Polymyositis is a rare and favourable outcome subtype of idiopathic inflammatory myopathy in Chinese patients

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Abstract

Objective

To investigate the prevalence and characteristics of typical polymyositis (PM) in Chinese patients with idiopathic inflammatory myopathy (IIM).

Methods

Patients diagnosed with IIM according to the 2017 EULAR/ACR criteria were included. Serological aspects including myositis-specific antibodies (MSA) and pathological data were re-evaluated. The diagnosis of typical PM was strictly done using the pathological criteria, while excluding other IIM subtypes such as dermatomyositis (DM), immune-mediated necrotising myopathies (IMNM), anti-synthetase syndrome (ASS), and sporadic inclusion body myositis (sIBM), based on their respective diagnostic criteria.

Results

A total of 544 IIM patients with muscle biopsy were involved, and 129 of them were diagnosed with initial PM according to the 2017 EULAR/ACR criteria. Only 6 (1.1%, 6/544) patients met the strict definition of typical PM after re-evaluation. Patients with typical PM were MSA-negative (100% vs. 35.7%, p=0.003) and had $CD8^+$ T cells surrounding or invading non-necrotic muscle fibres in muscle biopsies (100% vs. 7.8%, p<0.001) compared to the initially diagnosed PM patients. All typical PM patients achieved clinical remission at the second-year follow-up. Typical PM patients had a favourable prognosis compared to MSA-negative IMNM and unspecific myositis patients.

Conclusion

Strictly defined typical PM is a rare clinical subtype in Chinese IIM patients. Typical PM patients with classical pathology were MSA-negative and responded well to treatment and had a favourable prognosis. It is crucial for clinicians to combine clinical, serological, and pathological features to properly distinguish PM from other IIM subtypes.

Key words

polymyositis, prevalence, characteristics, pathology, prognosis

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Introduction

Idiopathic inflammatory myopathy (IIM) is a group of heterogenous autoimmune diseases characterised by muscle weakness and inflammatory infiltration of skeletal muscle (1). According to the 1975 Bohan & Peter criteria, IIM is classified into two clinical subtypes: polymyositis (PM) and dermatomyositis (DM) (2, 3). In 2004, the European NeuroMuscular Centre (ENMC) described the histopathological features of PM as endomysial inflammatory infiltration surrounding and invading non-necrotic muscle fibres. These features could be described as $CD8^+$ T cells surrounding non-necrotic muscle fibres, as well as overexpression of MHC-I on sarcolemma (4). In 2017, the European League Against Rheumatism (EULAR)/American College of Rheumatology (ACR) proposed the new classification criteria for IIM, dividing adult IIM into PM (which may include immune-mediated necrotizing myopathies (IMNM)), DM, and sporadic inclusion body myositis (sIBM) (5). Furthermore, the ENMC recommended diagnostic criteria for DM and IMNM in 2017 and 2018, respectively, based on clinical and pathological features, as well as the presence of myositis-specific antibodies (MSA) (6, 7).

However, previous studies have revealed that the prevalence of PM varies among different IIM cohorts according to the current criteria. For example, Jinin *et al*. reported a prevalence of 26.7% for PM in a large IIM cohort using the 2017 EULAR/ACR criteria (8). A recent study focusing on a sizable UK cohort found inconsistences in PM diagnosis between the 2017 EULAR/ACR criteria and expert opinion across all IIM subtypes. According to the 2017 EULAR/ACR criteria, 124 out of 255 IIM patients (48.6%) were classified as PM. However, only 37 patients (14.5%, 37/255) could be diagnosed as PM based on the expert consensus (9). A subsequent study conducted in the same cohort demonstrated that only nine out of 255 IIM patients (3.5%) could be ultimately diagnosed with PM after re-evaluating the data and when the current classification criteria were used (10). Reports from a Spanish study showed that after pathology re-evaluation and long-term follow-up of 1290 muscle biopsies, two-thirds of the patients who were initially diagnosed with PM were re-diagnosed as IBM and necrotizing myopathies (11). These studies suggest that PM is frequently misdiagnosed. Some experts highlighted that PM might be an over-diagnosed entity, and that the diagnosis of PM should exclude sIBM, IMNM, muscular dystrophy, metabolic myopathy and hereditary myopathy, as well as anti-synthetase syndrome (ASS), which is a unique subset of IIM characterised by interstitial lung disease (ILD) and serum positive anti-synthetase antibodies (12). Therefore, further studies are needed to accurately define PM and its characteristics.

In addition, the prevalence and clinical features of PM in Chinese IIM patients remain unclear. The aim of the study was to investigate the prevalence, characteristics, and long-term prognosis of PM in Chinese patients with IIM.

Patients and methods

Study population

Patients admitted to the department of Rheumatology of the China-Japan Friendship Hospital from 2015 to 2021 who performed muscle biopsy and were diagnosed as IIM according to the 2017 EULAR/ACR criteria were involved in the study. The clinical, serological and pathological data of patients were collected retrospectively from medical records. The study was approved by the Ethics Committee of the China-Japan Friendship Hospital (2019-SDZL-3). Oral consent was obtained from all participants. All methods used in this study adhered to the principles outlined in the Declaration of Helsinki.

Definition of typical PM

Typical PM was defined as MSA-negative, muscle biopsy findings with endomysial $CD8^+$T cells surrounding or invading non-necrotic muscle fibres, absence of rimmed vacuole, overexpression of MHC-I on sarcolemma, and exclusion of DM, IMNM, ASS, and sIBM. DM was diagnosed according to the 2017 ENMC criteria for DM. IMNM was diagnosed following the

2018 ENMC criteria for IMNM. ASS was diagnosed based on the 2011 Solomon criteria. The 2011 ENMC criteria for IBM were used to diagnose sIBM (4, 6, 7, 13). Patients who did not meet the above criteria and lacked typical PM pathological features were classified under unspecific myositis.

Evaluation of outcome

PM patients were followed-up from their initial visit to our department until June 30, 2023. Remission was defined as having muscle strength greater than grade 4 according to the Medical Research Council (MRC) grading system, normal creatine kinase (CK) level (less than 200 IU/L) and a glucocorticoid dose equivalent to prednisone less than or equal to 5 mg/d, with or without the use of immunosuppressants. Relapse referred to a deterioration in muscle strength, CK>200 IU/L, and the necessity to increase glucocorticoid dose or add additional immunosuppressants at any point during the follow-up period.

Statistical analysis

SPSS (V.24.0) and GraphPad Prism software (V.6.0; San Diego, California, USA) were used for statistical analyses. The data were expressed as medians with interquartile ranges for continuous variables and as numbers (percentages) for categorical variables. The Kruskal-Wallis H test (non-normal distribution) was used to analyse continuous data. The chi-square test and Fisher's exact test were used for categorical data. Two-sided *p*-values of <0.05 were considered statistically significant.

Results

The prevalence of typical PM in IIM

A total of 544 adult patients with IIM were involved in this study. According to the 2017 EULAR/ACR criteria, 129 were initially diagnosed with PM, after excluding DM, ADM, and sIBM. Among the initially diagnosed PM patients, 84 were MSA-positive and 45 were MSA-negative. After re-evaluating the serological and pathological data, the 84 patients who were MSA-positive were re-diagnosed with DM, IMNM, and ASS according to respective criteria. Among the 45 MSA-negative patients, re-diagnosis was done according to the pathological findings, resulting in the following reclassification: IMNM (29), unspecific myositis (10), and typical PM (6) (Fig. 1). The prevalence of typical PM in IIM was 1.1% (6/544).

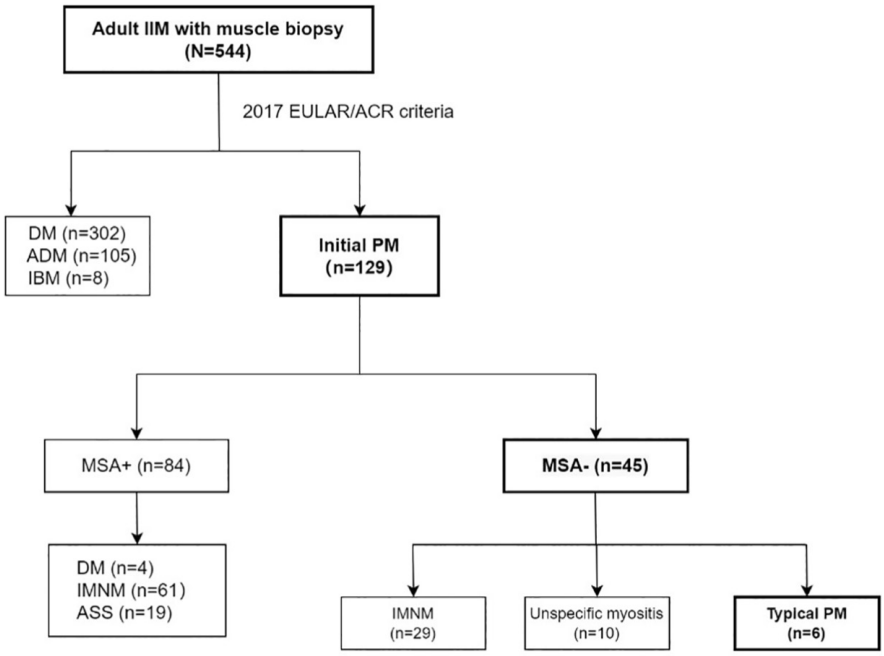

Fig. 1. Flowchart of the diagnosis of typical PM.

Comparison of the characteristics between initially diagnosed PM and typical PM patients

Upon comparing the characteristics between initially diagnosed PM and those of typical PM, no significant differences were observed in demographic and clinical features, including gender, age of onset, disease duration, muscle weakness, myalgia, and complication with other connective tissue diseases (CTD) or cancer. However, the initial PM had a higher proportion of MSA positivity than the typical PM (65.1% *vs*. 0, *p*=0.002). The most common MSA among initially diagnosed PM patients were anti-SRP and HMGCR antibodies, accounting for 34.1% and 13.2%, respectively. In addition, typical PM more frequently presented pathological features of $CD8^+$ T cells surrounding or invading non-necrotic muscle fibres compared with the initial PM group (100% *vs*. 7.8%, *p*<0.001). Other pathological features such as perifascicular atrophy, necrosis of muscle fibre, and overexpression of MHC-I on sarcolemma were found to be comparable between the two groups (Table I).

Comparison of clinical and pathological features among typical PM, MSA-negative IMNM and unspecific myositis patients

We further analysed the clinical and pathological characteristics of typical PM, MSA-negative IMNM, and unspecific myositis among 45 MSA-negative patients. Patients with typical PM exhibited the lowest incidence of myalgia compared to those with MSA-negative IMNM and unspecific myositis (16.7% *vs*. 27.6% and 80%, *p*=0.008). MSA-negative IMNM patients had the highest CK levels among the three groups (median, 2196 *vs*. 670 and 444 IU/L, *p*=0.007). Muscle biopsy with $CD8^+$ T cells surrounding or invading non-necrotic muscle fibres (100%, *vs*. 0 and 0, *p*<0.001) and overexpression of MHC-I on sarcolemma (100%, *vs*. 93.1% and 50%, *p*=0.01) were more prevalent in typical PM patients than in MSA-negative IMNM and unspecific myositis patients (Table II).

Outcome of typical PM patients

The typical PM, MSA-negative IMNM, and unspecific myositis patients were

Table I. Comparison of characteristics between initially diagnosed PM and typical PM patients.

Variables [a]	Initially diagnosed PM n=129	Typical PM n=6	*p*-value*
Female	94 (72.9)	5 (83.3)	1.0
Age onset (y)	48 (38, 59)	47 (35, 51)	0.539
Disease duration (m) [b]	3.0 (2.0, 8.5)	3.5 (2.8, 15.8)	0.435
Muscle weakness	113 (87.6)	6 (100)	1.000
Myalgia	51 (39.5)	1 (16.7)	0.405
Arthralgia	19 (14.7)	0	0.594
ILD	55 (42.6)	1 (16.7)	0.400
Cancer	5 (3.9)	0	1.000
Overlapped CTD	23 (17.8)	2 (33.3)	0.308
SSc	3 (2.3)	0	1.000
SLE	0	0	-
RA	4 (3.1)	1 (16.7)	0.206
pSS	16 (12.4)	1 (16.7)	0.561
MSA	84 (65.1)	0	0.002
SRP	44 (34.1)	-	-
HMGCR	17 (13.2)	-	-
Jo-1	6 (4.7)	-	-
PL-7	5 (3.9)	-	-
PL-12	3 (2.3)	-	-
EJ	4 (3.1)	-	-
OJ	1 (0.8)	-	-
SAE	1 (0.8)	-	-
Mi-2	1 (0.8)	-	-
NXP2	1 (0.8)	-	-
TIF1-γ	1 (0.8)	-	-
ANA	81 (62.8)	4 (66.7)	1.000
CK (IU/L)	1616 (531, 3377)	670 (539, 3863)	0.627
Pathological features			
Perifascicular atrophy [c]	2 (1.6)	0	1.0
Necrosis of muscle fibre	109 (84.5)	6 (100.0)	0.591
$CD8^+$T cells surrounding or invading non-necrotic muscle fibres[d]	10 (7.8)	6 (100.0)	<0.001
Overexpression of MHC-I on sarcolemma	106 (82.2)	6 (100.0)	0.589

ILD: interstitial lung diseases; CTD: connective tissue disease, including systemic sclerosis (SSc), systemic lupus erythematosus (SLE), rheumatoid arthritis (RA) and primary Sjogren's syndrome (pSS); MSA: myositis-specific antibody; MAA: myositis-associated antibody; ANA: anti-nuclear antibody; CK: creatine kinase.

[a]Continuous data are presented as median (first quartile, third quartile), and binary data are presented as n (%) of the patients.

[b]Disease duration: time from disease onset to hospitalisation.

[c]Two patients had perifascicular atrophy in initial PM, who were positive with anti-Jo-1 and PL-7 antibody respectively.

[d]Ten patients had the characteristic of CD8+T cells surrounding or invading non-necrotic muscle fibres in initially diagnosed PM, including 6 typical PM and other four with positive MSA: two anti-Jo-1, one anti-EJ and one anti-SRP.

*$p<0.05$ as significant.

followed-up for 2.8 to 8.6 years. Three MSA-negative IMNM patients were lost during the follow-up. But all the remaining patients are alive. During the first-year follow-up, no significant difference in remission rates was observed among the three groups. However, patients with typical PM exhibited higher remission rate than those with MSA-negative IMNM and unspecific myositis patients on the second-year follow-up (100% *vs*. 42.3% and 30%, $p=0.014$) (Fig. 2). The initial treatment did not differ significantly among the three groups (Supplementary Table S1). The clinical characteristics and outcomes of typical PM are presented in Table III. Two patients experienced recurrence once during the follow-up but achieved remission again after receiving a repeat administration of medial dose of glucocorticoid.

Discussion

The classification criteria for IIM, particularly the definition of PM, have been discussed for many years. Whether PM is an entity is still a controversy. It has been noted that MSA is associated with the clinical phenotype of IIM (1). Therefore, MSA can be used for classifying the IIM subtype. For instance, anti-Mi-2, -TIF1, -NXP2, -MDA5, and -SAE are specific antibodies for DM. Anti-SRP, and -HMGCR antibodies are used to diagnose IMNM. And anti-ARS antibodies are closely related to ASS (14-16). However, to date, no MSA that is related to PM has been identified. This may pose challenges in distinguishing PM from MSA-negative IIM and condition such as virus-triggered myositis (17). Therefore, the diagnosis of PM heavily depends on clinical manifestations and pathological findings. This study is the first to investigate the prevalence and characteristics of PM in a large Chinese IIM cohort, based on the MSA and muscle biopsy information. Our findings revealed that the proportion of typical PM in IIM was only 1.1%.

The EULAR/ACR classification criteria for IIM were established in 2017 and subsequently validated in different cohorts. When applying the new criteria to classify IIM, the proportions of PM in IIM were 14.5%, 41.7%, and 26.7% in the UK, Swedish, and Japan cohorts, respectively (8, 9, 18). In our study, 23.7% (129/544) of patients were initially diagnosed with PM using the 2017 EULAR/ACR criteria with muscle biopsy. However, 84 out of them had positive MSA including those related to DM, IMNM, and ASS. These were excluded from being diagnosed as PM, following the current criteria. In 1991, Dalakas *et al*. introduced endomysial inflammation surrounding or invading non-necrotic muscle fibres as a PM pathology (19). In 2004, the ENMC proposed the pathological criteria for definite and probable PM (4). In our study, we evaluated the typical PM using the strict definition in the initially diagnosed PM patients. Only six (1.1% of IIM) patients could ultimately be diagnosed with PM, which was significantly lower compared to previous

Table II. Comparison of characteristics among typical PM, MSA-negative IMNM and unspecific myositis patients.

Variables [a]	Typical PM n=6	MSA-negative IMNM n=29	Unspecific myositis n=10	*p*-value*
Female	5 (83.3)	19 (65.5)	10 (100)	0.088
Age onset (y)	47 (35, 51)	44 (33, 49)	50 (30, 58)	0.660
Disease duration (m)	3.5 (2.8, 15.8)	4.0 (2.0, 8.5)	2.5 (1.0,9.8)	0.610
Muscle weakness	6 (100.0)	27 (93.1)	8 (80.0)	0.285
Myalgia	1 (16.7)	8 (27.6)	8 (80.0)	0.008
Arthralgia	0	5 (17.2)	1 (10.0)	0.825
Dysphagia	1 (16.7)	9 (31.0)	4 (40.0)	0.644
Skin rash	0	0	0	-
Overlapped CTD	2 (33.3)	8 (27.6)	2 (20.0)	0.893
SSc	0	0	1 (10.0)	0.356
SLE	0	0	0	-
RA	1 (16.7)	2 (6.9)	0	0.456
pSS	1 (16.7)	6 (20.7)	1 (10.0)	0.851
Cancer	0	0	0	-
ILD	1 (16.7)	6 (20.7)	3 (30.0)	0.868
ANA	4 (66.7)	14 (48.3)	6 (60.0)	0.745
CK (IU/L)	670 (539, 3863)	2196 (1169, 4440)	444 (44, 849)	0.007
Pathological features				
Perifascicular atrophy	0	0	0	-
Necrosis of muscle fibre	6 (100.0)	29 (100.0)	4 (40.0)	<0.001
$CD8^+$ T cells surrounding or invading non-necrotic muscle fibres	6 (100.0)	0	0	<0.001
Overexpression of MHC-I on sarcolemma	6 (100.0)	27 (93.1)	5 (50.0)	0.01

ILD: interstitial lung diseases; CTD: connective tissue disease, including systemic sclerosis (SSc), systemic lupus erythematosus (SLE), rheumatoid arthritis (RA) and primary Sjogren's syndrome (pSS); MSA: myositis-specific antibody; MAA: myositis-associated antibody; ANA: anti-nuclear antibody; CK: creatine kinase.

[a]Continuous data are presented as median (first quartile, third quartile), and binary data are presented as n (%) of the patients.

*p<0.05 as significant.

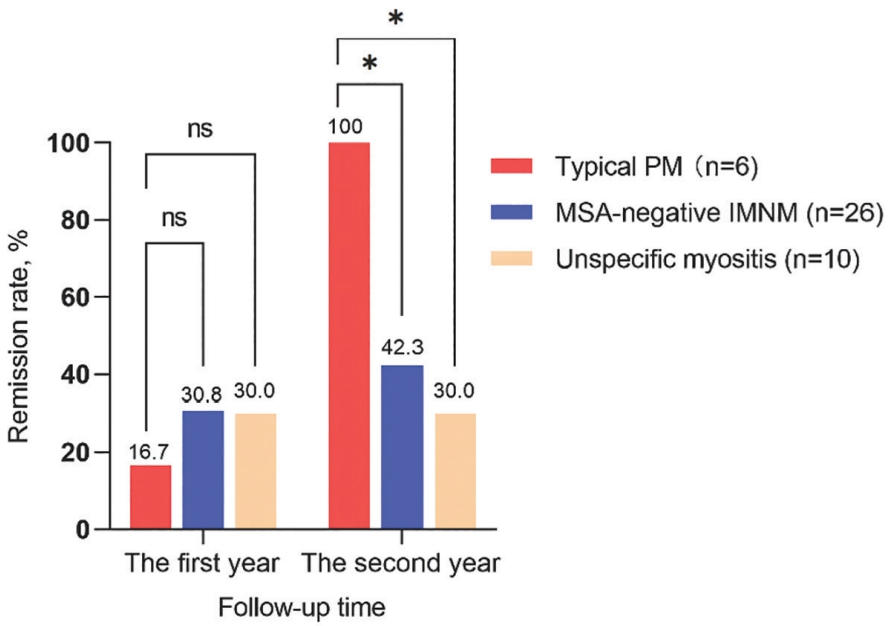

Fig. 2. The prognosis of typical PM, MSA-negative IMNM and unspecific myositis patients. The three groups were followed up for two years and remission rates were compared in the first and second year. *p<0.05.

studies. Lorce Martos *et al*. reported the prevalence of PM in a UK cohort, where only nine (3.5% out of IIM) patients could be diagnosed with PM, having excluded CTD-OM, IMNM, DM, and other myopathies in the previous 37 (14.5% out of IIM) PM patients (10). However, muscle pathology was not the inclusion criteria for diagnosing PM in their study, which might explain the significant difference in the proportion of PM between our study and previous studies. In a 2015 Spanish cohort study, Vilela *et al*. reported that after long-term follow-up, IBM, necrotizing autoimmune myositis (NAM) and CTD were excluded, and the incidence of PM in IIM was only 0.6%, which was similar to our study (11).

In some of the previous studies mentioned above, ASS patients were included in the category of PM. ASS patients had distinct characteristics with different serum anti-ARS antibodies. Some patients had typical DM-like skin rash and muscle involvement. Most of the previous studies showed that ASS presents unique muscle biopsy features such as perifascicular necrosis and myofascial fragmentation (20, 21). However, the 2018 ENMC criteria for DM recommended that patients with typical DM-like rash and positive anti-synthetase antibodies should be excluded when diagnosing DM (7). Thus, ASS is a distinct subtype of IIM, and experts suggested that it should be separately classified in PM. In this study, 129 patients were initially diagnosed with PM, but 19 of them were anti-ARS antibodies positive. These patients were excluded based on the strict definition of PM.

Forty-five of the patients who were initially diagnosed with PM were MSA-negative. After muscle biopsy evaluation, 29 of these patients met the 2018 ENMC recommended pathological criteria for IMNM. While ten of the patients matched unspecific myositis, based on the 2004 ENMC criteria, which is characterized by scattered inflammatory infiltration around blood vessels, degeneration of muscle fibre, and upregulation of MHC-I on sarcolemma (4). There are still controversies on whether these unspecific myositis

Table III. Clinical characteristics and outcome of typical polymyositis patients.

Variables	P1	P2	P3	P4	P5	P6
Sex	F	F	F	F	M	F
Age (y)	48	60	24	48	39	46
Disease duration (m)	3	2	4	10	3	13
Muscle strength*						
Proximal upper	4	3^+	4^+	5	5	4^+
Proximal lower	3	2	2^+	2	3^+	2
Neck flexor	3	3^+	4	3^+	5	4
Rash	N	N	N	N	N	N
Arthralgia	N	Y	N	N	N	N
Myalgia	Y	N	N	N	N	N
Dysphagia	Y	N	N	N	N	Y
ILD	N	N	N	Y	N	N
Cancer	N	N	N	N	N	N
Overlapped CTD	N	RA	N	N	pSS	N
CK (IU/L)	4362	702	638	510	549	3696
MAA	PM-Scl 75	N	Ku	N	Ro-52	N
ANA	Y	N	N	N	Y	N
Initial treatment	MP 40mg/d + CsA 200mg/d	Pred 40mg/d	Pred 40mg/d+ MTX 15mg/w	Pred 30mg/d+ MTX 12.5mg/w	Pred 30mg/d	Pred 30mg/d
Follow-up period (m)	47	103	63	49	44	46
Survival	Y	Y	Y	Y	Y	Y
Remission	Y	Y	Y	Y	Y	Y
Relapse times	0	1	0	0	0	1
Current treatment	MP 4.25mg/d+ CsA 150mg/d	Pred 5mg/d	Pred 5mg/d+ MTX 10mg/w	Pred 5mg/d	Pred 2.5mg/d	Pred 5mg/d

Pred: prednisone acetate; MP: methylprednisolone; CsA: cyclosporin; MTX: methotrexate. Additional definitions are provided in Table I.
*Muscle strength was evaluated according to Medical Research Council (MRC) grading system.

patients should be included in or excluded from the diagnosis of PM. Previous studies have shown that overlapping syndromes, ASS, and even DM patients presented unspecific muscle pathology (22-24). In our study, two had other CTD but none had rashes among the ten patients with unspecific pathology. Compared with typical PM and MSA-negative IMNM patients, the patients with unspecific myositis had mild abnormality in CK levels and pathological findings. However, only 30% (3/10) of the patients achieved remission after two-year treatment, and this could be attributed to a tendency of low/medial glucocorticoid dose (70% *vs*. 50% and 31%, $p>0.05$) and less possibility of adding immunosuppressants (60% *vs*. 66.7% and 75.9%, $p>0.05$) during the initial treatment (Supplementary Table S1). In addition, these patients tended to exhibit a higher prevalence of ILD (30% *vs*. 16.7% and 20.7%) and dysphagia (40% *vs*. 16.7% and 31%), though no significant differences were observed (Table II). The pathology may be related to the timing and location of the muscle biopsy, but it may also suggest that unspecific myositis and typical PM are different entities. Further research is needed to identify how to diagnose and classify unspecific myositis patients accurately.

IIM is a group of heterogeneous diseases characterised by significant differences in treatment response and prognosis among various subtypes. Patients with IMNM present with significantly elevated CK levels, severe muscle weakness, resistance to immunosuppressants, and an unfavourable prognosis (15). ASS patients are more prone to developing ILD and experiencing increased mortality, particularly those with the anti-PL-7 or PL-12 antibody (25). The prognosis of DM patients is closely linked to their MSA status. For example, anti-TIF1 positive DM patients were at a higher risk of developing cancer (26), while anti-MDA5 positive patients with DM are more susceptible to RP-ILD, which leads to high mortality rates (27). sIBM is a common subtype of IIM in elderly patients who do not respond to glucocorticoid, immunosuppressants, and even intravenous immunoglobin treatments (28). However, research that evaluates the long-term prognosis of PM patients is still lacking. This study demonstrates that typical PM patients respond well to steroid therapy and display a favourable prognosis. Although most patients had significant elevation in CK levels and muscle weakness (less than grade IV muscle strength) at the onset of the disease, 100% of them achieved remission within a two-year treatment period with glucocorticoids or combining with immunosuppressants. Two patients experienced one recurrence in the subsequent follow-up, and the condition improved after retreatment. No typical PM patient died during the follow-up period. Although the number of patients with typical PM was small in the study, the findings suggested that this type of IIM displays a favourable prognosis.

This study still had several limitations. Firstly, the study only included IIM patients who had muscle biopsy information, while excluding suspect PM patients who did not undergo muscle biopsy. Such a scenario may lead to a reduced frequency of PM. Secondly, as a single-centre study, there is a potential for data bias. Lastly, the small sample size of patients identified as having typical PM may not fully represent

clinical characteristics of PM. Validation should be conducted in a larger and multiple-centre cohort in future studies. To conclude, the definition of PM remains a topic of ongoing debate and consensus has not yet been reached. This study showed that typical PM is a rare subtype in Chinese IIM patients. The patients with PM exhibited good responses to treatment with favourable outcomes. These findings may provide data that aids the development of new classification criteria for IIM in the future.

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