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Idiopathic Inflammatory Myositis: A Hospital-based case review in the State of Perak, Malaysia

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INTRODUCTION

Idiopathic inflammatory myopathies (IIMs) are rare autoimmune conditions marked by muscle weakness and inflammation, with variable clinical features. Subtypes include dermatomyositis (DM), polymyositis (PM), inclusion body myositis (IBM), and juvenile dermatomyositis (JDM).

METHODOLOGY

This descriptive study reviewed records of patients diagnosed with DM/PM from 2013 to 2023 across four rheumatology clinics in Perak. The 2017 EULAR/ACR classification criteria were applied to assign probabilities—definite, probable, possible, or non-IIM—based on scores.

RESULTS

Among 50 patients, 82% had dermatomyositis (DM), 10% amyopathic DM (ADM), and 8% polymyositis (PM), with a mean age of 45.9 ± 18.1 years. Based on EULAR/ACR scores, 38% were definite IIM, 24% probable, 2% possible, and 36% were excluded. Biopsy confirmed DM in 10 cases. Juvenile onset occurred in five DM, one ADM, and one PM case. Most (95.6%) were diagnosed within a year. Proximal muscle weakness was more common in DM (P=0.001). Ethnicity: 46% Chinese, 42% Malay, 8% Indian; Malays had the earliest onset (mean age 39.8). Common skin signs included Gottron's papules (56%), heliotrope rash (52%), shawl sign (28%), and alopecia (20%). Shawl sign was more common in Chinese (P=0.018), telangiectasia in Malays (P=0.023). No skin sign was subtype-specific. CK levels were higher in DM (P=0.002) and varied by subtype (P=0.016). ILD was seen in five DM and one ADM patient (the latter anti-MDA5 positive). Five had malignancies; two were MSA-negative. Anti-MDA5 and anti-TIF1-γ antibodies were found in one DM and one juvenile ADM case.

CONCLUSIONS

The EULAR/ACR criteria effectively stratify IIM subtypes, with most DM and ADM cases classified as definite, probable, or possible. DM was the most common subtype, showing ethnic variations in age of onset and skin features. These results underscore the clinical diversity of IIM and validate the EULAR/ACR system in practice.

Table 1. Demographic and clinical characteristics (N=50)

		DM n (%)	ADM n (%)	PM n (%)	p
Patients (n/%)		41(82.0)	5(10.0)	4 (8.0)	
Gender	Male	7(70)	2 (20)	1 (10)	0.305
	Female	34 (85.0)	3 (7.5)	3(7.5)	
Ethnicity	Chinese	20 (87.0)	2 (8.7)	1 (4.35)	0.709
	Malay	16 (79.2)	2 (9.5)	3(14.3)	
	Indian	3 (75.0)	1 (25.0)	0	
	Others	0	2 (100)	0	
Age at diagnosis (Mean ± SD)		46.8 (±18.0)	45.8 (±20.5)	37.8 (±18.9)	
Age of Onset	< 18 years old	5 (71.4)	1 (14.3)	1 (14.3)	0.185
	18-39.9	7(77.8)	0	2 (22.2)	
	40 and above	29 (85.3)	4 (11.8)	1 (2.9)	
Duration from onset of symptoms to diagnosis	< 1 year	37 (86.1)	4 (9.3)	2 (4.6)	0.290
	1 to 2 years	1(50)	1 (50)	0	
Cutaneous manifestations	Gottron's papule	22 (78.6)	5(17.9)	1(3.6)	0.061
	Heliotrope	21 (80.8)	4 (15.4)	1(3.9)	0.306
	Shawl sign	12 (85.7)	2 (14.3)	0	0.502
	Alopecia	7 (70.0)	3 (30.0)	0	0.07
	V sign	5 (71.4)	2 (28.6)	0	0.220
	Poikiloderma	4 (66.7)	2 (33.3)	0	0.165
	Telangiectasia	4 (66.7)	2 (33.3)	0	0.165
	Mechanic's hand	1	0	-	-
Prox myopathy		32 (88.9)	0	4(11.1)	0.001
Skin Bx	Done	14 (82.4)	3(17.7)	0	-
Muscle Bx	Done	9 (90)	0	1 (10.0)	
EMG	Done	11(91.7)	0	1(8.3)	
Laboratory:	Raised CK	29 (87.9)	-	4 (12.1)	0.002
	CK (Mean±SD)	3975.3(±6837.6)	113.2(±57.2)	6645.3(±8127.2)	0.016
	LDH(Mean±SD)	588.5(± 554.3)	372(±182.4)	2088	0.258
	AST (Mean±SD)	192.2 (±285.2)	17.5(±2.121)	118.25(± 147.2)	0.118
	ESR (Mean±SD)	36.7(±26.0)	27.4 (± 10.73)	45.7 ((± 38.6)	0.816
	Positive ANA	28 (96.5)	1 (3.5)	0	0.001
ILD		5 (83.3)	1 (16.7)	0	0.717
Malignancy		4 (80.0)	0	1(20.0)	0.407

Table 2: Classification of IIM according to EULAR classification criteria scoring.

IIM	n (%)	Mean Score ± SD	Mean Probablity (%) ± SD	Diag	n	Mean_Score ± SD	Mean Probablity (%) ± SD
Definite_IIM	19 (38%)	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 (24%)	6.64 ± 0.71	74.1±10.5	PM	0		
				DM	11	6.58± 0.71	72.8 ± 10
				ADM	1	7.3	87.8
Possible_IIM	1 (2%)	6.4	47.8	PM	0		
				DM	1	6.4	47.8
				ADM	0		
Non-IIM	18 (36%)	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