Annals of Arthritis and Clinical Rheumatology

Clinical Image
Published: 21 Jul, 2020

# Interstitial Lung Disease as the Initial Manifestation of Anti-MDA-5 Positive Amyopathic Dermatomyositis

*Ambreesh Chawla*, Ann George, Shazia Beg and Sujatha Vuyyuru*

*Department of Rheumatology, University of Central Florida College of Medicine – Orlando, USA*

## Clinical Image

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by muscle weakness and distinctive skin findings. Interstitial Lung Disease (ILD) is a well-recognized complication and is seen in at least 10% of cases. Occasionally, the cutaneous manifestations of DM occur in the absence of clinically apparent muscle involvement, which is referred to as amyopathic dermatomyositis. The presence of myositis Specific Autoantibodies (MSA's) are felt to predict clinical manifestations and prognosis in DM. Of the MSA's, anti-Melanoma Differentiation-Associated gene 5 (MDA5 or CADM-140) is associated with rapidly progressive ILD. We report a unique case of amyopathic DM (with positive anti-MDA5) in a middle-aged woman initially presenting with dyspnea secondary to ILD.

A 54-year-old Caucasian woman with a history of recently diagnosed idiopathic pulmonary fibrosis presented to our clinic post hospitalization. Patient was sent to rheumatology for evaluation for connective tissue disease-ILD. Per patient, she was in her usual state of health until three months prior to hospitalization when she noticed development of a widespread photosensitive rash on her chest. The rash was accompanied with new-onset worsening dyspnea which ultimately led to a 7-day hospitalization for hypoxemic respiratory failure. PFT's revealed decreased FEV1 (72% predicted), FVC (69% predicted), TLC (73% predicted) and markedly diminished DLCO (27% predicted). High resolution CT-Chest revealed moderate traction bronchiectasis and honeycombing, consistent with usual interstitial pneumonia. In our clinic, the patient presented on home oxygen. Review of systems was positive for dyspnea and rash. Nail fold capillary microscopy revealed dilated loops. Exam also revealed 'V sign' rash over the anterior chest wall and lung auscultation demonstrated inspiratory velcro-like crackles. Muscle strength was intact. Labs revealed ANA 1:80 (homogenous) with normal ESR, CRP, LDH, CPK, Aldolase. All the following were negative: RF, CCP, SS-A/B, DsDNA, Smith and RNP. Further testing of DM and scleroderma-related autoantibodies were unremarkable with exception of the anti-MDA5 antibody. Urine studies were negative for occult blood and protein. She was started on a combination of high dose prednisone & mycophenolate.

The presence of MSA's can be helpful for both diagnostic and prognostic purposes in DM. The following MSA's have been well described in the literature for their association with ILD: Anti-synthetase antibodies (JO-1, PL-7, PL-12, OJ, EJ, KS, ZO), anti-MDA5 (anti-CADM140) and anti-PM-Scl. Anti-MDA5 is seen in about 10% Caucasian DM patients and 30% to 40% Asian DM patients. These patients may present with ulcerations, papules, arthritis, fever and rapidly progressive ILD. A subset of these patients has clinically amyopathic DM. Despite aggressive conventional treatments (high dose prednisone in combination with either cyclophosphamide, calcineurin inhibitors, mycophenolate, rituximab or tofacitinib), the 6-month mortality is around 50%. While antibody testing is helpful, early aggressive combined immunosuppressive therapy coupled with serial pulmonary surveillance may help improve their prognosis despite the risks of infection (Figure 1a,1b).

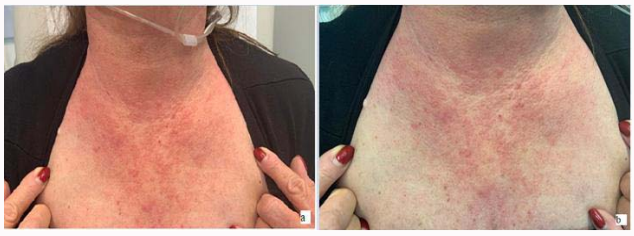

**Figure 1a,1b:** V-Sign as seen in our patient with Anti-MDA5-amyopathic dermatomyositis.

**OPEN ACCESS**

***Correspondence:***
*Ambreesh Chawla, Department of Rheumatology, University of Central Florida College of Medicine – Orlando, USA,*
*E-mail: ambreeshchawla@gmail.com*

**Received Date**: *27 Jun 2020*
**Accepted Date:** *16 Jul 2020*
**Published Date:** *21 Jul 2020*

***Citation:***
*Chawla A, George A, Beg S, Vuyyuru S. Interstitial Lung Disease as the Initial Manifestation of Anti-MDA-5 Positive Amyopathic Dermatomyositis. Ann Arthritis Clin Rheumatol. 2020; 3(2): 1020.*

***Copyright*** *© 2020 Ambreesh Chawla. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.*