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.

Table 1. Bohan and Peter diagnostic criteria for JDM [3]

A	Proximal and symmetrical muscle weakness of the pelvic and scapular girdle, anterior flexors of the neck, progressing for weeks to months, with or without dysphagia or involvement of reparatory muscles.
B	Elevation of the serum levels of skeletal muscle enzymes: creatine phosphokinase, aspartate aminotransferase, lactate dehydrogenase, and aldolase.
C	Electromyography characteristic of myopathy (short and small motor units, fibrillations, positive pointy waves, insertional irritability and repetitive high-frequency firing).
D	Muscle biopsy showing necrosis, phagocytosis, regeneration, perifascicular atrophy, perivascular inflammatory exudate.
E	Typical cutaneous changes: • heliotrope with periorbital edema and violaceous erythema; • Gottron's sign: vasculitis in the elbow, metacarpophalangeal, and proximal iterphalangeal joints.

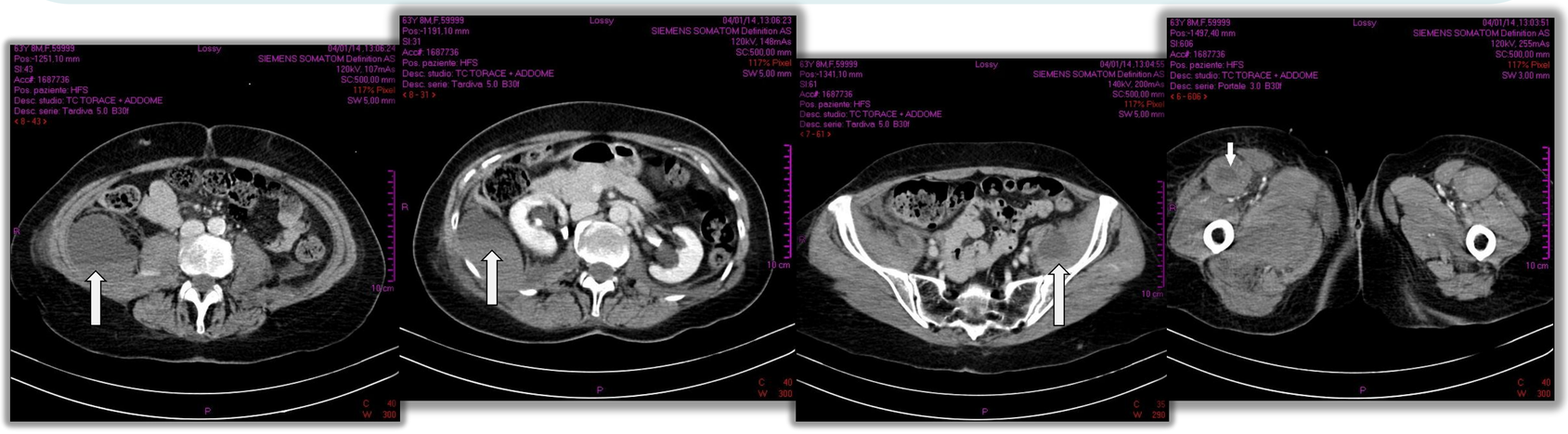
Criteria for DM	
Definitive	Three criteria (A, B, C or D) + E
Probable	Two criteria (A, B, C or D) + E
Possible	One criterion (A, B, C or D) + E

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.

CT abdomen scan

hemorrhage of both iliopsoas muscles



Conclusion

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 endomysial blood vessels, with involvement of Membrane Attack Complex, B cells and CD4 cells in the endomysial vasculature. Therefore, therapy with heparin and steroids needs careful consideration in patient with dermatomyositis.

Table 2 Muscle hematoma in dermatomyositis

Case	Age	Gender	Bleeding site	Heparin	Coagulability	Intravenous methylpredonisolone pulse	References
1	80	M	Left rectus sheath, oblique right thigh	Yes	APTT prolonged	Yes	[1]
2	50	F	Left rectus abdominis	No	Normal	No	[2]
3	11	F	Right reteroperitoneum	No	Normal	No	[2]
4	77	F	Left iliopsoas iliac, reteroperitoneum	Yes	APTT prolonged	Yes	[3]
5	64	F	Right reteroperitoneum, left rectus sheath	Yes	Normal	Yes	[4]
6	65	F	Iliopsoas both sides, thigh	Yes	APTT prolonged	Yes	[5]
7	60	M	Left trapezius	Yes	APTT prolonged	Yes	Our case

References

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