Spontaneous Muscle Hemorrhage Might Be Included into Systemic Manifestation of Dermatomyositis?


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Objective

to describe a case of spontaneous muscle hemorrhage in dermatomyositis.

Case Report

A 64-year-old female patient was admitted to our Clinic for muscle weakness and iperCKemia (3200 IU/L). She had a history of treated breast cancer. Physical examination revealed symmetrical muscle weakness in proximal limbs (MRC: F2/3), pain at rest and during muscle contraction, heliotrope skin lesion on the malar and supraorbital areas.

Work-up

• Laboratory findings → creatine-kinase 1201 IU/L (normal range 30-135), lactate-dehydrogenase 678 IU/L (113-229), aspartate-aminotransferase 174 IU/L (14-36), alanine-aminotransferase 135 IU/L (9-52), Erythrocyte sedimentation rate 35 mm/h (1-15).
• Battery of autoantibodies, anti-Jo-1 included→ negative.
• Chest CT scan→ pulmonary microembolism and several nonspecific lung nodules.

Electroneuromyography→ consistent with a myopathic process.

On the basis of these results, dermatomyositis was diagnosed (Tab. 1) (1). Treatment with prednisone 1 mg/Kg daily and fractioned heparin (6000 I.U. twice daily), to prevent a new episode of pulmonary embolism, were started.

Three days later, her blood pressure and hemoglobin dropped to 80/40 mmHg and 5.8 g/dl (12-16.5), respectively. Platelet count and coagulation screening were normal. Palpation of the abdomen revealed localized tenderness in right iliac fossa. A CT abdomen scan showed hemorrhage of both iliopsoas muscles. The patient was transferred to Intensive-Care Unit because of severe shock and underwent angiography and embolization of hypogastric artery to stop prolonged bleeding.

Over the next seven days, patient required assisted ventilation and several units of packed cells were transfused (hemoglobin was stable around 9-10 g/dl), but slight hemorrhage continued. During a transfusion the patient developed acute respiratory insufficiency and died.

Our experience, and at least seven similar cases in foreign clinics (Tab. 2) (2-3) suggests that patient with dermatomyositis may have intrinsic risk factors for life-threatening intramuscular hemorrhage and corticosteroids with heparin may increase this risk. Probably, microangiopathy may result from humoral attack against the endothelium of the endomyial blood vessels, with involvement of Membrane Attack Complex, B cells and CD4 cells in the endomyial vasculature. Therefore, therapy with heparin and steroids needs careful consideration in patient with dermatomyositis.

References