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CASE REPORT

# Ocular myasthenia gravis accompanied by anosmia

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## Abstract

We report a case of ocular myasthenia gravis (MG) accompanied by anosmia. A 76-year-old man had idiopathic anosmia of 2-year duration. Four months before consultation, he began to have drooping in the right upper eyelid along with muscle soreness, distension, and pain in the nape. His tongue was dark-red with a thin and white coating; his pulse was wiry and slippery. According to Traditional Chinese Medicine, eyelid drooping and anosmia are the main signs of liver constraint and spleen deficiency. In Western Medicine, the diagnosis was ocular MG and idiopathic anosmia. Our patient, along with the literature, suggests that anosmia may be an early symptom before MG. MG accompanied by anosmia could be a special subtype of MG according to antibody production and symptoms.



**Key words:** Myasthenia gravis; Olfaction disorders; Eyelids; Antibody

## INTRODUCTION

We report a case of ocular myasthenia gravis (MG) in an elderly gentleman. Before the initial consultation, he had suffered from loss of olfaction (anosmia) for 2 years as well as flaccidity and drooping in the right upper eyelid for 4 months.

After hospitalization, he was diagnosed with a drooping eyelid and anosmia in terms of Traditional Chinese Medicine (TCM), and as having ocular MG with anosmia in terms of Western Medicine. Analyses of the pathologic evidence suggested that the olfactory disorder could be manifestation of MG. We speculate that MG accompanied by anosmia is a special subtype of MG.

## CASE REPORT

The patient was a 76-year-old man. He complained of anosmia of 2-year duration, and flaccidity and drooping of the right upper eyelid for 4 months, and was hospitalized on 22 November 2012. He had developed anosmia with no obvious predisposing factor in 2010 and received no treatment. In July 2012, he went fishing and suddenly felt weak and found that his right upper eyelid was drooping, but he did not seek medical attention. The condition worsened gradually, and he tended to feel better in the morning than in the evening.

In August 2012, his vision was blocked partially by the drooping eyelid and his nape felt sore and painful. He sought help from our hospital. No abnormalities were found upon CT of the chest and he refused medications.

On 22 November 2012, his symptoms worsened and he was admitted to the Department of Encephalopathy of our hospital. His manifestations were anosmia, a drooping right upper eyelid, muscle soreness, and distension and pain in the nape. Diplopia, indifferent facial expressions, drooling, weakness in rotation of the neck, dyspnea, or weakness in the extremities were not observed.

He had a history of scrofula that had resolved. He had been diagnosed with coronary disease, but had never taken any medications for it. He did not smoke but

consumed a small amount of alcohol. His father had a history of eyelid drooping from at the age of 90 years, but no diagnosis had been made or treatment administered.

Clinical examination revealed: anosmia; drooping of the right upper eyelid: fissure in the right eyelid of 6 mm; fissure in the left eyelid of 10 mm; flexible movement of the eyeballs; pupil diameter (both sides) of 2.5 mm; both pupils sensitive to direct and indirect light reflex. The tongue and uvula protruded on the midline. Diplopia or forceful closing of the eyes was not identified. Breathing movement was symmetric on both sides, and muscle strength in the four extremities was grade 5. Volume and tone of muscle were normal. Tendon reflexes of the four extremities were symmetric, bilateral Babinski sign (and its equivalent signs) were negative, superficial and deep sensations of the trunk and four extremities were symmetric, and the ataxia test was negative. His tongue was dark red with a thin, white coating. His pulse was wiry and slippery.

On 27 November 2014, a fatigue test of the upper eyelids showed an initial fatigue time for the right upper eyelid to be 32 s (score, 1) whereas that of the left upper eyelid was > 60 s (score, 0). The neostigmine methylsulfate test showed a grade of ≥ 60%.

On 29 August 2012, routine and contrast-enhanced CT of the chest showed an old lesion in the right upper lung, as well as interstitial changes in the right lower lung. On 23 November 2012, MRI of the head showed ischemic infarction in bilateral frontal and parietal lobes, basal ganglia, and around the lateral ventricle; senile encephalic changes and minor inflammation of the ethmoid sinus were also observed.

On 29 November 2012, routine CT with three-dimensional reconstruction of paranasal sinuses showed local thickened mucus in the maxillary sinus and a bulging inferior nasal concha. Physicians from our ENT Department carried out nasal endoscopy, which showed mild congestion of nasal passages, no obvious swelling, and no neoplasm in the nasal septum or olfactory cleft. The patient refused an electrophysiology test. Causative factors of anosmia remained unknown. On 25 December, 2012, a serum test was positive for antibodies against titin, ryanodine receptors, and muscle-specific tyrosine kinase (MuSK), but negative for acetylcholine receptors and acetylcholine.

In terms of TCM, the diagnosis was a drooping eyelid and anosmia with a symptom pattern of liver constraint and spleen deficiency. In terms of Western Medicine, the diagnosis was ocular MG and idiopathic anosmia.

During hospitalization, the patient refused to take hormone and pyridostigmine therapies. Only cobamamide was administered to nourish the nerves. The TCM regimen was based on soothing the liver, strengthening the spleen, boosting *Qi*, and lifting *Yang*.

The patient was discharged on 30 December 2012 with an improved right upper eyelid. On 27 September 2014, he informed us by telephone that there was no improvement in olfaction. Since June 2014, his left eyelid began to feel weak and a drooping eyelid had developed. Currently, he is taking pyridostigmine tablets (30 mg, t.i.d.) and his condition is under control.

## DISCUSSION

### *Pathogenesis of MG*

MG is an acquired autoimmune disease that occurs mainly due to a defect/absence of nicotinic acetylcholine receptors at the post-synaptic neuromuscular junction. MG involves antibody-mediated destruction of acetylcholine receptors, is dependent on cell immunity, and is associated with complement. Causative factors include autoimmunity, passive immunity, and drug use. The typical clinical manifestation is fluctuating weakness and fatigue of certain groups of striated muscles (mostly extraocular muscles), a condition that is much milder in the morning and more severe in the evening, and which is aggravated by exertion and remitted by rest. The most common initial symptom is asymmetric ptosis and / or bilateral diplopia due to weakness of extraocular muscles.

According to the Osserman classification, MG can be categorized into five types. In type 1, only extraocular muscles are affected and no other muscle groups are involved within 2 years of the diagnosis. In type 2, mild, generalized MG involving at least one muscle group is observed. Type 3 is characterized by severe, generalized MG that occurs and progresses drastically, and respiratory muscles are often involved within 6 months of the diagnosis. In type 4, delayed, severe, generalized MG occurs in an insidious and gradual fashion, and respiratory muscles are involved within 2 years of the diagnosis. In type 5, skeletomuscular atrophy is noted within 6 months of the diagnosis.[1]

Recent studies have shown that multiple antibodies take part in MG, among which the mechanism of action of antibodies against acetylcholine receptors is relatively well characterized.[2]

Antibodies against titin are present in MG patients with antibodies against acetylcholine receptors, and suggest MG accompanied by a thymoma (MGT).[3,4] The possibility of a thymoma cannot be excluded even if a MG patient does not have antibodies against titin.[5] Studies have shown that MGT is associated with MG severity.[6,7]

Antibodies against MuSK are usually present in the serum of MG patients who do not have antibodies against acetylcholine receptors.[8] Its relevance is related to test methods, patient ethnicity[9-14] and MG type.[15,16] This type of MG occurs acutely and often involves the respiratory muscles.[17] It responds poorly to cholinesterase inhibitors and removal of the thymus gland.[18,19] A possible mechanism of action is that antibodies against MuSK influence accumulation of acetylcholine recep-