Between Symptoms and Diagnosis: Navigating the Uncertainty of Dermatomyositis Without a Muscle Biopsy

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Introduction

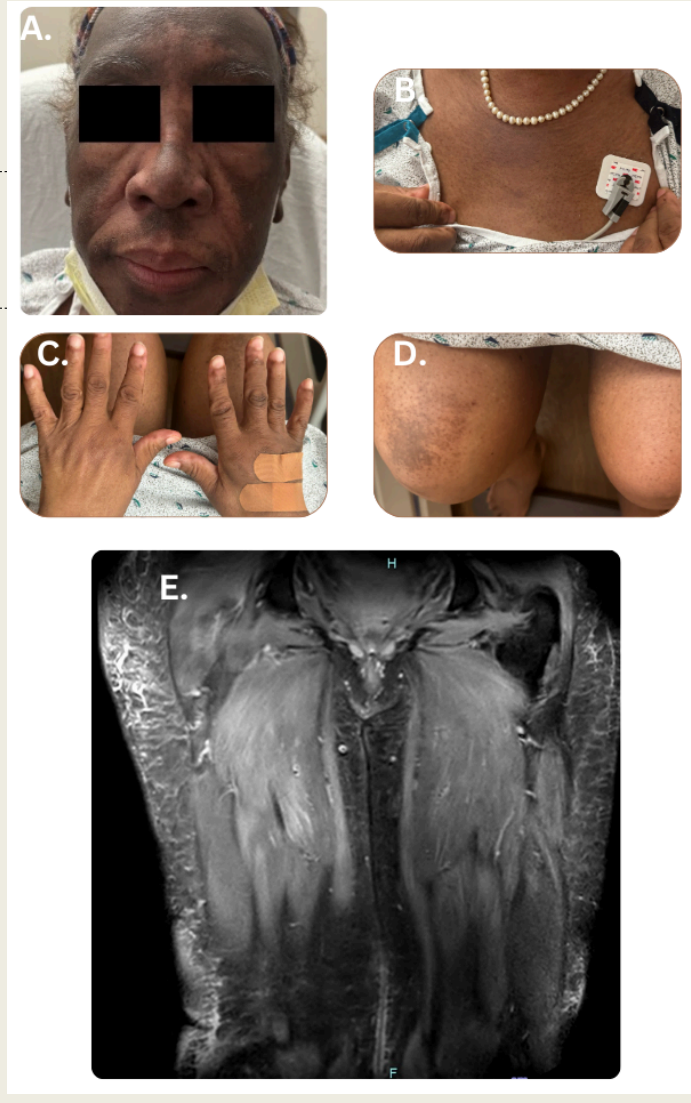
Dermatomyositis is a complex condition that manifests through a combination of muscle weakness and distinctive skin changes
It typically presents with symmetric proximal muscle weakness and is often accompanied by characteristic rashes such as heliotrope rash, Gottron's papules, the V sign, and the shawl sign.
The diagnosis traditionally relies on a combination of clinical presentation, laboratory results, and histological findings. However, in some cases where the clinical presentation is particularly clear, the role of a muscle biopsy may be reconsidered.

Patient Presentation

A 66-year-old woman with no significant medical history presented after a fall.
She reported progressive muscle weakness and skin hyperpigmentation in some areas for six months, unresponsive to physical therapy
The patient was not on any medication.
On physical examination, she had diffuse hyperpigmentation across her scalp, face, upper chest, and back, as well as erythema on her fingertips and small hyperpigmented papules on the DIP and PIP joints.
Her muscle strength was reduced to 2/5 in the hips and 3/5 in the shoulders.
Laboratory tests revealed elevated levels of creatinine kinase (CK) at 1500 U/L, C-reactive protein (CRP) of 21mg/dL, normal TSH , and a positive antinuclear antibody (ANA) (1:2560, homogenous pattern)
Additionally, the myositis panel was positive for high titer anti-Mi-2 alpha and beta antibodies and elevated aldolase levels.
An MRI of the lower extremities showed bilateral diffuse, symmetric intramuscular edema.
The patient refused to have a muscle biopsy. A skin biopsy revealed post-inflammatory changes and interface dermatitis.

Pictures

Figure A. Hyperpigmentation on scalp and face
Figure B. Hyperpigmentation over the chest
Figure C. Hyperpigmented papules on the DIP and PIP
Figure D. Hyperpigmentation on the Right Knee
Figure E. MRI of bilateral lower extremities (T2 STIR image) showing diffuse and symmetric intramuscular edema in the lower extremities bilaterally



Clinical Course

Prednisone 50mg daily was started, which led to an improvement in her symptoms.
At her two-week follow-up , her muscle weakness continued to improve, and azathioprine was added for long-term management.
TPMT was within normal limits
She will continue follow-up with primary care for age-appropriate cancer screening.

Discussion

The decision to diagnose dermatomyositis without a muscle biopsy presents a unique challenge, requiring a balance between clinical judgment and available evidence.1,2
Signs of dermatomyositis may present differently in people of color.
A case series show skin dyschromia were more pronounced in locations found in dermatomyositis compared to the erythematous/violaceous patches seen in patients with lighter skin tones.[3]
Single center study showed that clinicians took longer to diagnose and treat DM and Atypical DM in patients with dark skin.[4]
Familiarization of the clinical features in people with darker skin will help decrease racial disparities in our field.[5]
The EULAR/ACR criteria offer a robust framework, demonstrating high sensitivity and specificity even in the absence of a biopsy.
The concerns surrounding invasive procedures like muscle biopsies are real, and this case encourages us to reflect on their necessity in light of strong clinical and laboratory indicators.

References

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