False Reassurance: Complications of Mi2+ Dermatomyositis.

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INTRODUCTION

Myositis-specific autoantibodies have been beneficial for both diagnostic purposes as well as understanding the associated clinical phenotypes. Anti-Mi2-antibodies are strongly associated with dermatomyositis (DM) and have hallmark features of cutaneous disease, milder muscle involvement, and an overall positive prognosis[1]. Identification of typical characteristics of antibody-associated myositis can be helpful for both diagnostic purposes as well as screening for associated conditions.

CLINICAL CASE

A 53 year old male who was evaluated in the rheumatology office with increased myalgia after routine exercise, unintentional weight loss, skin changes, and rash over the bridge of his nose found to have abnormal capillary nailfold findings. Further workup showed elevated creatine kinase (CK), aldolase, liver function studies, and positive anti-Mi2 antibody (tested at Arup on a standardized myositis panel), consistent with the diagnosis of DM. Other contributing factors for myopathy were ruled out with thyroid screen, urine drug screen, vitamin D level, and viral serologies. For completeness, the patient underwent screening for malignancy and ILD with computed tomography of his chest/abdomen/pelvis yielding normal results. The patient was started on prednisone and methotrexate with initial notable clinical improvement. Upon further corticosteroid dose taper the patient developed profound ulcerations on the dorsal aspects of his metacarpal phalangeal joints, olecranon processes, and helix of the right ear. As these ulcerations are more characteristic of DM with anti-melanoma differentiation associated gene 5 antibodies (MDA-5) than anti-Mi2 antibodies, the patient was tested and found to be negative for the presence of MDA-5 antibodies. His prednisone dose was increased and methotrexate was continued with subsequent improvement and resolution of these lesions. Mycophenolate mofetil was added to the patient's regimen as combination therapy with methotrexate given the aggressive nature of the ulcerative lesions. Clinically, the cutaneous lesions improved, and he was able to be tapered off of his steroid dosing without recurrence of ulcerations.

Table 1. Features of Myositis-specific Autoantibodies

Myositis-specific autoantibody	Typical Characteristics/Associations
Anti-Mi2+ autoantibodies	• Cutaneous features: Gottron's papules, helio-trope rash, V-sign and shawl sign rashes, cuticular overgrowth. • Pulmonary features: no typical pulmonary involvement. • Typically respond well to immunosuppressive therapy.
Anti-MDA5 autoantibodies	• Mucocutaneous features: palmar erythema-tous macules+/-papules with higher likelihood of cutaneous ulceration. Gum pain, alopecia, mechanic hands, Gottron sign. • ILD-association, especially rapidly progres-sive lung disease.

Comment: *Typical features associated with anti-Mi2+ and anti-MDA5 autoantibodies[1,2].*

CONCLUSION

The skin ulcerations the patient developed were atypical for anti-Mi2 associated DM[1]. Research regarding the development of these ulcerations correlates more with patient's positivity for MDA-5 antibodies, which portends more severe disease with worse prognosis[2]. Identification of clinical phenotype of myositis-associated and myositis-specific autoantibodies assists in the diagnostic workup and management of related conditions. Although management can be adjusted by the provider when the disease does not fit the typical phenotype, concerns truly arise with respect to predicting complications of potentially severe disease such as ILD or risk of malignancy. This case demonstrates the clinical conundrum regarding screening and prognostic implications for patients who demonstrate worrisome features not usually associated with the patient's antibody phenotype.

References:

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