ORIGINAL ARTICLE

Idiopathic Inflammatory Myopathy: A Hospital-based Case Review in the State of Perak, Malaysia.

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Submitted: 20/05/2025. Revised edition: 05/08/2025. Accepted: 24/09/2025. Published online: 01/11/2025.

Abstract

Background: Idiopathic inflammatory myopathy (IIM) is a rare condition characterized by proximal muscle weakness and myositis. It includes dermatomyositis (DM), polymyositis (PM), inclusion body myositis (IBM), and juvenile dermatomyositis (JDM). They may share clinical and histological features but differ in underlying pathophysiology. **Objective:** To assess the diagnostic probabilities, and clinical characteristics of IIM. **Methods:** Clinical data from 50 IIM patients were retrospectively reviewed from Rheumatology and Dermatology clinics across four centres in Perak, Malaysia (2013–2023) and were evaluated using the 2017 The European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria for adult and juvenile IIM. **Results:** According to the EULAR/ACR criteria, 98.2% of cases were classified as definite IIM (mean score 10.6), 74.1% as probable (mean score 6.6), and 47.8% as possible (mean score 6.4). IIM was excluded in 36% of patients (mean score 3.9; probability 20.8%). Eighty-four percent had DM, 8% had ADM or PM, with a mean age of 45.9±18.1 years. 95.6% were diagnosed within a year of symptom onset, and 72% showed proximal myopathy, significantly common in DM (P=0.001). Ethnic breakdown showed 46% Chinese, 42% Malay, and 8% Indian patients, with Malays developing DM at a younger age (39.8±16.9 years). The shawl sign and telangiectasia were more common in Chinese (P=0.018) and Malay (P=0.023) patients, respectively. **Conclusion:** The EULAR/ACR scoring system effectively identified IIM subgroups. Despite varying probabilities, cases of both DM and ADM were classified into definite, probable, and possible categories, with or without biopsy data. DM was more prevalent than ADM, primarily affecting Chinese patients, while Malays had earlier onset.

Keywords: *Amyopathic dermatomyositis, clinical characteristics, dermatomyositis, EULAR/ACR criteria, idiopathic inflammatory myopathy, polymyositis.*

Introduction

Idiopathic inflammatory myopathy (IIM) is a rare group of chronic immune-mediated inflammatory myopathies that characteristically present with myopathy or myositis and a spectrum of clinical manifestations. It is classified into dermatomyositis (DM), amyopathic dermatomyositis (ADM), polymyositis (PM), inclusion body myositis (IBM), and juvenile dermatomyositis (JDM). The exact pathogenetic process of IIM is still unknown, but it is believed to involve genetic and environmental factors, including HLA alleles and viral triggers [1]. They may share clinical features but may be distinguished by certain specific presentations and histopathology findings. Nevertheless, cutaneous involvement is more common in DM. The patients may present with classical cutaneous lesion alone or with concomitant progressive myopathy and may develop complications such as pulmonary fibrosis and malignancy. Cutaneous manifestations may precede myositis by three to six months in 30-50% of patients, and 10% may present with myositis prior to the onset of cutaneous lesions [2,3]. DM with predominantly cutaneous phenotype without myopathy is classified as clinically amyopathic DM (CADM) which includes the amyopathic DM (ADM) and hypomyopathic DM (HDM) [3, 4, 5]. Due to the heterogeneity of IIM, the Bohan and Peter and the 2017 American College of Rheumatology (ACR)/European League Against Rheumatism (EULAR) criteria in the classification of myositis are widely used [6,7]. These criteria classify IIM into 'definite', 'probable' and 'possible' categories. Developed by EULAR/ACR in 2017, they enable better identification of IIM subgroup including juvenile IIM.

DM commonly affects middle-aged adult female. The prevalence and incidence of DM, however, vary widely with different geographical distributions and research methodology from previous reports [8,9,10]. Myositis-specific antibodies (MSA) and myositis-associated antibodies (MAA) such as the anti-melanocyte differentiation-associated gene 5 (MDA5) and anti-transcriptional intermediary factor 1 gamma (TIF1-γ) antibodies are important markers in determining the risk of DM or ADM developing interstitial lung disease (ILD) and malignancy.

Methods

This is a descriptive study analysing the clinical presentation of DM and PM patients in Perak state, Malaysia. The DM/PM patients in this study were ascertained based on a confirmed diagnosis by the rheumatologist according to the 2017 European League Against Rheumatism/
American College of Rheumatology (EULAR/ACR) classification criteria for adult and juvenile idiopathic inflammatory myopathies (IIM) [7]. Subgroups of IIM were determined by using scoring systems according to the criteria. Since DM is a considerably rare disease, all patients with confirmed diagnoses by the rheumatologist in the 10-year study period between 2013 and 2023 and currently still on follow-up were included in this study. Patients with myositis secondary to other causes such as drug-induced, electrolyte derangement (hypokalaemia), diabetes, thyroid disease, and neurological disorders as well as patients with no documented clinical and laboratory evidence of DM or PM were excluded.

Clinical data were retrieved from the patient medical records. The diagnostic criterion for DM includes classical cutaneous lesions, skin biopsy, clinically significant myopathy evaluated by electromyography (EMG), muscle enzymes i.e., creatine kinase ([CK], 24-173 IU/L), lactate dehydrogenase ([LDH], 20-350 IU/L), alanine aminotransferase ([ALT],10-36 IU/L), aspartate aminotransferase ([AST], 10-36 IU/L), muscle biopsy, myositis-specific or myositis-associated autoantibodies (MSA, MAA). The MSA profiles (anti-Jo-1 (histidyl-), anti-PL-7 (threonyl-), anti-PL-12 (alanyl-), anti-EJ (glycol-), anti-OJ (isoleucyl-tRNA synthetase), anti-KS (asparaginyl-), anti-SRP (signal recognition particle), anti-Mi-2, anti-TIF1-γ, anti-SAE (small ubiquitin-like modifier activating enzyme), anti-NXP-2 (nuclear matrix protein 2), anti-MDA5

(anti-melanoma differentiation-associated protein 5), and MAA profiles (anti-PM-Scl 75, anti-PM-Scl 100, anti-Ku, and anti-SS-A/Ro-52 kDa) were done by outsourcing the tests due to unavailability in public hospitals. Data extracted from the patient's clinical notes were demographics, cutaneous features, comorbidities (including malignancy, ILD), muscle enzymes, erythrocyte sedimentation rate ([ESR], <20 mm/1st hour), high sensitivity C-reactive protein ([CRP], <5 mg/L), serological and immunological markers (antinuclear antibody [ANA], extractable nuclear antigen panel [ENA], MSA, MAA), skin and muscle biopsy, electromyography (EMG), and imaging report (magnetic imaging resonance, MRI and high-resolution computed tomography (HRCT) if available.

Statistical analysis

Statistical analysis was performed using RStudio version 2023.06.1+524 (Mountain Hydrangea). Descriptive analysis was performed and presented as frequencies and proportions. The Fisher exact test was used to determine the significant association between the exploratory variables (i.e., the demographic and the clinical characteristics) and the type of myositis (i.e., DM or ADM). A p-value of less than 0.05 was considered statistically significant.

Results

Table 1 and 2 show distinct clinical features between Dermatomyositis (DM) and Amyopathic Dermatomyositis (ADM), with DM typically presenting with more significant muscle involvement, higher CK levels, positive ANA, and a greater risk for associated malignancy and ILD. Overall, the clinical features vary across gender and ethnicity in IIM patients. MSA and MAA show a variety of autoantibodies across ethnic groups and genders.

Most patients were diagnosed in adulthood, with a mean age of 45.9 years (SD = 18.1). However, seven patients were diagnosed during childhood (under 18 years of age). Among the DM (Dermatomyositis) patients, the majority were female, with a male-to-female ratio of 1:4. The largest proportion of patients were of Chinese descent, accounting for nearly half of the total (46.0%), followed by Malays at 42.0%, Indians at 8.0%, and others at 4.0%. There was no significant difference in the cutaneous features between DM and ADM. Shawl sign was the most common cutaneous lesion observed in Chinese ethnic group (P=0.018). There was a statistically significant difference in mean CK levels between DM, ADM and PM (P=0.016).

The probability scores for the IIM subgroups in Table 3 suggest that DM patients are more likely to meet the Definite IIM criteria, followed by ADM patients. There is also a notable portion of DM patients (n = 14) classified as non-IIM, indicating some overlap or uncertainty in the classification. PM patients are less represented in the table, but those included are close to the Definite IIM category. The EULAR/ACR classification criteria help stratify patients based on the likelihood of having IIM, with most patients falling into the Definite or Probable IIM categories.

Table 4 shows that the Definite and Probable IIM groups predominantly received a mix of traditional DMARDs and steroids. Non-IIM patients received more specialized treatments like rituximab and IV immunoglobulin. The Possible group received fewer treatments, with no patients in this category receiving multiple DMARDs, steroids, or NSAIDs. This treatment approach reflects a focus on controlling the autoimmune response in confirmed cases of IIM while more targeted therapies are employed in ambiguous or non-IIM diagnoses.

Discussion

The incidence of IIM has been reported as 11 per million person-years (10 for men and 13 for women) and a prevalence of 14/100,000 [11]. Whereas the global incidence and prevalence of DM vary widely from 1.97 to 21.5 per 100,000 population respectively [4,8,10,12].

In this study, the 2017 EULAR/ACR criteria for IIM were successfully used to classify IIM into subgroups according to the scores based on the suggested domains in the criteria. The probability of being classified as 'definite' for DM, ADM, and PM were 98.2%, 99.5%, and 97%, corresponding to scores of 10.4, 10.6, and 10.2, respectively. These results were consistent with the EULAR/ACR criteria, which define 'definite IIM' as a probability of ≥90%, with a score of ≥7.5 (≥8.7 with muscle biopsy). For 'probable' IIM, the probabilities for DM and ADM were 72.3% and 87.8%, corresponding to scores of 6.5 and 7.3, respectively, consistent with the criteria, which set the probability cut-off at 55%, corresponding to a score of 5.5 (6.7 with muscle biopsy). For DM, the probability of being classified as 'possible' was 47.8%, corresponding to a score of 6.4, which falls within the probability range of ≥50% to <55%.

IIM was ruled out in 18 patients, as their calculated probabilities were 21.2% and 17.3%, with scores of 3.9, 4.2 and 3.6 for DM, ADM and PM, respectively. These scores were below the 50% probability threshold, with scores lower than 5.3 (<6.5 with muscle biopsy) according to the criteria. These patients were categorized as non-IIM due to insufficient evidence in the criteria. However, despite the absence of certain criteria features, these patients were still treated as IIM cases, as they presented other signs such as myopathy, shawl sign, poikiloderma, and arthritis, which are not included in the scoring criteria. Therefore, while their probability scores did not meet the criteria for IIM, clinical judgment led to their continued treatment.

Females were found to have a greater risk of developing IIM, with a 2:1 female-to-male ratio, and the mean age of diagnosis among adults is between 40 and 60 years in other studies [4,13,14]. However, in this study, although a female preponderance is consistent with previous reports, the ratio was higher i.e., 4:1, which is almost similar to that in the study by Dourmishev i.e., 3.75:1[15]. This discrepancy is likely attributable to the multiethnic nature of this cohort. The mean age at diagnosis were 45.9±18.0 year for DM in this cohort, consistent with most reports in the literature [16, 17]. The age onset for JDM was 11 years old. DM was found to be common in Chinese ethnic group which is consistent with the study by Tong in Malaysia although the number was very small i.e., only eight patients [18]. The mean duration from birth to the onset of development of cutaneous manifestations was 11.2±6.4 years in JDM. The majority of patients were diagnosed in the same year when they developed the symptoms.

The term ADM was first coined by Carl Pearson [19] and recognized as a subtype of DM characterized by biopsy-confirmed cutaneous lesions, which are the hallmarks of DM, occurring for six months or longer with no clinical evidence of proximal myopathy and no serum muscle enzyme abnormalities [20]. It had been regarded and classified as a distinct entity among the spectrum of IIMs by Hoogendijk *et al*. in 2004 [21]. The incidence of ADM had been reported at 10% to 20% [4, 22] and was higher in Asian population (10% of DM cases) [5,23]. ADM is more often associated with fatal rapidly progressive interstitial lung disease (RP-ILD) and malignancy as compared to DM [4]. One ADM patient in this cohort with the presence of anti-MDA-5 antibodies had developed RP-ILD. This patient remains in clinical remission without development of malignancy over more than a three-year period.

In this study, the classical cutaneous manifestations i.e., heliotrope rash, Gottron's papules, poikiloderma, and photosensitivity occurred in both DM and ADM. Among the cutaneous lesions, Shawl's sign was commonly seen in Chinese ethnic group in this study (P = 0.018) compared to other ethnic groups. Although JDM and DM share common characteristics, cutaneous lesions especially calcinosis with skin ulceration were commonly found in JDM [24].

However, in the cohort of this study, these skin lesions were not documented in detail in the medical record.

Antinuclear antibody (ANA) is positive in more than 50% in confirmed cases of DM from previous report [25]. In this study, ANA was positive in 58% of patients. The relevancy of ANA in DM is still uncertain. There was no significant association between positive ANA with the cutaneous lesions of DM or the risk of malignancy in this study, similar to findings by Hoesly *et. al.* [25]. Four patients with DM were overlapped with systemic lupus erythematosus with positive anti-dsDNA.

DM has been recognized as paraneoplastic in established studies, estimated between 7% to 33% of DM patient particularly of adult onset [26,27]. MSA has been considered as important biomarker which provides guidance to the classifications of the disease and organ-specific involvement as well as the risk of malignancy [28]. However, it has been shown to be a poor diagnostic tool in DM patients [29] a finding supported by the present cohort. The presence of MSA i.e., anti-NXP-2 or anti-TIF1-γ antibodies, were found to be associated with the risk of malignancy [30, 31]. The prevalence of anti-TIF1-γ antibody is estimated to be between 18-35% in JDM [32]. The anti-TIF1-γ was positive in one of the juvenile-onset ADM and two DM patients in this study. Presence of anti-TIF1-γ in JCADM has been shown to have no association with risk of malignancy compared to adult DM or ADM [33] and the cutaneous lesions may take longer time to resolve. Meanwhile, among patients with the MSA test performed in this cohort, anti-Mi-2 autoantibodies were present in five (10%) of the CDM cases and none of them demonstrated any evidence of malignancy. The prevalence of anti-Mi-2 autoantibodies is estimated from 2% to 38% in adults with DM [26]. In contrast to anti-NXP-2 or anti-TIF1-γ, patients with anti-Mi-2 autoantibodies in general, had shown a lower risk of malignancy and a better response to treatment with better prognosis [34]. Adult DM had been reported to be commonly associated with occurrence of malignancy and interstitial lung disease (ILD) compared to JDM [35]. These complications of adult DM (ILD 83.3%, malignancy 10%) in this study had demonstrated similar incidence although the number was small and limited. However, among the patients with malignancy in this study who had the MSA test done, the antibodies were found to be negative.

In this study, skin biopsies were performed in 34% of patients as compared to muscle biopsies (20%). The majority of patients did not consent to or were not keen on muscle biopsy due to logistic problems. For those who had skin or muscle biopsies, the results were consistent with dermatomyositis and inflammatory myopathy changes, respectively. Electromyography (EMG) study findings were indicative of the disease in 24% of patients who underwent the procedure. However, EMG findings in DM are neither specific nor sensitive and may be negative or normal in 11% of patients [12]. A study by Constantinides *et al* revealed that muscle biopsy yielded excellent diagnostic accuracy compared to EMG (90% vs 70% respectively) [36]. In this study, the discordance between EMG and muscle biopsy findings was likely due to a significant number of patients who did not undergo these investigations and for those who had these tests done, there were normal biopsy findings despite myopathic EMG changes.

Corticosteroid and immunosuppressant had been the mainstay of treatment in all DM patients at any time period during the course of the disease, similar to all reviewed studies [37]. Anti-malarial drug (hydroxychloroquine) in combination with corticosteroid were used in majority of patients. Azathioprine (AZA) and methotrexate (MTX) were prescribed as corticosteroid-sparing agent and for patient who failed to respond to corticosteroid alone. MTX was more often used compared to AZA. It is evident that methotrexate is an effective corticosteroid-sparing agent to

treat the cutaneous lesions in DM [38]. Pulse intravenous (IV) cyclophosphamide (CYC) monthly was given to patients who were refractory to other immunosuppressants including corticosteroids especially those with ILD. Study by Shahin *et al* had indicated that CYC improves clinical response especially in refractory myositis [39]. Nine patients with severe cutaneous DM were administered with IV immunoglobulin (Ig) and better response to treatment was observed. However, due to its high cost, IVIg was only administered for short periods of time. Three patients who were resistant to steroid and other standard treatment were given rituximab (RTX).

Limitations of the study

This study's limitations are primarily due to the use of secondary data. A significant amount of missing data was encountered as information was extracted from medical records. Incomplete clinical and laboratory documentation further contributed to discrepancies in the study's results. Although myositis-specific antibodies (MSA) and myositis-associated antibodies (MAA) are crucial for diagnostic evaluation of idiopathic inflammatory myopathies (IIM), these tests have only been available in the Malaysian private hospitals over the past decade. Their high costs had made them inaccessible for many patients, with nearly half declining testing. As a result, only antinuclear antibody (ANA) and extractable nuclear antigens (ENA), which are available in public hospitals, were tested. Additionally, other essential ancillary tests such as muscle biopsy and MRI were also limited due to logistical challenges.

Conclusion

This study shows IIM is relatively uncommon in this region. The utility of the EULAR/ACR criteria is useful for diagnosing idiopathic inflammatory myopathies (IIM), even with its limitations. IIM should still be considered if clinical signs and lab findings suggest it—even if these features are not part of the diagnostic scoring system. DM was most common IIM subtype, more prevalent than polymyositis or inclusion body myositis.

Malay females were diagnosed with DM at a younger age, pointing to potential genetic or environmental factors. Additionally, DM was more common in the Chinese ethnic group, which suggests an ethnic predisposition. Rare complications such as ILD and malignancy were not commonly observed in the study population, indicating that these severe outcomes may not be as prevalent in this specific cohort.

These findings can help guide clinicians in recognizing DM in the Malaysian context and highlight the importance of early diagnosis, particularly in specific ethnic and gender groups. Further research may be needed to explore underlying reasons for the observed ethnic variations and the infrequent complications in this population.

Ethical approval

This study was approved by the Medical Research and Ethics Committee (MREC), Ministry of Health Malaysia (NMRR ID-23-01339-CWA) and the Universiti Kuala Lumpur Royal College of Medicine Perak Ethics Committee.

Authors' contributions

WS, ASSC, AFI, AH, ANZ, MAKHM, SKNAN, OPS, TJJ, LEL, NMH, PVRD, LHS responsible for conceptualizing, collecting clinical data; WS for report writing, and finalizing the manuscript; NAA for statistical analysis.

Acknowledgement

We want to express our gratitude to all healthcare providers involved in the care of this patient.

Conflict of interest

None.

Table 1. Demographic and clinical characteristics of classical DM and ADM (N=50)

		Total n (%)	DM n (%)	ADM n (%)	PM n (%)	*p*
Patients (n/%)		50 (100)	41(82.0)	5(10.0)	4 (8.0)	
Gender	Male	10 (20)	7(70)	2 (20)	1 (10)	0.305
	Female	40 (80)	34 (85.0)	3 (7.5)	3(7.5)	
Ethnicity	Chinese	23 (46)	20 (87.0)	2 (8.7)	1 (4.35)	0.709
	Malay	21 (42)	16 (79.2)	2 (9.5)	3(14.3)	
	Indian	4 (8)	3 (75.0)	1 (25.0)	0	
	Others	2 (4)	0	2 (100)	0	
Age at diagnosis (Mean ± SD)		45.9 (±18.1)	46.8 (±18.0)	45.8 (±20.5)	37.8 (±18.9)	
Age of Onset	< 18 years old	7 (14.0)	5 (71.4)	1 (14.3)	1 (14.3)	0.185
	18-39.9	9 (18.0)	7(77.8)	0	2 (22.2)	
	40 and above	34 (68.0)	29 (85.3)	4 (11.8)	1 (2.9)	
Duration from onset of symptoms to diagnosis	< 1 year	43 (95.6)	37 (86.1)	4 (9.3)	2 (4.6)	0.290
	1 to 2 years	2 (4.4)	1(50)	1 (50)	0	
Cutaneous manifestations	Gottron's papule	28 (56.0)	22 (78.6)	5(17.9)	1(3.6)	0.061
	Heliotrope	26 (52.0)	21 (80.8)	4 (15.4)	1(3.9)	0.306
	Shawl sign	14 (28.0)	12 (85.7)	2 (14.3)	0	0.502
	Alopecia	10 (20.0)	7 (70.0)	3 (30.0)	0	0.070
	V sign	7 (14.0)	5 (71.4)	2 (28.6)	0	0.220
	Poikiloderma	6 (12.0)	4 (66.7)	2 (33.3)	0	0.165
	Telangiectasia	6 (12.0)	4 (66.7)	2 (33.3)	0	0.165
	Mechanic's hand	1 (2.0)	1	0	-	-
Proximal myopathy		36 (72.0)	32 (88.9)	0	4(11.1)	**0.001**
Skin Biopsy	Done	17 (34.0)	14 (82.4)	3(17.7)	0	-
Muscle Biopsy	Done	10 (20.0)	9 (90)	0	1 (10.0)	
EMG	Done	**12 (24)**	11(91.7)	0	1(8.3)	
Laboratory:	Raised CK	33 (66.0)	29 (87.9)	-	4 (12.1)	**0.002**
	CK (Mean±SD)	3802.7(±6654.6)	3975.3(±6837.6)	113.2(±57.2)	6645.3(±8127.2)	**0.016**
	LDH(Mean±SD)	628.1 (± 610.1)	588.5(± 554.3)	372(±182.4)	2088	0.258
	AST (Mean±SD)	176.1 (± 269.0)	192.2 (±285.2)	17.5(±2.121)	118.25(± 147.2)	0.118
	ESR (Mean±SD)	36.5 (± 25.8)	36.7(±26.0)	27.4 (± 10.73)	45.7 ((± 38.6)	0.816
	Positive ANA	29 (58.0)	28 (96.5)	1 (3.5)	0	**0.001**
Interstitial lung disease (ILD)		6 (12.0)	5 (83.3)	1 (16.7)	0	0.717
Malignancy		5 (10.0)	4 (80.0)	0	1(20.0)	0.407

DM, dermatomyositis; ADM, amyopathic dermatomyositis; PM, polymyositis; EMG, electromyography; CK creatine kinase, ESR, erythrocyte sedimentation rate; ANA, anti-nuclear antibody; ILD, interstitial lung disease
P-value < 0.05 is considered significant.

Table 2. Clinical characteristics by major ethnic groups and genders.

Clinical characteristic	Gender n (%)		P	Ethnicity n (%)				P
	Male	Female		Malay	Chinese	Indian	Others	
Age at diagnosis (Mean±SD)	42.9±20.9	46.7±17.5	-	39.8±16.9	50.4±19.1	48.0±9.1	55.5±23.3	
Cutaneous lesions:								
Gottron's papule	7 (25.0)	21 (75.0)	0.263	11 (39.3)	14 (50.0)	2 (7.1)	1 (3.6)	0.955
Heliotrope	5 (19.2)	21 (80.8)	1.000	12 (46.2)	12 (46.2)	1 (3.9)	1 (3.9)	0.785
Shawl sign	3 (21.4)	11 (78.6)	1.000	2 (14.3)	8 (57.1)	3 (21.4)	1 (7.1)	**0.018**
Alopecia	0	10 (100.0)	0.179	6 (60.0)	3 (30.3)	1 (10.5)	0	0.546
V sign	2 (28.6)	5 (71.4)	0.616	1 (14.3)	4 (57.1)	2 (28.6)	0	0.100
Poikiloderma	0	6 (12.0)	0.327	3 (50.0)	2 (33.3)	1 (16.7)	0	0.546
Telangiectasia	0	6 (100)	0.327	4 (66.7)	0	1 (16.7)	1 (16.7)	**0.023**
Mechanic's hand	0	1 (100)	-	0	1	0	0	-
Laboratory:								
Raised CK	7 (21.2)	26 (78.8)	0.765	15 (45.5)	14(42.4)	3(9.1)	1(3.1)	0.821
ESR (Mean ± SD)	22.1(±19.7)	39.97(±26.1)	**0.021**	42.0(±28.6)	31.9(±25.3)	30.3(±11.9)	38.5(±9.2)	0.606
Positive ANA	3 (10.3)	26 (89.7)	**0.048**	12 (41.4)	13 (44.8)	2 (6.9)	2 (6.9)	0.498

CK, creatine kinase; ESR, erythrocyte sedimentation rate; ANA, anti-nuclear antibody;
P-value < 0.05 is considered significant.

Table 3. Classification of IIM according to EULAR classification criteria scoring.

IIM	n	Mean Score ± SD	Mean Probablity (%) ± SD	Diagnosis	n	Mean_Score ± SD	Mean Probability (%) ± SD
Definite_IIM	19	10.6 ± 2.08	98.2± 2.13	PM	1	10.2	97.6
				DM	16	10.7 ± 2.28	98± 2.28
				ADM	2	10.6	99.5
Probable_IIM	12	6.64 ± 0.71	74.1±10.5	PM	0		
				DM	11	6.58± 0.711	72.8 ± 10
				ADM	1	7.3	87.8
Possible_IIM	1	6.4	47.8	PM	0		
				DM	1	6.4	47.8
				ADM	0		
Non-IIM	18	3.86 ± 1.10	20.8± 15.6	PM	3	3.6±0.87	17.3 ± 13.2
				DM	14	3.89 ± 1.2	21.2 ± 16.9
				ADM	1	4.2	24.4

DM, dermatomyositis; ADM, amyopathic dermatomyositis; PM, polymyositis; IIM, Idiopathic Inflammatory Myositis.

Table 4. Treatment administered.

	Definite		Probable		Possible		Non-IIM	
	n	%	n	%	n	%	n	%
DMARDs								
Methotrexate	5	26.3	6	50	-	-	2	11.1
Cyclosporine	2	10.5	1	8.3	-	-	2	11.1
Hydroxychloroquine	9	47.4	10	83.3	1	100	7	38.9
Mycophenolate mofetil	2	10.3	1	8.3	-	-	1	5.6
Azathioprine	9	47.4	6	50	-	-	5	27.8
≥ 2 cDMARDs	8	42.1	8	66.7	-	-	4	22.2
≥ 3 cDMARDs	1	5.3	4	33.3	-	-	-	-
Steroids	19	100	11	91.7	-	-	15	83.3
NSAIDs	4	21.1	1	8.3	-	-	2	11.1
Combined treatment								
cDMARDs+Steroid	17	89.5	10	83.3	-	-	13	72.2
Other treatment								
Rituximab	2	10.5	-	-	-	-	1	5.6
IV Ig	1	5.3	1	8.3	-	-	7	38.9

Percentage per column total.
NSAIDs, non-steroidal anti-inflammatory drugs; cDMARDs, conventional disease modifying agent for rheumatic diseases; IV Ig, intravenous immunoglobulin.

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