Examining the Matrix: A Case of Anti-Nuclear Matrix Protein 2 (NXP-2) Positive Dermatomyositis

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Title:

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Background:
Dermatomyositis is a disease characterized by proximal muscle weakness, elevated muscle enzymes and cutaneous skin findings including heliotrope rash, periungual erythema, Gottron's papules/sign and shawl-sign. Myositis-specific autoantibodies (MSA) can be used to predict disease manifestations, response to therapy and prognosis. Specifically, patients with dermatomyositis with positive anti-nuclear matrix protein (NXP-2) typically present with classical skin findings, subcutaneous edema, profound muscle weakness, severe dysphagia and hypophonia.

Case Presentation:
A 21-year-old Hispanic lady presented to the hospital for severe muscle weakness. She reported soreness in her thighs for three months prior to presentation followed by arm soreness. She also developed a facial and left neck rash along with complaints of dysphagia and hypophonia. She denied joint pain or constitutional symptoms. Exam was notable for marked edema of upper and lower extremities, malar erythema not sparing the nasolabial folds, heliotrope rash, and a crusting lesion on the left breast that was previously reported as necrotic. Muscle strength was diminished in the proximal aspect of extremities; patient was unable to stand from a seated position. Her initial CPK was 14000. In addition, AST and ALT were elevated and ANA was positive with a nuclear speckled pattern. Chest CT was remarkable for a focal opacity in the posterior right upper lobe, most likely focal aspiration pneumonia for which the patient was placed on antibiotics. She was initially managed with vigorous IV fluids for suspected rhabdomyolysis, and then subsequently with oral prednisone with improvement of her CPK values and modest improvement of her muscle weakness. The 11 antibody myositis panel was positive for the NXP-2 antibody. Due to the patient's presentation of profound muscle weakness and suggestive rash, in addition to high titer myositis specific antibodies, she was diagnosed with dermatomyositis and started on pulse IV steroids, intravenous immunoglobins and azathioprine. In the following days, patient was able to ambulate and CPK, aldolase and liver function tests were down trending.

Conclusions:
Myositis specific antibodies are a group of antibodies that have been associated with idiopathic inflammatory myopathies (IIM) and can be utilized in the diagnosis, assessment, management and prognosis of IIM. These antibodies are generally mutually exclusive and may help in characterization of a phenotypic presentation. Furthermore, MSA may be valuable in targeted therapies. Our patient had high titer NXP2 antibodies. These antibodies are more common in the juvenile form of dermatomyositis. Anti-NXP2 autoantibodies are found to confer a risk of developing five clinical characteristics: peripheral edema, muscle weakness, myalgia, dysphagia, and a reduced risk of interstitial lung disease.