the left eye was dull. The muscle tone was normal, and there was no muscle atrophy or muscle weakness. The deep tendon reflexes were normal, and the Babinski sign was negative. The sensory system and the coordinated movement were normal. In a Schellong study, systolic blood pressure decreased to 70mmHg from 110mmHg, indicating orthostatic hypotension. The blood count and the biochemistry were normal in a laboratory examination. Immunoelectrophoresis did not have abnormal findings. Antinuclear antibody including antiSS-A antibody, antiSS-B antibody, PR3-ANCA, MPO-ANCA, anti-GAD antibody were negative but anti-gAChR antibody was positive. The results of examination of cerebrospinal fluid were: cell count 1/3(mononuclear leukocyte), sugar 59 mg/dl, protein 52.5 mg/dl, IgG 6.1 mg/dl, IgG index 0.48, oligoclonal band negative. There was no cerebellar atrophy in the head MRI of the brainstem. A large quantity of intestinal tract gas accumulated in abdominal simple X-p. CVR-R decreased to 1.28% in the electrocardiogram. In the head-up tilt test, the systolic blood pressure decreased to 63mmHg from 122mmHg. The diastolic blood pressure decreased to 27mmHg from 72mmHg (Figure 1). Because antigAChR antibody was positive, AAG was diagnosed, and immune therapy was provided. Large quantities of immunoglobulins administered by IV therapy and plasma exchange had no effect. A steroid pulse was performed following the plasma exchange. After

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starting prednisolone internal use (60 mg/day), the dizziness on rising disappeared, and the pupillary light reflex of both eyes recovered. Disappearance of the orthostatic hypotension was shown in the Schellong study (Table 1). After large quantities of immunoglobulins were administered by IV therapy, the right foot drop developed without precipitants in particular. However, a sense of pain and a tactile fall appeared in the right culf exterior. Therefore, the right peroneal nerve palsy was suspected. The right foot drop was restored spontaneously.

Discussion

It was reported recently that a group with reversible cognitive functional disorder was present in AAG [3]. The right peroneal nerve palsy of the present case developed after large quantities of immunoglobulins were administered by IV therapy. However, the blood symptoms did not recur after the plasma exchange and the steroid pulse therapy was effective. Therefore, there is a possibility that peripheral neuropathy developed associated with the condition of a patient with AAG. As the mechanism of the bloodstream accommodation, a disorder of the vasa nervorum with the dysautonomia may be involved. There is a merger of autoimmune disease such as Sjogren syndrome or the myasthenia gravis in approximately 20% of AAG cases. It may be said that it is not rare that an autoantibody of antigAChR antibody 以外 develops in AAG. [2,4,5] AAG has been thought to be present only in autonomic nervous system disorders, but the clinical manifestations are varied. This disease may be accompanied with peripheral neuropathy as well as a central nerve disorder. For a diagnosis of AAG and development of treatment, the study of further cases is expected.

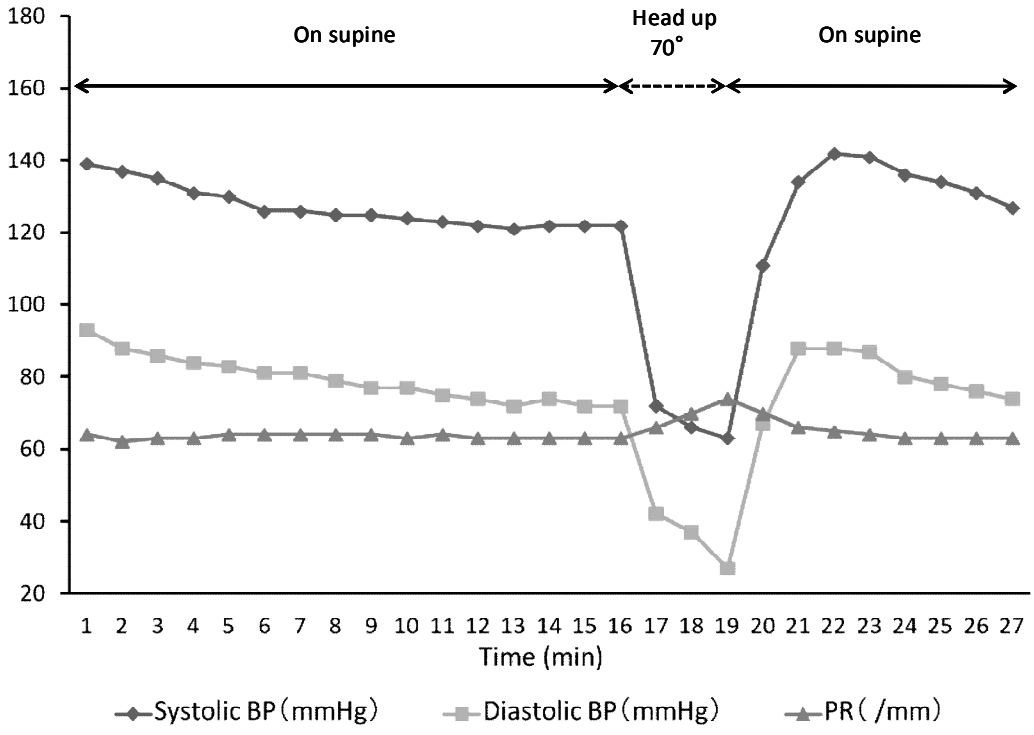

Figure 1. Head-up tilt test.

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a) Before plasma exchange and steroid pulse

	On supine	Standing			
		0 min	1 min	2 min	3 min
Systolic BP (mmHg)	137	90	89	90	91
Diastolic BP (mmHg)	79	49	46	48	50
PR (/min)	65	84	86	86	86

b) After plasma exchange and steroid pulse

	On supine	Standing			
		0 min	1 min	2 min	3 min
Systolic BP (mmHg)	101	91	94	90	91
Diastolic BP (mmHg)	56	48	45	52	52
PR (/min)	71	96	92	91	91

Figure 2. Change in response of blood pressure before and after plasma exchange and steroid pulse therapy.