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Alvaro J Ramos-Rodriguez et al., Intern Med 2017, 7:4 (Suppl)
DOI: 10.4172/2165-8048-C1-006

2nd International Conference on

Internal Medicine & Hospital Medicine

September 13-14, 2017 Dallas, USA

An uncommon condition: Hypomyopathic dermatomyositis

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Background: Hypomyopathic dermatomyositis (HDM) is a very rare condition with a reported incidence of 2 in 1,000,000; it represents less than 5% of all dermatomyositis diagnoses. It is characterized by the classical dermatomyositis cutaneous findings with less evident muscle damage. HDM has been associated with rapidly progressing and refractory interstitial lung disease (ILD) in multiple studies hence may carry a worse prognosis. The diagnosis is usually established with imaging and muscle biopsy along with clinical findings as serological testing may be negative for muscle enzymes. We present a rare case of hypomyopathic dermatomyositis in an African-American adult with impressive cutaneous findings.

Case Report: A 45-year-old African-American female with past medical history of asthma presenting to the clinic complaining of 1 month of generalized rash is considered. Review of systems was negative except for mild proximal muscle weakness. Physical exam was remarkable for skin phototype VI, edematous and violaceous plaques on periorbital skin (Heliotrope rash), erythematous eruption in the chest, back and abdomen and prominent Gottron's papules on extensor surface of bilateral hands with corkscrewing of nail fold capillaries. Laboratories were unremarkable including normal levels of creatinin phosphokinase and aldolase. Autoimmune panel was positive for antinuclear antibody and anti-RO/SSA (126 U). Electromyography of the 4 extremities was inconclusive. Magnetic resonance imaging revealed increased intensity in biceps, triceps, deltoids, gluteus, and abductor muscles of the proximal lower extremities. Skin biopsy reported non-specific findings of muscle inflammation with dense lymphocytic infiltrate and atrophy of muscle fibers. Patient was subsequently diagnosed with hypomyopathic dermatomyositis and was started on prednisone 50 mg and hydroxyzine 25 mg daily and instructed to avoid further sun exposure. Follow-up computerized tomography of chest, abdomen and pelvis was negative for malignancy and lung disease.

Conclusion: Hypomyopathic dermatomyositis is an uncommon condition that may be easily overseen by physicians as there is usually no serological evidence of muscle damage. This case highlights the importance of a good dermatologic exam to diagnose internal diseases and prevent potential life-threatening complications. In our case, we ruled out malignancy given the high association with dermatomyositis, and our patient was monitored closely in the outpatient clinic for development of ILD.

Biography

Alvaro J Ramos-Rodriguez is currently a Medical Resident Physician at the Icahn School of Medicine at Mount Sinai West. He is the author of the recently published textbook *Dermatology for the USMLE*. He has dedicated a major part of his medical career to teaching and helping students prepare for the USMLE, including teaching review courses. His interest in dermatology includes DRESS syndrome, toxic erythema of chemotherapy, atopic dermatitis, hidradenitis suppurativa, dermatomyositis, psoriasis and infectious skin disorders.

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