One case of myotonic dystrophy that developed cerebral infarction

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Abstract

We report on a 53-year-old woman with myotonic dystrophy (MyD) that developed cerebral infarction. This patient had cognitive dysfunction and dysphagia with MyD. Hyperlipidemia and hypotension were found, and this case developed infarction in the anterior cerebral artery region. With MyD, we are careful about hypotension, and it seemed to be necessary to prevent cerebral infarction.

Key Words:

myotonic dystrophy, cerebral infarction, cognitive dysfunction, autonomic nervous system disorders, hypotension

Case Report

The patient was a 53 years old, women. As for the family history, two siblings of mother and mother presented with clinical manifestations of myotonic dystrophy (MyD). There is cholecystolithiasis as a medical history. From about 30 years old, cervical muscle weakness developed. In 42 years, she had a diagnosis of MyD. We started night noninvasive positive pressure ventilation therapy (NPPV) after 45 years old. Dysphagia worsened after 50 years old and was admitted to Tokushima national Hospital. The consciousness of disease was poor (MMSE 29 points, FAB 15 points), and the lower limbs function was a wheelchair level. On April 14, 2016, speech of unknown meaning, memory impairment and balance disorder of the trunk developed from an early morning. The height was 158cm, and the weight was 54 kg (21.6 kg/square meter of BMI). At the onset of cerebral infarction, as for the pulse, 103 a minute was regular. The blood pressure was 140/95mmHg. When she became the locus, blood pressure systolic decreased from 100mmHg to 80mmHg. The both eyes had a cataract, but the fund is normal. We do not hear the cervical bruit, and there is no cardiac murmur. There was disorientation in the consciousness, and long-term memory impairment was found from a short term, but the memory was preserved immediately. There is motor aphasia. There is bilateral blepharoptosis, and there is muscle weakness of the facial muscle, and there is dysphagia. Dysequilibrium of the trunk, cervical significant muscle weakness and grip myotonia were found. As for the limbs muscular strength, proximal part decreased slightness, and distal part decreased moderate degree. The deep tendon reflexes decreased with limbs, and the pathologic reflex was negative. There is no sensory disturbance. The bladder rectal disorder was not found.

As for the laboratory findings, the urinalysis

was normal. WBCs increased by the examination for peripheral blood with 11,000/μl. The blood chemistry presented with mild liver function abnormality. CPK 83IU/l, blood glucose 73 mg/dl, HbA1c 5.3%, HMA-R 0.34%, T-cho 271 mg/dl, TG 266 mg/dl, CRP 2.1 mg/dl, IgG 849 mg/dl, the thyroid function were normal, D-dimer 3.2 μg/ml, Fib 403mg/d, FDPs 4.0 μg/ml. The number of the CTG repeat was 1,100 times by the DMPK genetic screening. There are gallstones and aortic calcification by thoracicoab dominal part CT. BNP was 8.9 pg/ml. There are no abnormal findings to an electrocardiogram and a Holter electrocardiogram. CVR-R was 2.82%. An echocardiography did not have the abnormal findings. In the carotid echo, the plaque was not seen. In the head MRI diffusion weighted image of the next day of the cerebral infarction onset, a high signal was found in a left basal ganglia part mainly on the left head of caudate nucleus, the right head of caudate nucleus and genu of corpus callosum (Figure 1). Both anterior cerebral arteries were not depicted in the head MRA (Figure 2). From April 15 through April 16, there was not the exacerbation of the disturbance of movement of extremities. The blood pressure was 130/95mmHg from 99/78mmHg.

Discussion

MyD merges the organ damage of many systems, including diabetes, hyperlipidemia, arrhythmia, respiratory control disorder, and autonomic nervous system disorders. Intellectual disturbance is found in MyD [1]. The consciousness of disease lacked in this case before the cerebral infarction onset. Because the presenting symptoms of cerebral infarction of this case were cognitive function decreases, we racked our brains about early diagnosis. The responsibility lesion of memory impairment of this case was regarded as bilateral caudate. Elevated blood pressure was not found for an acute phase at cerebral infarction onset, and this case had decreased blood pressure by a changing position to locus. The examination of circadian rhythm measurement of the blood pressure revealed hypotension early in the morning. In MyD and the report of the merger of cerebral infarction, cardiogenic cerebral embolism by the mitral valve prolapse is reported [2]. In addition, a stroke risk factor of MyD includes hyperlipidemia, diabetes, a report of the atrial fibrillation. As a stroke risk factor of this case, hypotension and hyperlipidemia were found. About autonomic nervous system disorders of MyD, hypotension is reported [3]. Orthostatic hypotension is found in 37% of MyD [4]. Anterior cerebral artery region infarction was thought to develop in this case by a thrombotic mechanism or a hemodynamic mechanism as a base for atherosclerosis. When a stroke risk factor was poor in MyD, we were careful about hypotension, and it seemed to be necessary to prevent cerebral infarction.

References

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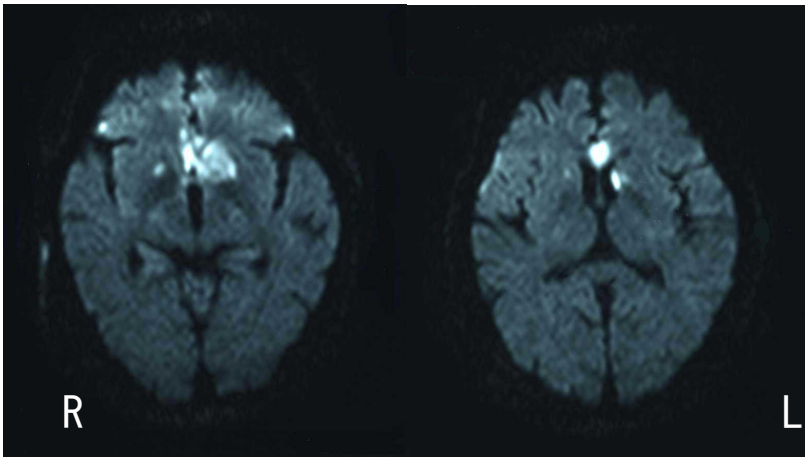

Figure 1:

Head MRI diffusion weighted image (the day after the cerebral infarction onset). A high signal was seen in the left basal ganglia part, mainly on the left head of the caudate nucleus, the right head of the caudate nucleus and the genu of the corpus callosum.

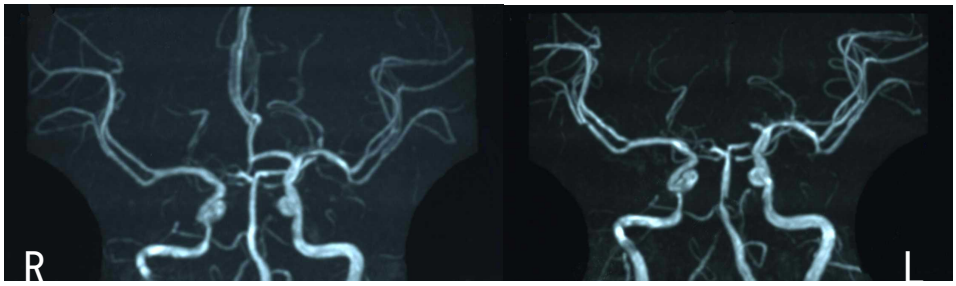

Figure 2:

Head MRA (left: The cerebral infarction pre-critical / right: The day after the cerebral infarction onset). They were after the cerebral infarction onset, and both anterior cerebral arteries are not depicted.