15. Redefining Dermatomyositis: Description of new diagnostic criteria that differentiate pure dermatomyositis from overlap myositis with dermatomyositis features.

Marie-Pier Payette, Yves Troyanov, MD, FRCPC, Ira N. Targoff, MD, Jean-Pierre Raynauld, MD, FRCPC, Suzanne Chartier, MD, FRCPC, Jean-Richard Goulet, MD, FRCPC, Josiane Bourré-Tessier, MD, FRCPC, Eric Rich, MD, FRCPC, Tamara Grodzicky, MD, FRCPC, Marvin Fritzler, MD, PhD, FRC
Department of Medicine, Divisions of Rheumatology (YT, MPP, JPR, JRG, JBT, ER, TG, JLS), Internal Medicine (FJ, MK) and Dermatology (SC), Centre Hospitalier de l'Université de Montréal, University of Montreal School of Medicine, Montreal, Quebec, Canada; Department of Medicine, Division of Rheumatology, Hôpital du Sacré-Coeur (YT), University of Montreal School of Medicine, Montreal, Quebec, Canada; Veterans Affairs Medical Center (IT), University of Oklahoma Health Sciences Center, and Oklahoma Medical Research Foundation, Oklahoma City, OK, USA; and Mitogen Advanced Diagnostics Laboratory (MJF), Faculty of Medicine, University of Calgary, Calgary, Alberta, Canada

Background: Dermatomyositis (DM) is a major form of autoimmune myositis (AIM). The characteristic DM rash (Gottron's papules, heliotrope rash) and perifascicular atrophy (PFA) at muscle biopsy are regarded as diagnostic. However, new concepts are challenging the definition of DM. A modified Bohan and Peter clinical classification (mcB&P) of AIM was proposed. In the mcB&P, overlap features in presence of myositis allow a diagnosis of overlap myositis (OM), irrespective of the presence or absence of the DM rash or PFA. Therefore, our objective was to further differentiate DM from OM.

Methods: Using the mcB&P, we performed a longitudinal study of 100 AIM patients, including 44 patients with a DM phenotype, defined as DM rash, and/or DM-type calcinosis and/or PFA at biopsy. Overlap features, DM rash course, adermatopathic DM (aDM), DM-specific and overlap autoantibodies, cancer and survival were evaluated.

Results: Two subsets were identified in patients with a DM phenotype: pure DM (n=24) and OM with DM features, or OMDM (n=20). In pure DM, the rash of DM was the first disease manifestation, was always present at the time of myositis diagnosis, and was chronic and associated with a high cutaneous score. Concurrent heliotrope rash and Gottron papules (PPV 91%), as well as the V-sign and/or shawl sign (PPV 100%), were diagnostic of pure DM. Anti-Mi-2, anti-MJ and anti-p155 autoantibodies were restricted to pure DM (PPV 100%) and present in 50% of patients. 21% of patients had cancer. Fifteen-year survival was excellent (92%). In contrast, in OMDM the first manifestation was proximal muscle weakness or other skeletal muscle-related complaints. The DM rash appeared at diagnosis or followup and was associated with a low cutaneous score. aDM, absent in pure DM, predicted OMDM (PPV 100%). Autoantibodies, found in 70% of patients, included anti-Jo-1, anti-PL-7, anti-PM-Scl, anti-U1RNP and anti-U5-RNP. OMDM was not associated with cancer but 15-year survival was only 65%.
PFA occurred as commonly in OMDM (n=6/20 patients, 30%) as in pure DM (n=4/24, 17%). These 6 OMDM patients had aDM at the time of myositis diagnosis. Only one of them developed a DM rash at follow-up, emphasizing the lack of specificity of PFA for pure DM.

Conclusion: Using the mcB&P allowed identification of OMDM, a new clinical subset of OM. Furthermore, identification of OMDM allowed in turn recognition of pure DM as a new entity, distinct from OMDM or from OM without DM features. However, the absolute specificity of a DM rash and PFA for the diagnosis of pure DM was lost.