

Clinical Findings, Antibody Panel and Pathology of Patients with Inflammatory Myopathies in Isfahan Province, Iran

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Abstract

Background: In the current study, we aimed to evaluate the characteristics of individuals with autoimmune inflammatory myopathies (AIMs) in Isfahan.

Material and Methods: In this retrospective, observational study, the clinical, histopathological, immunohistochemical staining, electromyography, and autoantibody profile of the patients with AIM in subgroups of dermatomyositis (DM), necrotizing autoimmune myopathy (NAM), polymyositis (PM), and antisynthetase syndrome (AS) were assessed.

Results: Seventy-seven patients with AIMs including 38 (49.3%), 21 (27.3%), 14 (18.2%), and 4 (5.2%) with NAM, DM, PM, and antisynthetase diagnoses were enrolled, respectively. Proximal weakness (94.80%), myalgia (32.46%), skin rash (25.97%), and dysphagia (25.97%) were the most common presentations, among which skin rash was statistically more frequent in DM (P value = 0.001). Fiber necrosis (100%), CD68 (92.1%), and C5-b9 (60.5%) positivity, irritable myopathy (78.9%), and anti-Ro-52 (7.1%) positivity were the most common findings in NAM. Perivascular infiltration (71.4%), fiber degeneration (66.7%) and necrosis (52.4%), CD68 (100%), CD20 (71.4%), and CD8 (71.4%), both irritable (57.1%) and non-irritable (38.1%) myopathy, anti-Mi-2 (19%), anti-NXP-2 (19%), and anti-MDA-5 (14.3%) autoantibodies were the detected characteristics of DM patients. PM individuals were characterized with endomysial infiltration (92.9%), fiber necrosis (71.4%) and perivascular infiltration (64.3%), CD68 (100%), CD20 (100%) and CD8 (92.9%) positivity, both irritable (57.1%) and non-irritable (42.9%) myopathy and anti-Mi-2 (14.3%), and anti-ANA (14.3%) antibodies. Fiber necrosis (100%) and degeneration (75%), CD68 (75%), CD8 (75%), and C5-b9 (75%), irritable myopathy (50%), and anti-Jo-1 antibody (100%) positivity were the diagnostic tools for AS.

Conclusion: According to the outcomes of the current study, the clinical, histopathological, and autoantibody profiles of the patients with AIM are variably different with remarkable overlaps. Therefore, thorough assessments are required to make the final diagnosis.

Keywords: Antisynthetase syndrome, autoimmune diseases, dermatomyositis, muscular diseases, myositis, polymyositis

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INTRODUCTION

Autoimmune inflammatory myopathies (AIMs) are a set of heterogeneous treatable myopathies and constitute a subgroup of very heterogeneous muscular diseases by their clinical presentation, their histological profile and serological, their association or not with extra muscular manifestations, and by their response to treatment. AIMs are

generally rare sporadic conditions involving approximately 1 in 100,000 annually. These disorders predominantly affect women.^[1] It is rare in adults, with an estimated incidence of 2 cases per 100,000 per year and a prevalence of 30 cases per 100,000 individuals.^[2] Muscle weakness is the key point in AIMS and inflammatory manifestations play an integral role in their pathophysiology.^[3] AIMs are characterized

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by subacute-onset and often severe progressive proximal muscle weakness.^[3]

AIMs at present are classified into four subgroups: dermatomyositis (DM), necrotizing autoimmune myopathy (NAM), polymyositis (PM)/overlap myositis (OM), and antisynthetase syndrome (AS). Those who are primarily diagnosed with PM might later be diagnosed as OM. However, both children and adults might present DM, while all other types of myositis mostly involve middle-aged individuals.^[4]

Standard laboratory testing such as aspartate aminotransferase, alanine aminotransferase, serum creatine kinase, lactate dehydrogenase, and aldolase, as well as imaging modalities such as ultrasonography or magnetic resonance imaging are widely used to diagnose AIMs. Other diagnostic tools include myositis autoantibody testing, muscle biopsy, and electromyography (EMG). More novel modalities, such as myositis-specific autoantibody testing and ultrasound were accompanied by encouraging results.^[5]

Muscle biopsy plays a crucial role in diagnosis particularly for those with negative myositis-specific antibodies. The findings of biopsy in immunohistochemical dying show autoantibodies against T and B lymphocytes (CD4, CD8, and CD20), macrophages (CD68), major histocompatibility complex class I antigen, and membrane attack complexes.^[6,7]

In the present study, we try to present a thorough view of inflammatory myopathies in Isfahan province, Iran.

MATERIALS AND METHODS

In the current retrospective observational study, data from 77 patients with documented diagnoses of AIMs were recruited. The patients were selected among those who were admitted at Al-Zahra and Kashani Tertiary Centers, affiliated with Isfahan University of Medical Sciences, Isfahan, Iran from March 2020 to March 2022.

The study was designed based on the tenets of the Helsinki declaration and approved by the Ethics Committee of Isfahan University of Medical Sciences via code number IR.MUI.MED.REC.1401.163. All the patients were informed about the probable use of their medical data for scientific research at the time of admission and requested to sign a written consent for the use of their medical data.

The patients were categorized into four diagnostic subgroups including necrotizing NAM, DM, and AS, and those who did not have a specific pathology were classified into the PM subgroup.

Demographic (age and gender), clinical symptoms (proximal weakness, myalgia, skin rash, dysphagia, weight loss, lung involvement, head drop, generalized weakness, lower limb weakness, distal weakness, hoarseness, calf hypertrophy, dysarthria, and splenomegaly), reports of muscle biopsy (fiber necrosis and degeneration, endomysial infiltration, perivascular infiltration, perimysial infiltration, perifascicular atrophy and

perifascicular necrosis), immunohistochemical staining (CD68, CD8, C5-b9, CD20, MHC-1, and CD4), anti-nuclear autoantibodies (anti-Mi-2, anti-MDA-5, anti-NXP-2, anti-TIF-1 γ , anti-Jo-1, anti-Ro-52, anti-PM/Scl 75, and anti-SRP), and EMG reports of the patients were recruited from the archives of the hospitals.

Statistical analyses

Data were analyzed using SPSS version 18.0 (SPSS Inc., Chicago, IL, USA). Mean and standard deviation were applied to present continuous variables. Categorical data were presented in absolute numbers and percentages. Continuous variables were compared using an independent *t*-test, while the categorical ones were compared by applying the Mann-Whitney, and the Chi-square or Fisher's exact tests. The *P* value of less than 0.05 was considered statistically significant.

RESULTS

Data from 77 patients with documented diagnoses of AIMs were evaluated. The studied population predominantly consisted of females ($n = 60$, 77.9%) and had a mean age of 49.2 ± 15.76 years old. The subtypes of AIMs in the studied patients included NAM, DM, PM, and AS with frequencies of 49.3%, 27.3%, 18.2%, and 5.2%, respectively.

Considering the type of AIMs, the groups were not statistically different considering their mean age (*P* value = 0.107). Gender distribution statistically differed among the subgroups; however, all the AIM subtypes predominantly consisted of females (*P* value = 0.020) [Table 1].

The clinical symptoms including proximal weakness (94.80%), myalgia (32.46%), skin rash (25.97%), and dysphagia (25.97%) were the most common presentations for which AIM diagnosis was made. Further symptoms are presented in Table 1 in detail. The comparison of the AIM subtypes in terms of the clinical presentations revealed insignificant differences except for skin rash that was remarkably more prevalent in DM and AS than in the other groups (*P* value = 0.001) [Table 1].

The major histopathological findings in the patients included necrotic fibers (81.8%), fiber degeneration (51.9%), endomysial infiltration (48.1%), and perivascular infiltration (40.3%). Considering the subtype of AIMs, the histopathological characteristics variably differed. Necrotic fiber was present in all cases with NAM and AS, while it was significantly less common in PM (71.4%) and DM (52.4%) (*P* value = 0.001). Endomysial infiltration was the most common finding of PM (92.9%) compared to other diseases ($\leq 50\%$) (*P* value = 0.001). Perivascular infiltration was mostly noted in DM (71.4%) and PM (64.3%) than the others (*P* value = 0.001). Perimysial infiltration (*P* value = 0.007) and perifascicular atrophy (*P* value = 0.011) were commonly seen in DM as compared to other AIMs. Detailed information is presented in Table 2.

Table 1: Clinical and therapeutic characteristics of patients in subgroups of AIMs

Variables	AS (n=4)	DM (n=21)	NAM (n=38)	PM (n=14)	Total (n=77)	P
Age/year (Mean±SD)	44.3±17.32	45.3±11.36	52.7±16.82	44.8±16.60	49.2±15.76	0.107
Sex						
Female	3 (75.0)	18 (85.7)	25 (65.8)	14 (100.0)	60 (77.92)	0.020
Male	1 (25.0)	3 (14.3)	13 (34.2)	0 (0.0)	17 (22.07)	
Clinical symptoms						
Proximal weakness	2 (50.0)	19 (90.5)	38 (100.0)	14 (100.0)	73 (94.80)	0.719
Myalgia	1 (25.0)	8 (38.1)	10 (26.3)	6 (50.0)	25 (32.46)	0.218
Skin rash	2 (50.0)	13 (61.9)	4 (10.5)	1 (12.5)	20 (25.97)	0.001
Dysphagia	1 (25.0)	5 (23.8)	9 (23.7)	5 (25.0)	20 (25.97)	0.557
Weight loss	1 (25.0)	2 (9.5)	8 (21.1)	1 (12.5)	12 (15.58)	0.496
Lung involvement	2 (50.0)	0 (0.0)	5 (13.2)	2 (12.5)	9 (11.68)	0.092
Head drop	0 (0.0)	2 (9.5)	5 (13.2)	0 (0.0)	7 (9.09)	0.490
Generalized weakness	0 (0.0)	1 (4.8)	3 (7.9)	0 (0.0)	4 (5.19)	0.813
Lower limb weakness	1 (25.0)	1 (4.8)	1 (2.6)	0 (0.0)	3 (3.89)	1.000
Distal weakness	1 (25.0)	1 (4.8)	1 (2.6)	0 (0.0)	3 (3.89)	1.000
Hoarseness	0 (0.0)	1 (4.8)	1 (2.6)	0 (0.0)	2 (2.59)	0.455
Calf hypertrophy	0 (0.0)	0 (0.0)	1 (2.6)	0 (0.0)	1 (1.29)	1.000
Dysarthria	0 (0.0)	0 (0.0)	1 (2.6)	0 (0.0)	1 (1.29)	1.000
Splenomegaly	0 (0.0)	0 (0.0)	0 (0.0)	1 (12.5)	1 (1.29)	0.182

DM=Dermatomyositis, NAM=Necrotizing autoimmune myopathy, PM=Polymyositis

Immunohistochemical staining of the muscle biopsies revealed that the majority of biopsies were positive for CD68 (94.8%), CD8 (61.03%), C5B59 (57.1%), and CD20 (53.2%). CD8 positivity was remarkably high in those diagnosed with PM (92.9%), AS (75%), and DM (71.4%) compared with NAM cases (2.1%) (P value = 0.004). A significantly high rate of positive biopsies for CD20 was found in PM (100%) and DM (71.4%) compared to the other diagnoses (P value = 0.001). Other data are shown in Table 2.

Irritable myopathy (67.5%) was the most common finding of EMG in the studied population, while the subgroups did not differ in terms of EMG reports (P value = 0.101) [Table 2].

The assessment of antinuclear antibodies revealed that 48.1% of the patients were negative for all the antibodies. Nevertheless, all the patients with AS were positive for anti-Jo-1. The other antibodies were variably present in different AIMs [Table 2].

DISCUSSION

In the current study the characteristics of AIMs, a set of heterogeneous autoimmune diseases affecting muscles, were evaluated in 77 patients including 38 (49.3%), 21 (27.3%), 14 (18.2%), and 4 (5.2%) with NAM, DM, PM, and AS, respectively.

Women were the dominant population of this study, who were in their 5th–6th decades of life. Those with NAM were relatively but not significantly older than the others. Surfing the studies in the literature have shown outcomes in line with our findings representing that the peak age of AIM incidence is over 40–50; however, it variably differs according to the subtype of AIM.^[8,9] Moreover, similar to most autoimmune

disorders, AIMs predominantly occur in females rather than males.^[10]

Proximal weakness (94.80%), myalgia (32.46%), skin rash (25.97%), and dysphagia (25.97%) were the most common clinical presentations of the studied patients. Dysphagia was more frequent in NAM patients, while skin lesions and lung involvement were noted in half of the AS patients. The patients did not differ regarding their clinical characteristics except for skin rash, which was remarkably higher among the individuals with DM. Proximal weakness is the major characteristic of all types of AIMs; however, other features help to differentiate these diseases.^[11] NAM is known to be mainly presented with symmetrical proximal weakness.^[12] Dysphagia and cardiac involvement are common extramuscular manifestations of NAM.^[13] Gottron's papules and heliotrope rash are the major cutaneous presentations of DM, while muscle weakness initiates within 6 months after developing of these skin alterations. Other features include photosensitivity, poikiloderma, diffuse erythema, palmar papules, and diffuse hair loss, as well as nail changes including ragged cuticles and periungual erythema.^[14,15] Dysphagia and lung involvement are the other presentations of DM^[16,17]; however, we found no lung involvement in our DM patients. Similar to the other AIMs, proximal muscle weakness is the major complaint of PM, as well.^[4] AS generally might be represented with characteristics of DM or PM; however, recent genetic studies have included this myopathy as overlap myositis. Extramuscular involvement is common in AS. Interstitial lung disease is the most devastating morbidity in AS; however, other manifestations include arthritis, Raynaud phenomenon, and cutaneous involvement called mechanic's hands, characterized by hyperkeratosis and fissuring of the palmar skin.^[18,19]

Table 2: Electroneuromyography and antibodies of patients in subgroups of AIMs

Variables	AS (n=4) n (%)	DM (n=21) n (%)	NAM (n=38) n (%)	PM (n=14) n (%)	Total (n=77) n (%)	P
Histopathological factors						
Necrotic fibers	4 (100.0)	11 (52.4)	38 (100.0)	10 (71.4)	63 (81.8)	0.001
Fiber degeneration	3 (75.00)	14 (66.7)	19 (50.0)	7 (50.0)	40 (51.9)	0.272
Endomysial infiltration	2 (50.0)	9 (42.9)	13 (34.2)	13 (92.9)	37 (48.1)	0.001
Perivascular infiltration	1 (25.0)	15 (71.4)	6 (15.8)	9 (64.3)	31 (40.3)	0.001
Perimysial infiltration	1 (25.0)	11 (52.4)	6 (15.8)	2 (14.3)	20 (26.0)	0.007
Perifascicular atrophy	1 (25.0)	11 (52.3)	0 (0.0)	0 (0.00)	13 (16.9)	0.011
Perifascicular necrosis	1 (25.0)	2 (9.5)	1 (2.6)	1 (7.1)	4 (5.2)	0.382
Immunohistochemical staining						
CD68	3 (75.00)	21 (100.0)	35 (92.1)	14 (100.0)	73 (94.8)	1.000
CD8	3 (75.00)	15 (71.4)	16 (2.1)	13 (92.9)	47 (61.03)	0.004
C5-b9	3 (75.00)	12 (57.1)	23 (60.5)	6 (42.9)	44 (57.1)	1.000
CD20	1 (25.0)	15 (71.4)	11 (28.9)	14 (100.0)	41 (53.2)	0.001
MHC1	1 (25.0)	10 (47.6)	11 (28.9)	5 (35.7)	27 (35.1)	0.795
CD4	1 (25.0)	7 (33.3)	11 (28.9)	7 (50.0)	26 (33.7)	0.394
Electroneuromyography						
Irritable myopathy	2 (50.0)	12 (57.1)	30 (78.9)	8 (57.1)	52 (67.5)	0.101
Non irritable myopathy	1 (25.0)	8 (38.1)	6 (15.8)	6 (42.9)	21 (27.3)	
Irritable proximal myopathy with neuropathy	0 (0.0)	0 (0.0)	3 (7.9)	0 (0.0)	3 (3.9)	
Irritable generalized myopathy with myotonic discharge	0 (0.0)	1 (4.8)	0 (0.0)	0 (0.0)	1 (1.3)	
Antinuclear antibodies						
Anti-Mi-2	0 (0.0)	4 (19.0)	1 (2.4)	2 (14.3)	7 (9.1)	0.001
Anti-MDA-5	0 (0.0)	3 (14.3)	2 (4.8)	1 (7.1)	6 (7.8)	
Anti-NXP-2	0 (0.0)	4 (19.0)	2 (4.8)	0 (0.0)	6 (7.8)	
Anti-ANA	0 (0.0)	2 (9.5)	1 (2.4)	2 (14.3)	5 (6.5)	
Anti-TIF-1 γ	0 (0.0)	2 (9.5)	2 (4.8)	0 (0.0)	4 (5.2)	
Anti-Jo-1	4 (100.0)	0 (0.0)	0 (0.0)	0 (0.0)	4 (5.2)	
Anti-Ro-52	0 (0.0)	0 (0.0)	3 (7.1)	1 (7.1)	4 (5.2)	
Anti-PM/Scl 75	0 (0.0)	1 (4.8)	2 (4.8)	1 (7.1)	4 (5.2)	
Anti-SRP	0 (0.0)	0 (0.0)	2 (4.8)	0 (0.0)	2 (2.6)	
Unknown	0 (0.0)	10 (47.6)	21 (59.5)	6 (42.9)	37 (48.1)	

AS=Antisynthetase syndrome, DM=Dermatomyositis, NAM=Necrotizing autoimmune myopathy, PM=Polymyositis

The evaluated NAM patients in our study had fiber necrosis (100%), half of them had degeneration, and perimysial, endomysial, and perivascular infiltration were the other findings in NAM. Other biomarkers were positive in less than 30% of the patients. Similarly, studies in the literature have insisted on the significance of necrosis in NAM; however, in line with our study they have mentioned fiber degeneration and regeneration as well as endomysial infiltration as the other features of muscular biopsy in NAM.^[20,21]

Perivasclar infiltration, fiber degeneration, perimysial infiltration, and atrophy and perifascicular infiltration were notably high in DM. Perifascicular infiltration that was positive in more than 50% of our patients is a known characterizing finding of DM presented in other studies. Accordingly, the researchers have emphasized on the significance of perifascicular and perivascular infiltration as well as reduced number of capillaries in DM.^[10,18,22]

Endomysial infiltration, fiber necrosis, perivascular infiltration, and fiber degeneration were the hallmarks found in PM

patients. Lundberg and colleagues also presented fiber necrosis and degeneration as the histopathological finding of PM. They continued that endomysial infiltration by T cells can be seen in PM.^[10,23]

The histopathological study of AS patients' biopsy showed necrotic and degenerated fibers and endomysial infiltration. Muscle biopsy in AS may show variable characteristics such as perivascular, perimysial, and endomysial infiltrations.^[24,25] However, although fascicular necrosis is a usual finding in AS, we observed it in other AIMs, as well.

Immunohistochemical staining of the muscles was positive for CD68 in nearly all the patients. The next biomarker was CD8; however, it was rarely positive in NAM. CD68 is the biomarker of macrophages and a representative of inflammation in muscles. Therefore, its positivity in AIM individuals was not surprising. CD8 is the symbol of cytotoxic T cells. However, it is mostly positive in AIM, its presence is not necessarily showing AIM, as it might be present in other diagnoses.^[11,26]

NAM patients were majorly positive for CD68 and C5-b9. These two findings are common immunohistochemical characteristics of NAM. The first one is representative of macrophage activity and the latter shows (the membrane attack complex on sarcolemma.^[13]

CD68, CD8, CD20, C5B9, and MHC1 were remarkably positive in DM individuals, while CD4 positivity was limited to 33% of the cases. The pathophysiology of DM involves all B cells, T cells, and macrophages. Therefore, coloring of CD4, CD20, CD8, and CD68 might variably be seen in these individuals.^[27]

The distribution of immunohistochemical staining in our patients diagnosed with PM revealed the considerable positivity of CD68, CD20, and CD8 in these patients. MHC-1 was positive in 35% of the cases. Autoaggressive inflammatory cells of CD8 and CD68 positive cells surround the myofibers and destroy the normal appearance of these fibers. This is the Hallmark characteristic of PM.^[28] Overexpression of MHC-I, even on unaffected myofibers is another finding in PM.^[15]

The main reason to administer EMG in these individuals is to rule out other differential diagnoses including neurogenic or neuromuscular junction disorders. Proximal muscular irritability is frequent in these patients. Nevertheless, irritable myopathy is the most common finding, which is compatible with our study. EMG study shows small myopathic units, with fibrillation potentials, and the absence of large neurogenic units.^[18]

The majority of the patients were not positive for any of the antinuclear antibodies. This issue has been presented in several other studies, as well. For instance, Betteridge *et al.*^[29] presented that over 30% of AIMs do not have any specific anti-nuclear autoantibody or might not be positive for any of them. However, we assume the quality of our kits might be responsible for this high rate of negative responses. Nevertheless, anti-Ro52, anti-PMscl, anti-Ku, and anti-U1RNP are the most common diagnostic ones in individuals with AIM. This panel is well-known as myositis-associated autoantibodies.^[10] However, the positivity for these antibodies was limited to less than 10% in our patients.

Anti-SRP and anti-HMGCR are frequently found in NAM. It has been estimated that up to 40% of NAM individuals are negative for all the anti-nuclear autoantibodies.^[13] In our study, approximately 62% of dermatomyositis patients had one of the four dermatomyositis-specific autoantibodies (anti-Mi2, anti-TIF1- γ , anti-NXP-2, and anti-MDA-5). This finding is compatible with a study by Mammen *et al.*,^[30] who showed that over 65% of DM patients have at least one of the autoantibodies that are specific for DM. Anti-Mi2 is one of the autoantibodies classically associated with skin lesions and related to the incidence of calcinosis. It has been documented that the presence of anti-Mi2 is a good Hallmark for the response to immunosuppressant medications.^[4] Anti-Mi-2 and anti-ANA were detected in 15% of PM patients. Anti-Jo

as the marker of AS diagnosis was positive in 100% of our patients. This autoantibody is the Hallmark for AS diagnosis; however, cannot predict the probability of lung involvement and disease prognosis.^[10,26]

Despite the value of our study in the assessment of AIM as these myopathies are relatively rare, it has several limitations. The lack of examination of alkaline phosphatase, and acid phosphatase in patients was the main limitation in this study. Furthermore, the retrospective observational design of the study might be accompanied by several weak points including data loss or collection errors as well as applying non-uniform therapeutic approaches. In addition, the non-randomized data registry might have led to remarkable biases. Despite the value of this study in presenting noteworthy data about AIMs, large prospective randomized controlled studies are strongly recommended.

CONCLUSION

The results of the current study imply that the subgroups of AIMs have remarkable overlaps considering the symptoms, histopathological, immunohistochemical, EMG, and autoantibody profile. Accordingly, the correct diagnosis and management of these disorders require comprehensive assessments of clinical symptoms, pathological studies and autoantibodies investigations.

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Ethics approval and consent to participate

The study was approved by the Ethical Committee of the Isfahan University of Medical Sciences, Iran (IR.MUI.MED.REC.1401.163). The study protocol was explained to the patients, and they were reassured regarding the confidentiality of their personal information and signed written consent for participation in the study.

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Conflicts of interest

There are no conflicts of interest.

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