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"Coat-hanger" 통증으로 발현한 다계통 위축 환자

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A Case of Multiple System Atrophy Presenting with "Coat-Hanger" Pain

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ABSTRACT

Symptoms of cerebral ischemia, such as dizziness, visual disturbance, and fainting, and their relation to the head-up posture are important indicators in the diagnosis of orthostatic hypotension and autonomic failure. Moreover, the additional symptom of neck pain in the suboccipital and paracervical regions, i.e., coat-hanger pain, is often related to a postural change in these patients. However, discomfort caused by coat-hanger pain from orthostatic hypotension in multiple system atrophy has received little attention. We report the case of a woman who presented with coat-hanger pain due to orthostatic hypotension, which markedly impaired her routine activities.

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KEYWORDS Coat-hanger pain, Multiple system atrophy, Orthostatic hypotension

Coat-hanger pain describes the specific neck pain felt in the suboccipital and paracervical regions in patients with orthostatic hypotension due to pure autonomic failure, multiple system atrophy (MSA), and spinal cord injury.[1,2] This pain has been attributed to muscle hypoperfusion/ischemia in the presence of systemic hypotension in the upright position.[3] A previous study showed that the prevalence of coat-hanger pain is ~50% in patients with MSA.[2] However, this has not been described previously in Korea. Here, we report a patient with MSA presenting with coat-hanger pain associated with changes in orthostatic blood pressure.

Case

A 46-year-old woman was referred for evaluation of severe cervical and shoulder discomfort associated with orthostatic dizziness for 5 years. Sometimes, she felt chest pain when standing. The nature of the pain was described as cramp-like, gripping, and squeezing. The symptoms were relieved in the supine position and the relief was sustained unless the precipitating activity,

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such as a head-up postural change, was resumed. Her past medical and family history was unremarkable. She was alert and had a full range of extraocular movements. Hypermetric saccade and bilaterally decreased gain of smooth pursuit were observed. Gaze-evoked nystagmus was observed during leftward gaze. Horizontal head shaking induced downbeat nystagmus. She exhibited scanning speech and bilateral dysmetria in both arms and legs. Truncal sway was evident. The deep tendon reflex was hyperactive. A fundoscopic examination showed a normal posterior pole in both eyes. Other findings of neurological and neurotological examinations were normal. Her pain in the cervical and shoulder areas accompanied by dizziness was evoked by a head-up tilt; the most severe pain was scored when the blood pressure was at the lowermost point, and disappeared in the supine position (Fig. 1). EKG and cardiac enzymes were also normal during chest pain. Review of the previous brain MRI revealed mild diffuse atrophy of the cerebellum (Fig. 2). There was no abnormal finding in cervical-spine MRI. Genetic testing for spinocerebellar ataxia types 1, 2, 3, 6, 7, and 8 was negative. Laboratory evidence led to the exclusion of multiple sclerosis, vitamin E deficiency, paraneoplastic cerebellar degeneration, and viral cerebellitis. On review, the presence of progressive cerebellar ataxia without evidence of a focal or nonfocal symptomatic origin of the disease, severe autonomic dysfunction, and cerebellar atrophy on MRI led us to establish a diagnosis of clinically probable MSA-cerebellar type presenting with coat-hanger pain.[4] She was managed using nonpharmacological measures, such as increased salt intake and elastic stocking application, and pharmacological treatment, including fludrocortisone and midodrine. However, her symptoms were sustained, and she was confined to bed.

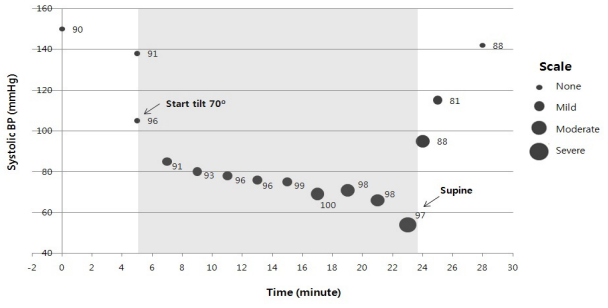

Figure 1. This figure demonstrates the substantial drop in blood pressure (systolic blood pressure from 150 mmHg to 53 mmHg) accompanied by coat-hanger pain and dizziness during the head-up tilt table test (gray column). The discomfort recovers about 2 min later, after return to the supine position, as the blood pressure rise. The size of circle indicates intensity of pain and numbers next to circle mean heart rate. The most severe pain is scored when the blood pressure is at the lowermost point.

Discussion

We described a patient with MSA presenting with pain of the neck and shoulder areas during upright posture due to orthostatic hypotension, in addition to imbalance and dizziness. This type of neck pain in the suboccipital and paracervical regions (coat-hanger configuration) has been reported by patients with autonomic failure and orthostatic hypotension.[1] It differs from other types of neck pain by its manifestation in the standing position.[5] In addition, the neck pain is related to the degree of orthostatic hypotension, as observed in our patient who complained of severest pain at the lowest blood pressure, and is relieved in the supine position.[1] These symptoms are often increased by physical activity and by arm movement alone, such as during hair combing, ironing, dishwashing, and reaching up to perform tasks such as to hang up washed clothes. Eating also reportedly worsens symptoms, presumably through splanchnic vasodilation.[2] In other patients, warm temperature worsens symptoms, probably through cutaneous vasodilation.[2] Although the

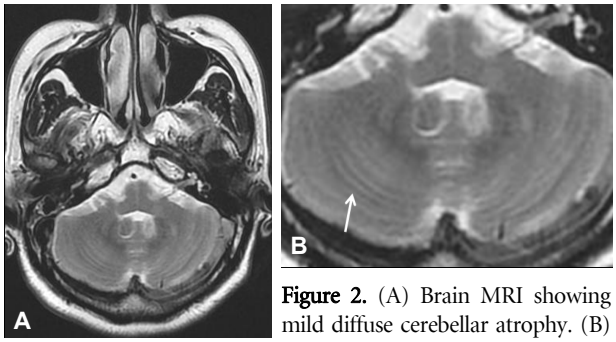

Figure 2. (A) Brain MRI showing mild diffuse cerebellar atrophy. (B) More prominent folia markings (arrow) are noted in the magnified image.

exact mechanisms underlying this phenomenon are not clear, muscle ischemia is the most probable explanation. Namely, the low blood pressure may cause hypoperfusion of the neck muscles. Pain consisting of aching, cramping, or squeezing is similar to ischemic muscle pain.[6] The paracervical muscles need to be tonically active to keep the head upright; during autonomic failure, the combination of underperfusion and such muscle contraction might cause pain. This hypothesis is also supported by the results of recent research based on the velocity recovery cycles of muscle action potentials.[3] Our patient also complained of chest pain. It is known that chest pain may occur in MSA, even in young individuals with normal coronary arteries, and that it may be caused by ischemia of the chest wall. Moreover, some patients complain of lumbosacral and gluteal muscle discomfort, and calf claudication may occur in other patients.[2]

This is the first documentation of coat-hanger pain associated with orthostatic hypotension in a patient with MSA in Korea. The presentation of this case indicates that coat-hanger pain may be the initial manifestation in patients with orthostatic hypotension due to MSA.

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