Hypokalemic Periodic Paralysis Accompanied with Myasthenia Gravis: A Case Report

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Citation

Abstract
Myasthenia gravis and hypokalemic periodic paralysis are commonly related with hyperthyroidism but rarely occur together. Here, in this article a case of hypokalemic periodic paralysis in a north Indian male accompanied with myasthenia gravis has been reported. He had motor weakness despite a normal level of serum potassium. Edrophonium test was positive. He dramatically improved after pyridostigmine treatment. Autoantibodies to nicotinic AchR-Ab and dihydropyridine receptor or L-type voltage gated calcium channel were postulated to explain these associated diseases.

INTRODUCTION
Myasthenia gravis (MG) is an acquired neuromuscular junction disorder while as hypokalemic periodic paralysis (HPP) may be familial disease with autosomal dominant inheritance. Autoimmune disease and thymic tumors are associated with myasthenia (1). Both MG and HPP are commonly associated with hyperthyroidism (2,3,4,5,6). Here, in this article a case of MG accompanied with HPP has been reported, in a north Indian man who was admitted and treated in I.C.U of Institute Of Medical Sciences, Banaras Hindu University, Varanasi – 05, Uttarpradesh, INDIA.

CASE REPORT
A 49 year old male presented with proximal muscle weakness for 10 days and was admitted to I.C.U of Institute Of Medical Sciences, Banaras Hindu University, Varanasi – 05, Uttarpradesh, INDIA. Previously, he had experienced intermittent proximal muscle weakness for three years and was diagnosed as HPP. His serum potassium was 1.8 mEq/L with normal level of bicarbonate and pH of arterial blood at the time of paralysis attack. He denied a history of drug usage, vaccinations and family members with periodic paralysis. In the past three years, he had several attacks of weakness with low serum potassium level and fully recovered by potassium replacement.

One week before, he again presented with proximal muscle weakness without a history of fluctuation, ptosis, or dysphagia. His potassium level was normal.

Physical examination revealed normal cranial nerve, proximal muscle weakness (biceps grade III/V, wrist flexion grade V/V bilaterally, hamstring grade III/V, ankle flexion grade IV/V bilaterally, generalized hyporeflexia, no sensory loss, plantar flexion bilaterally, clonus was negative bilaterally. He denied doing an electromyography test.

Urinalysis, complete blood count, serum creatinine kinase, thyroid function test, and blood test for autoimmune diseases were normal. Edrophonium test was done with positive result. His motor weakness has still completely responded to pyridostigmine 360 mg/day after 6 months follow up.

DISCUSSION
HPP is an uncommon disease of uncertain cause and may be a familial disorder. It is characterized by proximal motor weakness, hyporeflexia, and low serum potassium level without acidosis or alkalosis. Repeated episodes of muscle weakness may occur. Fatal attacks of muscle weakness or paralysis happen because of the involvement of respiratory muscle (10).

The pathogenesis of HPP is now better understood.

The abnormal gene in most patients is located on chromosome 1q. This leads to abnormal alpha-1 subunit of the dihydropyridine-sensitive calcium channel in skeletal muscle (11). Nevertheless, the mechanism of leading episodic influx potassium into the cells is not clear. On the other hand, MG is a disorder mediated by auto-antibodies against the acetylcholine receptor (AChR-Ab) (12). The history of fluctuation of motor weakness during the day is the cornerstone to differentiate from HPP. Respiratory failure...
can also be observed as in HPP. The recurrent attacks in HPP patients with normal plasma potassium level between episodes distinguish HPP from other causes. The other differential diagnoses in this patient were thyrotoxic periodic paralysis, recurrent Cushing’s syndrome, inflammatory myositis, GB syndrome and MG. Thyrotoxic periodic paralysis usually occurs in Asian men(3) with hypokalemia. The other two differential diagnoses (recurrent Cushing’s syndrome and inflammatory myositis) should demonstrate clinical symptoms and signs. Laboratory results were used to exclude these three diseases. MG patients usually show normal deep tendon reflex. However, the presented case showed hyporeflexia. Because of several episodes of HPP attacks, these may cause hyporeflexia. Therefore, we proceeded to exclude MG by Edrophonium test, the diagnostic test for generalized MG. Ocular involvement and history of fluctuation can be found 75.3% and 91.2% of generalized MG patients(11). The positive Edrophonium test and clinical response made the diagnosis of MG. Even though, both diseases are commonly associated with hyperthyroidism (1,11) The occurrence of them together rarely occurs and is not well understood. Both nicotinic AchR-Ab in myasthenia gravis and dihydropyridine receptor or L-type voltage gated calcium channel in HPP have been found to be affected by autoantibodies and/or genetic anomalies(13).

Because of good response to pyridogtigmine, thymectomy was not considered.

In conclusion, MG and HPP may be associated diseases.

References

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