Letter to Editor

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Giant cell myositis associated with myasthenia gravis and thymoma

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Dear Editor,

Myasthenia gravis (MG) is an autoimmune disease of the neuromuscular junction and 10-15% of patients with MG have thymoma (1). Thymomas are also associated with other autoimmune disorders such as pure red cell aplasia, systemic lupus erythematosus, polymyositis, and Good's syndrome (2). Giant cell myositis (GCM) is another inflammatory entity that is commonly associated with MG or thymoma (2). However, there are few reports of combined GCM, thymoma, and MG.

We report a case of 72-year-old male patient who developed weakness and stiffness in all limbs after heavy exercise. He had an upper respiratory tract infection 2 weeks ago and unintentionally lost 10 kilograms last year. A neurologic examination revealed mild weakness of neck flexors and proximal muscles of all extremities [4/5 according to the Medical Research Council (MRC) manual muscle testing scale]. Deep tendon reflexes were preserved. A blood test revealed elevated creatine kinase (1597 U/L; normal <171 U/L) and myoglobin (1445 ng/mL; normal range 17.4-105.7 ng/mL) concentrations. Cranial and cervical magnetic resonance imaging showed no significant pathology. Nerve conduction studies, needle electromyography, and jitter analysis of the right frontalis muscle using a concentric needle electrode (CNE-jitter) were normal except for increased insertional activity. Although a neostigmine test was negative, oral pyridostigmine bromide (3x60 mg/day) was given empirically. After four days, the pyridostigmine was stopped due to a lack of clinical response. A biopsy of the left biceps brachii muscle, performed due to increased creatine kinase and proximal-dominant muscle weakness, showed necrotic fibers, inflammatory cell infiltrate, and the presence of giant cells compatible with GCM. Intravenous methylprednisolone (1000 mg/day) was started. During the follow-up, the patient developed dysphagia, respiratory distress, and ptosis. Repeated electrophysiologic tests revealed a significant decremental response in 2, 3, and 5 Hz repetitive nerve stimulations in the right nasal (9%, 10%, and 12%, respectively) and abductor digiti minimi muscles (18%, 25%, and 49%, respectively), and increased jitter in CNE-jitter analysis performed in the right semispinalis muscle. Acetylcholine receptor antibody was found as 52.80 nmol/L (radioimmunoassay; normal <0.25 nmol/L) and anti-titin antibodies were found positive in an immunoblot. A paraneoplastic screen was negative for anti-amphiphysin, CV2, Ma2, Ri, Yo, Hu, and Recoverin, but positive for anti-GAD65 antibody. Oral pyridostigmine treatment was tried for the second time and plasmapheresis was started due to rapid deterioration of swallowing and respiratory functions.

Thorax computed tomography revealed an anterior mediastinal mass and thymectomy was performed. The histologic examination showed type B1 thymoma without capsular invasion. The patient has been on pyridostigmine, azathioprine, and monthly intravenous immunoglobulin treatments for 20 months. His respiratory and swallowing functions are good, but muscle weakness persists (4/5 according to the MRC scale).

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Thymoma, MG, and GCM can rarely occur simultaneously. In these cases, myositis may either occur concurrently or precede the development of MG (3). Therefore, close follow-up for MG and thymoma is important for patients with GCM. Similarly, early recognition of the presence of GCM in patients with MG is important because it affects the treatment strategy (3). Although there is no standard treatment for MG complicated by myositis, it is critical to start immunotherapy upon suspicion because it can be fatal (4).

Informed Consent: Informed consent was not obtained from the patient who participated in this study because no picture or information was used that could identify the patient.

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