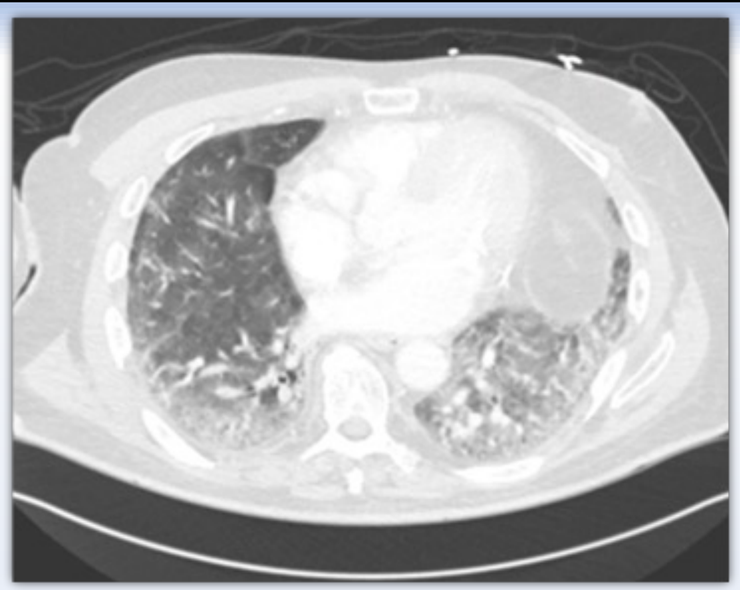
The Perilous Progression: Anti-MDA5 Dermatomyositis Leading to Rapidly Progressive ILD

Divya Samat MD, Tahminah Kausar MD, Haider Khan MD, Shivam Singh MD

Tower Health-Reading Hospital, PA

BACKGROUND

Anti-melanoma differentiation-associated gene 5 (MDA5) dermatomyositis (DM) is a subtype characterized by mucocutaneous ulceration, palmar papules, alopecia, arthritis, and significantly increased risk of rapidly progressive interstitial lung disease (RP-ILD). Patients with anti-MDA5+ DM have over a 20-fold higher risk of RP-ILD compared to anti-MDA5- patients. Systemic involvement in these patients poses a substantial mortality risk.



Rapidly Progressive ILD

CONCLUSION

Anti-MDA5 amyopathic dermatomyositis presents atypically compared to cutaneous DM, often leading to delayed diagnosis and poor outcomes. Early identification and aggressive intervention are crucial, as prompt treatment can improve outcomes even after ILD has developed.

LEARNING POINTS

- For patients presenting with RP-ILD, anti-MDA5 DM should be considered in the differential, even in the absence of muscular or cutaneous symptoms.
- Additionally, patients with anti-MDA5 antibodies are at increased risk of RP-ILD and should undergo HRCT screening at diagnosis.

CASE PRESENTATION

An 85-year-old man with recently diagnosed interstitial lung disease (ILD) on extended prednisone taper, hypertension, and type 2 diabetes presented with acute worsening of shortness of breath. He was hypoxic (SaO2=88% on room air) but normotensive. Three months prior, he had developed productive cough with clear sputum, congestion, and progressive dyspnea that worsened when lying down. He had no fever, chills, chest tightness, wheezing, or lower extremity edema. Lung sounds were clear, and chest X-ray was inconclusive for pneumonia. Treated for rhinosinusitis with antibiotics and symptomatic management, his symptoms persisted, leading to a hospital visit 4 days later. Investigations revealed negative sputum culture and BNP of 56 pg/ml. Chest X-ray showed chronic bibasilar opacities, particularly on the left, with no acute cardiopulmonary findings. HRCT revealed subpleural ILD/pulmonary fibrosis with a Usual Interstitial Pneumonia (UIP) pattern. He improved slightly with iv steroids and nebulizers and was discharged on inhaled steroids with a follow-up appointment. Subsequent serologies were positive for ANA and MDA5 antibodies, and he was started on prednisone taper.

He later presented with worsening dyspnea and right lower limb edema. He denied chest pain, palpitations, dizziness, recent travel, prolonged immobilization, sick contacts, smoking, muscle pain, fatigue, rashes, or a history of cancer or hypercoagulable disorders. CT angiography confirmed an acute pulmonary embolism with small clot burden, diffuse ground-glass opacities, and bibasilar pulmonary fibrosis. DVT screening was positive. He was admitted for treatment with Eliquis and anticipated early discharge. Despite these measures, his condition deteriorated, leading to acute hypoxic respiratory failure and escalation to IMU-level care. He was placed on HFNC, and Pulmonology and Rheumatology were consulted. Treatment for severe and rapidly progressive ILD in the context of anti-MDA5 dermatomyositis included prednisone 50 mg, intravenous immunoglobulin for 5 days, and mycophenolate mofetil 1.5 g daily. Despite maximal treatment, he remained severely dyspneic and could not tolerate weaning. Due to rapid progression, he chose to transition to comfort care and passed away the following day.

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