

Rapidly Progressive Interstitial Lung Disease in Melanoma Differentiation-Associated Gene 5 (MDA5) Antibody Positive Dermatomyositis



Carmen Montagnon, M.D.^a, Caitrin Coffey, M.D.^b, Shreyasee Amin, M.D., M.P.H^b, and Ashima Makol, M.B.B.S^b

- a. Mayo Clinic Department of Internal Medicine, Rochester, MN, 55905, USA
- b. Mayo Clinic Division of Rheumatology, Rochester, MN, 55905, USA

Abstract

A 56-year-old male presented to clinic with a 3-month history of increasing dyspnea, 1-month history of skin lesions, and 2-week history of arthralgias and decreased muscle endurance. Physical exam revealed inspiratory crackles to the midlungs bilaterally, intact muscle strength but fatigability, stiffness of the hands, raised rash on his torso, dry scaly plaques on the fingers, and tender, papular lesions on the palms. He was Jo-1 negative, but melanoma differentiation-associated gene 5 (MDA5) antibody positive. CK and aldolase were normal, but MRI of lower extremity muscles showed patchy muscular edema/inflammation. CT chest showed patchy bilateral groundglass and reticular opacities suggestive of organizing pneumonia. Skin biopsy was consistent with dermatomyositis. The patient was diagnosed with MDA5-positive dermatomyositis with interstitial lung disease (ILD) and initiated on prednisone 40 mg daily within 2 weeks of presentation; higher doses were precluded due to side effects. His arthralgias and rash improved, however dyspnea continued to worsen over the next 2 weeks. Mycophenolate mofetil was added within a month of presentation. Unfortunately,

his respiratory status rapidly deteriorated over the next 2 weeks, requiring intensive care unit admission and did not significantly improve despite pulse IV methylprednisolone, cyclophosphamide, plasmapheresis, and ECMO support. Due to minimal clinical improvement, he is currently being considered for lung transplantation while rituximab is being added. ILD accompanying MDA5 dermatomyositis is often rapidly progressive and associated with a poor prognosis.¹⁻³ early recognition with aggressive upfront treatment with immunosuppression is critical for management, but deterioration may still occur.^{1,3}

Biography:

Dr. Montagnon attended Mayo Clinic Alix School of Medicine where she graduated in 2019. She is currently an internal medicine preliminary year resident and will begin dermatology residency at Mayo Clinic in July 2020.

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Presenting author details:

Name: Dr. Carmen Montagnon

Contact number: (925) 759-2280

Email address: montagnon.carmen@mayo.edu

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