

agglutinin disease. The patient had no further complications.

Review of the medical records revealed that in 9 of the patient's complete blood counts over the previous 2 years, the laboratory had commented that the patient's blood sample "required rewarming" and appeared "markedly agglutinated." Laboratory results from the previous year showed a progressive decrease in hemoglobin and gradual increases in lactate dehydrogenase and mean corpuscular volume, indicating progression of CAD. An increase in the CD4-to-CD8 ratio was also observed, indicating simultaneous progression of SS. These findings suggest that the patient may have had pathologic CAs causing low-grade hemolysis and agglutination for 2 years while undergoing ECP.

The incidence of CAD is 1 per million per year, and the disease is divided into primary and secondary subtypes.¹ Primary CAD is monoclonal, chronic, and associated with lymphoproliferative malignancies, most frequently CLL. Secondary CAD is transient, polyclonal, and associated with viral and mycoplasma infections.¹ CAs are found at low levels in the sera of most healthy individuals and react at low temperatures. They may become pathologic at high titers or high thermal amplitudes, the temperature at which the autoantibodies react.¹⁻⁴ Activation of CAs—precipitated by exposure to low temperatures and acute phase reactants—can result in cold-induced circulatory symptoms such as acrocyanosis, as well as catastrophic hemagglutination and hemolysis.^{1,2} Clues to the diagnosis of CAD in our patient include chronic macrocytic anemia, a history of CLL, and temperature-dependent hemagglutination.¹⁻⁵ Although his CAD and SS were progressing simultaneously, there are no reports of a correlation between the 2 conditions. We attribute the patient's CAD to his underlying CLL. Hemagglutination may have been precipitated by his recent viral illness and environmental conditions during ECP. We have presented a potentially serious complication that occurred during the use of a therapy that is broadly applied by multiple medical specialties. Awareness of the risk factors, signs, and symptoms of CAD, and appropriate pretreatment screening may help avoid this rare but significant adverse event in patients with CAs undergoing ECP.

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A case of Wong-type dermatomyositis with concomitant anti-MDA5 features

To the Editor: Dermatomyositis (DM) is a multisystem autoimmune disease for which multiple phenotypes have been described. Here we report the case of a patient with both Wong-type DM and features of a DM phenotype associated with anti-melanoma differentiation-associated gene 5 antibodies (anti-MDA5 DM).

A previously healthy 54-year-old man presented with a 1-year history of a progressive eruption previously diagnosed as pityriasis rubra pilaris (PRP) by 2 independent dermatologists. He had failed topical corticosteroids and 2 courses of oral prednisone pulses, and subsequently weakness and dyspnea on exertion developed. His past medical history was unremarkable.

Exam revealed confluent pink-orange patches and plaques over the trunk with islands of sparing (Fig 1), and his extremities displayed hyperkeratotic follicular papules. He also had painful violaceous palmar papules (Fig 2), ragged cuticles, rough radial palms and digits, and subtle violaceous periorbital edema. Proximal muscle strength was 4-5+.



Fig 1. Wong-type dermatomyositis. Widespread confluent salmon pink/orange colored patches and scaly plaques with well defined islands of sparing over the trunk.



Fig 2. MDA5 dermatomyositis. Multiple violaceous palmar and digital papules and rough radial palms and digits.

Histology demonstrated superficial perivascular lymphocytes and rare eosinophils below a thin epidermis with vacuolar interface change and necrotic keratinocytes. There was increased mucin in the superficial reticular dermis and hyperkeratosis over follicular infundibulum.

A mild elevation of creatine kinase (CK) and inflammatory markers were noted. Results were positive for antinuclear antibodies (speckled

pattern) and negative for antibodies to Jo1, SSA, and SSB. Electromyography (EMG) and muscle biopsy were consistent with DM. High-resolution chest computed tomography and an age-appropriate malignancy work-up were unremarkable.

Taken together, his mixed DM and PRP-like presentation, histopathology, EMG, muscle biopsy, and CK elevation were consistent with Wong-type DM. His distinct painful palmar papules raised concern for the anti-MDA5 antibody positive DM subtype. Subsequent enzyme-linked immunosorbent assay for anti-MDA5 was positive. He was treated with intravenous methylprednisolone (subsequently transitioned to a slow prednisone taper), intravenous immunoglobulin, mycophenolic mofetil, and hydroxychloroquine. He experienced rapid recovery and demonstrated only minimal cutaneous symptoms and no internal manifestations after 10 months.

Wong-type DM is an exceedingly rare DM subtype characterized by PRP-like morphology,¹ which has been described in all 22 prior cases.² Histologic findings vary, with features ranging from those typical of DM or PRP alone, to a combination of both (as seen in our patient). Hyperkeratosis overlying follicular infundibulum, as reported here, is common.²

Similar to typical DM, Wong-type appears to carry a risk for myositis and interstitial lung disease (ILD). Unlike other subtypes, Wong-type DM is not associated with internal malignancy.² Our patient's malignancy work-up was negative.

Anti-MDA5 DM is a recently described unique DM phenotype with a number of distinct morphologic features, with painful palmar papules and mucocutaneous ulcerations being among the most characteristic.³ Incidence ranges from 10% to 20% in Japan to 7% to 13% in the United States.³⁻⁵ Our patient both displayed many anti-MDA5 features, including painful violaceous palmar papules, and had a positive anti-MDA5 ELISA.

The most important consequence of anti-MDA5 DM is an increased risk for rapidly progressive ILD and resultant decreased survival, relative to other subtypes.^{3,4} Therefore some recommend early and aggressive treatment in suspected cases of anti-MDA5 DM.⁴ Of note, internal malignancy appears to be less common in anti-MDA5 DM. Fortunately, our patient had neither ILD nor malignancy.

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Dermatomyositis associated with anti-tumor necrosis factor therapy in a patient with psoriasis

To the Editor: Anti-tumor necrosis factor (a-TNF) agents have become a common treatment for many different autoimmune diseases. Due to conflicting effects mediated by TNF, a-TNF therapies both treat and induce autoimmune disease. Herein we discuss a novel case of a-TNF-induced amyopathic dermatomyositis (DM) in a patient with a history of psoriasis.

A 37-year-old Brazilian woman was referred to our medical center for management of psoriasis. She had a 7-year history of biopsy-proven psoriatic plaques that had failed topical therapies and were exacerbated by UV therapy. Physical exam revealed a large 20- × 30-cm erythematous, scaly plaque on her back, and more than 20 guttate papules on her elbows and extensor forearms. She was started on adalimumab 40 mg biweekly following routine lab work and a negative tuberculosis test.

One month later, her psoriasiform plaques had markedly improved, but indurated violaceous plaques overlying her knuckles with ragged cuticles and looped telangiectasias developed (Fig 1). Because she was satisfied with the resolving psoriasis, adalimumab was continued.



Fig 1. Dermatomyositis. Erythematous papules overlying the metacarpal and interphalangeal joints (Gottron papules).

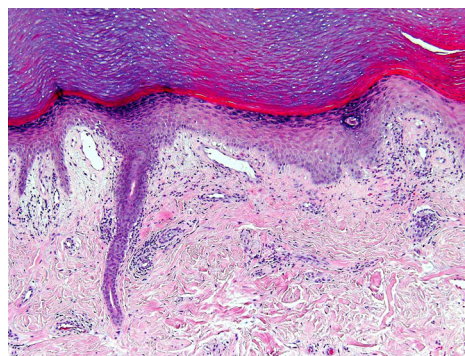


Fig 2. Dermatomyositis. Hematoxylin and eosin stain of dorsal hand punch biopsy. Vacuolar interface dermatitis with sparse perivascular inflammation.

A biopsy specimen from the periarticular dorsal hand skin papule was obtained (Fig 2).

Biopsy revealed an atrophic epidermis and subtle vacuolar interface change associated with sparse superficial perivascular inflammation with increased dermal mucin. She denied myalgias or muscle weakness. Laboratory findings revealed a negative ANA, negative SCL70, a creatine kinase of 136 U/L, and a jo-1 < 1. Based on the clinical, laboratory, and pathologic findings, the diagnosis of DM was made. The patient has had negative results on age-appropriate cancer screenings, including gynecologic exam, computed tomography of abdomen/pelvis, and chest roentgenogram, as well as normal pulmonary function tests. Over the subsequent 10 months, she has had no other manifestations of DM. Her Gottron papules have softened somewhat with the addition of methotrexate. To our knowledge, anti-TNF-induced DM has not been previously reported in a patient with psoriasis.

Vasculitis, psoriasiform eruptions, and a lupus-like syndrome are the most common a-TNF-induced