CASE REPORT

Dropped Head Sign as the Only Symptom of Myasthenia Gravis

Hiroaki Yaguchi, Asako Takei, Sanae Honma, Isao Yamashita, Shizuki Doi and Takeshi Hamada

Abstract

Neck extensor muscle weakness and the dropped head sign are associated with various neuromuscular disorders. However, these symptoms are comparatively rare in myasthenia gravis (MG). We report a MG case that presented with dropped head sign as the main symptom. A 55-year-old man developed subacute weakness of the neck extensor muscle and presented with dropped head. We established a diagnosis of MG based on the results of an edrophonium test and a voluntary single fiber electromyogram (vSFEMG), and a high serum antiacetylcholine receptor antibody level. This patient was treated with pyridostigmine and his neurological symptoms improved. There are reported cases of dropped head sign as the first symptom of MG, however, in those cases, other muscles showed weakness during the first few months after onset. In the present case, throughout the clinical course no other symptoms outside of dropped head sign were seen.

Key words: dropped head sign, myasthenia gravis, neck extensor muscles, single fiber electromyogram, edrophonium test

Introduction

The dropped head sign is an occasional symptom in patients with neuromuscular disease and occurs when the extensor muscles of the neck lack sufficient strength to maintain the head in an upright position (1, 2). MG is a disorder of the neuromuscular junction that leads to rapid muscle fatigue, and can be a cause of dropped head. However, the dropped head sign rarely occurs as the first symptom of MG. While there are previous reports of the dropped head sign in patients with MG (2, 3), dropped head sign as the only symptom of MG has not been reported. Here, we describe a patient who presented with dropped head sign as the only symptom, and who was eventually diagnosed as having MG.

Case Report

A 55-year-old man complained of difficulty in holding his head erect for the previous 2 months. He was unable to hold his head upright by the time he was referred to our hospital. Neurological examination revealed that the strength of the neck extensor muscles was decreased to 2/5 of normal, while the patient’s other muscles were normal, including the levator palpebrae superioris and the neck flexors. Dysarthria and dysphasia were not noticed. Weakness of the neck extensor muscles was more severe in the evening than in the morning. Deep tendon reflexes were present and symmetrical. Muscle tonus was normal. A superficial or deep sensory disturbance was not detected. The patient’s cerebellar function and gait were also normal. There was no muscular atrophy and no involuntary movements. His family history and past history were unremarkable. The results of laboratory tests, including creatine kinase, aldolase, myoglobin, sodium, potassium, calcium, and C-reactive protein, were within normal limits. Endocrine and metabolic parameters were also within their reference ranges. Computed tomography (CT) of the neck muscles, magnetic resonance imaging (MRI) of the brain, and cervical MRI were normal. The repetitive nerve stimulation test was recorded over the right abductor digitii minimi muscle, right deltoid muscle, right sternocleidomastoid muscle, right orbicularis oculi muscle, and right orbicularis oris muscle. No waxing or waning of

1 Neurology, Kushiro Rousai Hospital, 2 Neurology, Hokuyukai Neurology Hospital and 3 Minami-ku Neurology, Sapporo Minami Hospital

Received for publication August 6, 2006; Accepted for publication October 30, 2006

Correspondence to Dr. Hiroaki Yaguchi, yaguchi@xa2.so-net.ne.jp

DOI: 10.2169/internalmedicine.46.6167
Figure 1. Edrophonium test results of our patient. (A) Dropped head sign. (B) Edrophonium improves weakness of the neck extensor muscles.

the compound muscle action potential (CMAP) was found at any of these sites. Although no abnormality of the repetitive nerve stimulation test was found, an edrophonium test was performed with intravenous edrophonium. Within 60 seconds of edrophonium administration, the strength of the patient’s neck extensors returned to 5/5 (Fig. 1). Since the edrophonium test was considered to be positive, we examined the patient’s serum antiacetylcholine receptor antibody level, and found that the levels were elevated (1.9 nmol/l; normal range 0.0-0.2 nmol/l). Further, we performed a vSFEMG study of the right extensor carpi radialis muscle in order to establish a diagnosis. The mean consecutive difference (MCD) was 149 ms, which was increased, and blocking was also seen. Therefore a diagnosis of MG was established. There was no evidence of a thymoma on chest MRI.

Since his symptoms remained mild and confined to the neck extensors, the patient was started on oral pyridostigmine bromide (120 mg/day). He tolerated this regimen well, and returned to his normal daily activities. At present, the patient is taking the same dose of oral pyridostigmine bromide and is able to maintain daily life activities by himself.

Discussion

Neck extensor muscle weakness and the dropped head sign are associated with various neuromuscular disorders (1, 2). In the present patient, the dropped head sign was the only presenting feature, therefore, we considered all of the neuromuscular diseases as possible diagnoses. Although the repetitive stimulation test and chest MRI were negative, the positive edrophonium test, the slightly increased serum levels of anti-acetylcholine receptor antibody, and the results of the vSFEMG study indicated a diagnosis of MG.

MG is known to affect the neck muscles, leading to difficulty in maintaining an upright head position. However, MG more commonly affects the flexor muscles of the neck (3-5). In most cases, other muscles are involved as weakness of the neck extensors becomes apparent. Two previous studies have reported dropped head as the first symptom of MG (2, 3). In one case, other muscles were found to be involved when the patient was examined due to weakness of his legs and difficulty rising from a chair. In the other patient, limb-girdle and facial muscle weakness with ptosis developed over 2 months. Thus, in those two cases, the symptoms became worse and systemic muscle involvement was seen within a few months after the onset. MG was diagnosed only after systemic muscle involvement became apparent (2, 3). The present case is unique in that dropped head sign was the only symptom throughout the clinical course.

In MG patients, the high frequency of extraocular muscle weakness depends on a high density of acetylcholine receptor. Considering that point and this case, there could be a possibility that the distribution of acetylcholine receptor differs greatly in individuals.

In the present case, since there was no thymoma and his weakness had improved due to only pyridostigmine bromide, it was not necessary to perform thymectomy or to give medication of prednisolone. MG is a treatable disease, and appropriate therapy can improve the quality of life. In patients with dropped head sign as the only symptom, the possibility of MG should be considered and a diagnosis can be obtained with the edrophonium test and an SFEMG study.
### References


© 2007 The Japanese Society of Internal Medicine
http://www.naika.or.jp/imindex.html