Acute crisis of hypertension, tachycardia and flush of Amyotrophic lateral sclerosis

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INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is characterized by progressive muscular weakness caused by loss of motor neurons in the precentral gyrus, brainstem and spinal cord. Several reports indicated that the presence of autonomic dysfunction of sweat gland, salivary gland, gastrointestinal tract, urinary sphincter and pupillary muscles in ALS (1, 2). However, the pathophysiological mechanism and possible treatment of autonomic dysfunctions has not been well described in ALS. We present a rare case of ALS with acute crisis of severe hypertension, tachycardia and flush as well as successful treatments.

CASE REPORT

A 68-year-old man was admitted to our hospital because of progressive weakness of the upper extremities for six months. Neurological examination showed dysarthria and atrophy of the tongue with fasciculation. The muscular atrophy and fasciculation were also seen in the upper and lower extremities in which the weakness was moderately present. The tendon reflexes were slightly increased and pathologic reflexes were observed. The results of electromyograms were consistent with ALS. Because the level of \( \text{SpO}_2 \) was from 70 to 90% during the sleep, he was immediately placed under the bi-level positive airway pressure (BiPAP) care. However, he suddenly collapsed and was eventually placed under the mechanical ventilator on the seventh hospital day with tracheostomy. BP had been maintained at the normal range 120-140/60-80 mmHg before and after his admission without any medications. On the 12 \(^{th}\) hospital day, he suddenly developed severe hypertension and tachycardia. BP rapidly increased by 240/120 mmHg and heart rate reached to 150/min. He showed a flush of the face and chest during the attack. Although he complained of severe dyspnea, neither severe hypoxia (\( \text{SpO}_2 \) 98%) nor abnormal bronchial sound was observed during the attack. ECG showed sinus tachycardia and chest X-ray was normal. Those symptoms improved for a couple of hours after the onset by supportive care. Subsequently, BP dropped by 70/30 mmHg with heavily sweating. Using drip infusion of electrolytes, BP returned to normal range for less than one hour. During the attack, the serum level of noradrenalin (NA) was slightly increased (0.62 ng/ml) (normal < 0.05 ng/ml). No adrenal tumors were seen.

Based on the clinical course of the first episode, we attempted to use the diazepam to control the symptoms at the second attack. It was very effective and his clinical symptoms rapidly improved. In order to prevent the repeated episodes of the attack, we started daily diazepam and bunazosin hydrochloride, alpha receptor antagonist, through the nasogastric tube. In the following two months, he showed five times of the hypertensive attack during the daytime or sleep. However, he experienced an improvement of degree of severity of the attacks on each occasion using the both medicines. Eventually the hypertensive attack was completely repressed.
DISCUSSION
To our knowledge, the clinical presentation of the present case is quite rare in ALS individuals. An advanced and respiratory dependent ALS cases may show cardiovascular autonomic dysfunction such as orthostatic hypotension and fluctuation of blood pressure (BP) (1, 3). It was difficult to interpret the mild elevation of NA level during the attack in the present case. Although we could not obtain the permission to carry out a detailed examination of autonomic nervous system, the previous report suggested the presence of alpha-sympathetic hyperactivity in ALS. Furthermore, enhanced sympathetic drive of central origin may be associated with dysfunction of cardiovascular system (4). We believe that alpha receptor antagonist might reduce sympathetic activity and diazepam control central vasocontrol systems in the present case (4, 5).

The autonomic disturbance is one of the important factors in association with sudden death of ALS. Since not only neurologist but also many family physicians have been a chance to see and care for ALS individuals, we have to become aware of the presence of severe autonomic dysfunction of ALS.

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