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Clinical Image
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Favorably Unfavorable: Interstitial Lung Disease as the Initial Manifestation of Anti-Mi-2 Positive Dermatomyositis

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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 of DM and is seen in at least 10% of cases. The presences of Myositis-Specific Autoantibodies (MSA's) are felt to predict clinical manifestations (including ILD) and prognosis in DM. Of the MSA's, anti-Mi-2 (which targets the chromodomain helicase DNA binding protein, CHD4) is classically associated with skin and muscle features of DM, and to be protective for ILD with a favourable prognosis (5-year survival >90%). We report a unique case of DM (with positive anti-Mi-2) in a young woman initially presenting with dyspnea secondary to ILD.

A 25-year-old Hispanic woman with a one-year history of Raynaud's Phenomenon (RP) presented to our clinic for initial evaluation. Outside of RP, she had no prior medical history and was not on any medications. Review of systems was positive for dyspnea on exertion. On exam, the patient demonstrated severe asymmetric digital blanching with sequential color changes and evidence of an unhealed digital ulceration. Nail fold capillary microscopy revealed dilated loops and avascularity. Auscultation of the lungs demonstrated fine inspiratory velcro-like crackles. Labs revealed ANA 1:640 (dual pattern: Homogenous, speckled) with elevated CPK, Aldolase, RF, ESR, CRP and mild transaminitis. All the following serologies were unremarkable: DsDNA, Smith, RNP, Chromatin, SS-A/B, ACPA, antiphospholipid antibody panel. Urine studies were negative for occult blood and protein. Further testing of DM and scleroderma-related autoantibodies were unremarkable with exception of the anti-Mi-2 antibody. PFT's revealed a decreased FEV1 (71% predicted), FVC (65% predicted), TLC (80% predicted) and markedly diminished DLCO (45% predicted). High resolution CT-Chest revealed bilateral subpleural reticulation and honeycombing, consistent with Usual Interstitial Pneumonia (UIP). During subsequent clinic visits, the patient experienced proximal muscle weakness (UE>LE) and developed a cutaneous eruption over the posterior shoulders and proximal UE. She was maintained on a combination of high dose prednisone & mycophenolate, in addition to pharmacotherapy for RP.

The presence of MSA's directed against cytoplasmic proteins, ribonucleoproteins and other

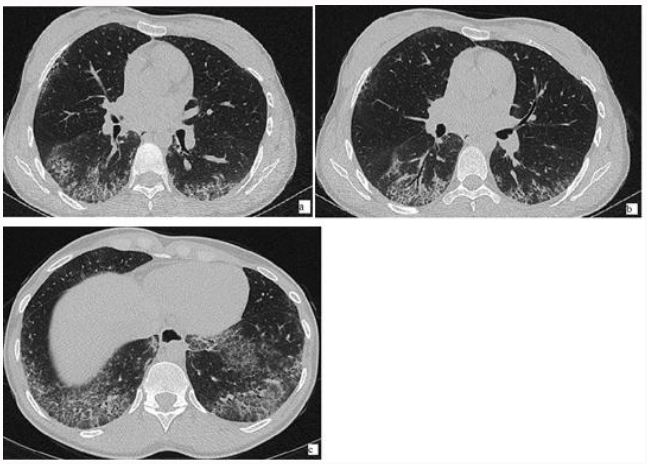

Figure 1a-1c: HRCT demonstrating honeycombing and traction bronchiectasis consistent with Usual Interstitial Pneumonia (UIP) ILD. Traction bronchiectasis is caused by the fibrotic lung pulling on the bronchi, causing irreversible dilatation.

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nuclear antigens 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 antibody (polymyositis/scleroderma overlap). Anti-Mi-2 can be seen in both adult and juvenile DM and is usually associated with cutaneous disease, myositis with good response to treatment and a lack of ILD. Our case demonstrates a noteworthy presentation where UIP-ILD coincides with the presence of a 'favourable' MSA (anti-Mi-2). While antibody testing is helpful for both diagnostic and prognostic purposes, our case delineates the gravity of screening for less frequently involved organs irrespective of a patient's MSA profile (Figure 1a-1c).