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BRIEF REPORT

Comparison of the 2017 EULAR/ACR criteria with Bohan and Peter criteria for the classification of idiopathic inflammatory myopathies

Benzeeta Pinto[1] · Ramya Janardana[1] · Raghunanadan Nadig[2] · Anita Mahadevan[3] · Anusha S. Bhatt[3] · John Michael Raj[4] · Vineeta Shobha[1]

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Abstract

Bohan and Peter is the oldest criteria for the classification of idiopathic inflammatory myopathies (IIM). Recently, 2017 EULAR/ACR criteria were introduced which were validated against a control group. The objective of this study was to assess the performance of the 2017 EULAR/ACR criteria in retrospective cohort of adult and juvenile idiopathic inflammatory myopathies and compare with Bohan and Peter criteria. This was a retrospective study of patients clinically diagnosed to have IIM in a tertiary care center in the last 10 years. Only patients with a minimum follow-up of 6 months and response to steroids and immunosuppression were included in the study. Performance of both the criteria in the cohort was assessed and compared with clinical diagnosis. Hundred and eleven patients (87 females) were included in the study. Eleven patients had juvenile onset. Ninety-three patients (83.8%) were classified as probable/definite myositis using the Bohan and Peter criteria. Eighty-nine (80.2%) patients were classified as having probable/definite inflammatory myositis using the new criteria. Agreement between the two criteria was weak in our cohort (κ-0.331). Complete details of muscle biopsy were available in 52 patients. In this subgroup, 96% were classified by Bohan and Peter and 80.8% by EULAR/ACR criteria. Bohan and Peter classified 73% and EULAR/ACR 82% of patients when biopsy was excluded ($n = 111$). Both criteria classified over 90% of the patients with dermatomyositis. Forty-two patients were clinically diagnosed as polymyositis, of these 32 patients had myositis overlap syndrome. Bohan and Peter classified 66.7% and EULAR/ACR classified 64.3% in this subset. Bohan and Peter criteria had high sensitivity in the presence of muscle biopsy compared with EULAR/ACR. The performance of the EULAR/ACR criteria was similar to Bohan and Peter in the absence of muscle biopsy. Both criteria had poor sensitivity in polymyositis.

Keywords 2017 EULAR/ACR classification criteria · Bohan and Peter · Inflammatory myopathy

Introduction

Inflammatory myositis is a heterogeneous disease with well-defined subsets. The classification of IIMs has been a subject of debate for decades [1, 2]. In 1975, Bohan and Peter published their criteria for classification of IIM. Although these criteria have not been validated, they continue to be the most widely used criteria for classification of myositis [3, 4]. These criteria have been criticized due to various limitations including inclusion of non-specific myositis features (electromyogram), lack of specific description of skin rashes, muscle biopsy features, and well-defined exclusion criteria. Many modifications have been proposed to these criteria but none of these are validated against a control group [2]. In 2017, as a part of a collaborative effort between American College of Rheumatology and European League Against Rheumatism, new classification criteria were proposed which were validated in 976 IIM patients and 624 non-IIM controls. These criteria were found to have a sensitivity of 93% and specificity of 88% including muscle biopsy features. The sensitivity and specificity were slightly lower without biopsy (sensitivity and specificity 87% and 82%, respectively) [5,

✉ Vineeta Shobha
vineeta_shobha@yahoo.co.in

[1] Department of Clinical Immunology and Rheumatology, St John's Medical College, Sarjapur Road, Bengaluru 560034, India

[2] Department of Neurology, St John's Medical College, Bengaluru, India

[3] Department of Neuropathology, National Institute of Mental Health and Neurosciences (NIMHANS), Bengaluru, India

[4] Department of Biostatistics, St John's Medical College, Bengaluru, India

6]. The performance of these criteria was studied by Hočevar A et al. [7] in 95 patients of inflammatory myositis. In this cohort, muscle biopsy was available in 91.6%. The sensitivity was lower (78.9%) than in the validation study. We conducted this study to compare the performance of the EULAR/ACR 2017 criteria with Bohan and Peter criteria in our cohort of clinically diagnosed IIM.

Methods

This was a retrospective study conducted in a tertiary care referral center. All patients clinically diagnosed to have IIM in the last 10 years and with a follow-up of at least 6 months and response to steroids and immunosuppression were considered for the study. The case details were reviewed by three rheumatologists (BP, RJ, and VS) and one neurologist (RN) and a consensus was reached. Data was derived from structured proformas being maintained prospectively for the cohort. Additional details were derived from inpatient and outpatient records as applicable. Patients with insufficient data to enable classification were excluded from the study. We included muscle biopsies only when full details were available. Muscle biopsies were studied on paraffin sections and cryosections and enzyme histochemistry was performed on all biopsies. Testing for antinuclear antibody (ANA) was done using indirect immunofluorescence (IIF). Screening for myositis specific antibodies (MSA) and myositis-associated antibodies (MAA) was done by immunoblot. Anti Ro, anti Ro52, U1RNP PM-Scl, and Anti Jo1 were the MAAs and MSA included in the immunoblot. Electromyogram was considered as positive in the presence of short, small polyphasic motor units, with insertional activity, positive sharp waves, and fibrillations. Diagnosis of myositis overlap syndrome was based on suggestions of Troyonov et al. [8]. Diagnosis of Systemic lupus erythematosus (SLE) was according to ACR or SLICC criteria [9]. Systemic sclerosis was diagnosed based on ACR/EULAR criteria, and mixed connective tissue (MCTD) was diagnosed based on Kahn Criteria [10, 11]. Patients who did not fulfill any particular criteria but had at least one clinical (arthritis, raynaud's, sclerodactyl, puffy fingers, interstitial lung disease) and one serological feature (ANA/ MAA) of connective tissue disease (CTD) were diagnosed as undifferentiated CTD (UCTD).

Statistical tests were performed using the Statistical Package for the Social Sciences (SPSS 18.0, 2009; SPSS Inc., Chicago, IL, USA). Quantitative variables were described as mean ± standard deviation and qualitative variables as number (%). Agreement between classification criteria sets was evaluated using Cohen's kappa coefficient (κ). Ethical clearance was obtained from the Institutional Ethics Committee and guidelines set by ICMR 1994 and Helsinki declaration (modified, 1989) were adhered to.

Results

A total of 111 patients with IIM were included in this study. Eleven patients had a juvenile onset. (10 Juvenile Dermatomyositis, one juvenile onset overlap myositis). The clinical features are summarized in Table 1. Median follow-up duration of the cohort 24 months (IQR 55.5 months). Muscle biopsy was available in 52 patients. EMG was available in 70 patients and was suggestive of myositis in 52 patients. Immunoblot was available in 86 patients and 43 patients were positive for at least one MAA (Table 1).

Sixty-nine patients had rashes suggestive of dermatomyositis (DM) as per the description by Bohan and Peter (56 DM, 3 ADM, and 10 JDM). Only 49 of these had any one of the rashes included in the present criteria (Table 1). The other rashes that were seen in our patients include V sign, shawl sign, holster sign, periorbital edema without erythema, and erythematous rashes on extensor aspect of hand, forearms, and arms.

Myositis overlap syndrome was diagnosed in 32 patients, of these two had overlap with systemic lupus erythematosus, three had mixed connective tissue disease, 14 had systemic sclerosis and 13 patients had undifferentiated connective tissue disease. All patients with UCTD had a positive ANA/MAA in addition to at least one clinical feature other than myositis.

Ninety-three patients (83.8%) were classified as probable/definite myositis using the Bohan and Peter criteria. Eighty-nine patients (80.2%) were classified as having probable/definite IIM using the new criteria (score ≥ 5.5 without biopsy or ≥ 6.7 with biopsy). The agreement between the two classification criteria was weak (κ-0.331) (Table 2).

Muscle biopsy was performed in 72 patients but we were able to trace the full details only in 52 patients. Hence, only these were included for analysis. Fifty patients were classified by Bohan and Peter criteria and 42 by EULAR/ACR criteria (Table 2). Forty-three (82.7%) biopsies had at least one of the features included in the EULAR/ACR criteria (Table 1). Only these were taken as positive in the Bohan and Peter criteria. In patients with positive biopsies ($n = 43$), all 43 (100%) were classified by Bohan and Peter and 37 (86%) by EULAR/ACR criteria.

Discussion

In our cohort of 111 patients, only 80.2% were classifiable by the EULAR /ACR criteria. The most important area of discrepancy was the muscle biopsy. Bohan and Peter criteria had a very good sensitivity in patients with biopsy. The EULAR/ACR criteria performed similarly with and without biopsies. Forty-three of 52 biopsies were reported as positive by the pathologist and had at least one feature included in EULAR/ACR criteria. The new criteria include only four features with highest weightage given to rimmed vacuoles followed by perifascicular atrophy. Most of our patients had perimysial/

Table 1 Clinical features of patients with idiopathic inflammatory myositis

Age (at diagnosis)	38.22 years	ANA positivity	63 (56.7%)
Sex (female: male)	3.6:1	Anti Jo1 positivity ($n = 86$)	5 (5.2%)
Median follow-up duration	24 months	MAA ($n = 86$)	43 (50%)
Proximal muscle weakness	108 (97.3%)	Rnp/Sm	26 (30.2%)
		Ro52	26 (30.2%)
		PmScl	4 (4.6%)
Raynaud's	22 (19.8%)	Elevated muscle enzymes (CK /LDH/AST/ALT)	100 (90%)
Dysphagia	36 (32.4%)	Cutaneous involvement (s/o DM)	69 (62.1%)
Arthralgia/arthritis	73 (65.7%)	Heliotrope	38 (34.2%)
ILD	10 (9%)	Gottron's papules	22 (19.8%)
Clinical diagnosis		Gottron's sign	12 (10.8%)
Dermatomyositis (two cancer associated)	56 (50.4%)	Muscle biopsies	52
Amyopathic DM	3 (2.7%)	Positive for IIM	43
Juvenile dermatomyositis	10 (9%)	Endomysial inflammation	5
Polymyositis	42 (37.8%)	Perimysial/perivascular inflammation	38
Myositis overlap syndrome	32 (76.2%)	Perifascicular astrophy	19
Cancer-associated PM	1 (0.9%)	Rimmed vacuoles	0
Others	9 (8.1%)		

DM, dermatomyositis; *ILD*, interstitial lung disease; *MAA*, myositis-associated antibodies

perivascular inflammation which is given low weightage in the new classification. Muscle biopsy may not be positive in all patients to be taken as the gold standard [12, 13]. However, it does serve to exclude other muscle diseases and strengthens the diagnosis of IIM [14].

Both the criteria had poor sensitivity in patients with polymyositis. On reviewing the patients classified as polymyositis, we found that 32 of the 42 patients with PM had overlap myositis. Nineteen patients had well-defined overlap with a CTD (18 adults, 1 juvenile). Thirteen patients had UCTD. These patients are classified by EULAR/ACR criteria as polymyositis. The existence of polymyositis has been under a lot of debate [15]. In our cohort, only nine patients with polymyositis did not have any other features to suggest CTD. Diagnosis of inflammatory myositis in a patient with a CTD does not require an extensive work up. In this subset proximal muscle weakness, autoantibody positivity with elevated muscle enzymes may suffice for a diagnosis. A clinicoserologic approach has previously been suggested by Troyanov et al. [8]. They used this approach on 100 patients with myositis and found the frequency of polymyositis reduced from 45% as diagnosed by Bohan and Peter to 9% with the new classification. A similar observation has also been made by van der Meulen et al. [16]. CTD myositis is a distinct entity and needs to be better defined and differentiated from polymyositis.

Bohan and Peter criteria was able to classify more patients of DM than the new criteria. While the former criteria include all rashes suggestive of DM, the latter includes only heliotrope rash, Gottron's papules and Gottron's sign. These are difficult to identify in dark skinned individuals. The subclassification scheme in the EULAR/ACR is based entirely on the presence or absence of these three skin findings leading to misclassification of some DM patients as PM. Perifascicular atrophy which is diagnostic of DM on biopsy is not included in the subclassification scheme.

Table 2 Classification using Bohan and Peter and 2017 EULAR/ACR criteria

	Bohan and Peter *n* (%)	EULAR/ACR *n* (%)
Overall ($N = 111$)	93 (83.8)	89 (80.2)
Without biopsies (N = 111)	81 (73)	91 (82)
With biopsies ($N = 52$)	50 (96.2)	42 (80.8)
DM ($N = 56$)	55 (98.2)	52 (92.9)
ADM ($N = 3$)	0	1
PM ($N = 42$)	28 (66.7)	27 (64.3)
JDM ($N = 10$)	10 (100)	9 (90)

DM, dermatomyositis; *ADM*, amyopathic dermatomyositis;*JDM*, juvenile dermatomyositis; *PM*, polymyositis

Our study has several limitations. Data was analyzed retrospectively. Muscle biopsy data was available for only half of the patients. We did not screen specifically for MSAs other than Jo1. Due to lack of an appropriate control group, we were unable to analyze the specificity of these criteria in our cohort. We did not include patients with IBM as they were not being followed-up in the unit. We did not have any patients with necrotizing myositis. It is possible that these patients have been missed or diagnosed as polymyositis.

In conclusion, both EULAR/ACR and Bohan and Peter criteria had a lower sensitivity in our cohort than reported previously. The EULAR/ACR criteria performed similarly with and without muscle biopsies. Since IIM is a heterogeneous disease, a single classification criterion may not suffice for all disease subsets. There is no clarity on the classification of overlap myositis which was the second largest subset of IIM in our cohort.

Compliance with ethical standards

Ethical clearance was obtained from the Institutional Ethics Committee and guidelines set by ICMR 1994 and Helsinki declaration (modified, 1989) were adhered to.

Conflict of interest The authors declare that they have no conflict of interest.

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