Correspondence on 'EULAR/ACR classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups'

The publication of the European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) 2017 classification criteria for adult and juvenile idiopathic inflammatory myopathy is a landmark with 186 citations as of date.[1] Patients with dermatomyositis (DM) and polymyositis (PM) differ with respect to clinical features, autoantibody profile and treatment response and have been subgrouped for a long time now.[2]

We studied 26 consecutive adult patients over 1 year seen in a tertiary teaching hospital in south India, satisfying either the 2017 criteria or 1975 Bohan-Peter criteria for idiopathic inflammatory myositis (IIM). Patients were subclassified using the 2017 criteria subclassification tree. Those with typical skin rash (heliotrope, Gottron papule or Gottrons sign) were classified as DM (n=14) and the rest as PM (n=12). There were no patients satisfying the criteria for IBM. Overlap myositis was excluded.

As is evident from table 1; seven patients in the PM group had non-classic skin rash (generalised erythematous rash or malar rash). These patients had received a bedside diagnosis of DM. Five of these patients had DM-associated myositis-specific antibodies (MSA) in serum (four anti-Mi2, one anti-NXP2) and perifasicular atrophy on muscle biopsy, another characteristic features of classical DM (specificity >90%, sensitivity 30%–50%).

Using the International Myositis Classification Criteria Project[1] database, the 2017 criteria classified 7% of 214 patients from the DM subgroup as PM. The agreement between physician subtype diagnosis and the 2017 subclassification tree at a probability of 55% and 90% was 0.89 and 0.94, respectively, being the lowest for DM compared with other subtypes.[1] Cutaneous findings of heliotrope, Gottron's papule or Gottron's sign have been assigned a higher weightage in the 2017 criteria and are also the only cutaneous findings considered for differentiating between DM and PM in the subgroup classification tree. This is justified because for pure DM, the positive predictive value of concurrent heliotrope rash and Gottron's papules of 91% rises to 100% with the presence of V-sign and/or shawl sign.[3] It is however worth noting that the sensitivity for Gottron's papule at 60%–80% and heliotrope rash at 30% for diagnosis of DM is low. Patel *et al* retrospectively evaluated the likelihood of the skin variables included in the EULAR/ACR criteria in classifying patients with amyopathic DM (ADM) and found that 26.3% of ADM would not meet the suggested 55% minimum probability cut-off to be classified as IIM on the basis of the EULAR/ACR criteria. Also 6% of ADM did not have any of the three skin variables. They suggested that subtyping of ADM can be improved by expanding the skin variables included.[4]

In the Euro myositis registry cohort (n=3067), a DM-specific rash was seen in 3.4% of patients classified as PM using the Bohan Peter IIM classification criteria.[5]

When patients in our study were subclassified using existing 2017 criteria, there was weak agreement between physician and ACR/EULAR criteria (Cohens kappa coefficient=0.43).The lower coefficient of agreement between physician diagnosis (considered gold standard) and 2017 criteria in our cohort most likely suggest ethnic/geographic variation in prevalence of classic DM cutaneous features. Addition of MSA positivity and perifascicular atrophy (PFA) on muscle biopsy to the EULAR/ACR criteria in our cohort lead to moderate agreement between physician and ACR/EULAR criteria (Cohens kappa coefficient=0.77).

While discussing the 2017 EULAR/ACR criteria,[1] the authors do mention that 'a future update of the EULAR/ACR classification criteria should include the more recently identified MSA'. We suggest that just addition of non-classic cutaneous findings at the node of the subclassification tree would substantially improve the sensitivity of the criteria for subtyping patients of DM across different populations.

Table 1 Characteristics of DM and PM subgroup as per ACR/EULAR criteria

	DM (n=14)	PM (n=12)
Mean age in years±SD	46.3±12.8	40.3±10.8
Median time to diagnosis in months (IQR)	8 (6.2–9.7)	2 (1–3)
Female:male	11:3	6:6
Organ system involved	14	12
Muscle	14	7
Cutaneous	13	11
Constitutional	11	6
Skeletal	4	10
Gastrointestinal	8	8
Pulmonary	5	3
Severity of myositis at onset		
Mild	2	1
Moderate	0	0
Severe	12	11
Cutaneous manifestations		
Heliotrope rash	5	na
Gottrons papule	4	na
Gottrons sign	11	na
Non-specific rash	6	7

ACR/EULAR, American College of Rheumatology/European League Against Rheumatism; DM, dermatomyositis; na, not applicable; PM, polymyositis.

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