2015 Persian Gulf Criteria for Early Diagnosis of Polymyositis/Dermatomyositis

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Abstract

The idiopathic inflammatory myopathies are a group of chronic inflammatory diseases with unknown etiology involving muscle resulting myositis. It may be an autoimmune process and multiple autoantibodies have been detected in this group of disorders. Polymyositis (PM) is a member of this group in which Myositis is the predominant feature of a systemic disease involving muscle (Myositis), heart (Myocarditis, conduction abnormalities, arrhythmia and coronary heart disease), lung (Interstitial lung disease), Gastrointestinal (dysphagia, nasal regurgitation and/or aspiration), joints (arthralgia/arthritis) and so on. Myositis as the cardinal involvement in PM is presented as symmetric proximal muscle weakness. Climbing stairs and getting up with difficulty are the history of these patients. Muscle enzymes including Creatine Kinase (CK), Lactate Dehydrogenase (LDH), aldolase, aspartate aminotransferase (AST) and alanine aminotransferase (ALT) can be increased in patients with myositis. In Dermatomyositis, skin involvement is a major finding along with other manifestations as same as all the features can be seen in PM. As same as other systemic inflammatory Rheumatic diseases the diagnosis of PM/DM can be established by clinical/laboratory judgment of an expert rheumatologist. Bohan and Peter classification criteria for PM/DM has been used since 1975. We know that, Bohan and Peter criteria cannot exactly differentiate the polymyositis from muscular dystrophies and many other myopathies and after that, the discovery of MSA and detection of the role of MRI in diagnosis of muscle inflammation have been introduced. In this letter, the corresponding author wants to deliver a new diagnostic criteria for early detection of PM/DM along with a guideline for approaching to diagnosis of PM/DM.

Keywords
Polymyositis, Dermatomyositis, Bohan and Peter Criteria, Persian Gulf Criteria

1. Introduction

The idiopathic inflammatory myopathies are a group of chronic inflammatory diseases with unknown etiology involving muscle resulting myositis. It may be an autoimmune process and multiple autoantibodies have been detected in this group of disorders (1).

The members of this group of diseases are including (1):
• Polymyositis (PM)
• Dermatomyositis (DM)
• Childhood (Juvenile) PM/DM
•Overlap syndrome
• PM/DM accompanied by malignancies
• Inclusion Body Myositis (IBM)

2. Main Body

In this letter, we want to discuss only about the diagnosis of PM/DM.

Polymyositis is a member of this group in which Myositis is the predominant feature of a systemic disease involving muscle (Myositis), heart (Myocarditis, conduction abnormalities, arrhythmia and coronary heart disease), lung (Interstitial lung disease), Gastrointestinal (dysphagia, nasal regurgitation and/or aspiration), joints (arthralgia/arthritis) and so on (2-5).

Myositis as the cardinal involvement in PM is presented as symmetric proximal muscle weakness. Climbing stairs and getting up with difficulty are the history of these patients. Muscle enzymes including Creatine Kinase (CK), Lactate Dehydrogenase (LDH), aldolase, aspartate aminotransferase (AST) and alanine aminotransferase (ALT) can be increased
in patients with myositis as same as other muscle disorders such as muscle dystrophies and myopathies (5,6).

Autoantibodies seen in patients with myositis are divided into two groups (7):

- Myositis-specific autoantibodies (MSA)
- Myositis-associated autoantibodies (MAA)

The most important autoantibodies belong to MSA are (8):

- Antisynthetase antibodies especially Anti-Jo-1
- Anti-Signal Recognition Particle; Anti SRP
- Anti-Mi-2

The most important autoantibody belongs to MAA is ANA that is positive in up to 80% of patients with PM/DM (9). Others, e.g. anti-Ro, anti La, anti-sm, anti RNP and so on are positive in overlaps of PM/DM with other systemic Rheumatic diseases (10).

Myogenic pattern in electromyography (EMG) can be seen not only in myositis but also in myopathies and muscle dystrophies and normal EMG cannot rule out myositis. It shows neurogenic pattern in neuropathies and mixed pattern of myogenic/neurogenic in Inclusion Body Myositis. Magnetic Resonance Imaging (MRI) can detect all regions of myogenic/neurogenic in Inclusion Body Myositis. The diagnosis of PM/DM can be established by clinical/laboratory judgment of an expert rheumatologist.

Myogenic pattern in electromyography (EMG) can be seen not only in myositis but also in myopathies and muscle dystrophies and normal EMG cannot rule out myositis. It shows neurogenic pattern in neuropathies and mixed pattern of myogenic/neurogenic in Inclusion Body Myositis. Magnetic Resonance Imaging (MRI) can detect all regions of inflammation in whole muscles of the body. But, its findings are not characteristic for myositis and it cannot differentiate the myositis from myopathies and muscle dystrophies (11).

In Dermatomyositis, skin involvement is a major finding along with other manifestations as same as all the features can be seen in PM. These skin findings are including (12):

- Gottron’s sign/papules
- Heliotrope rash
- Shawl sign
- V sign
- Holster sign
- Erythroderma
- Periungual abnormalities
- Mechanic’s hands
- Calcinosis cutis and so on

Gottron’s papule is the erythematous papules can be seen upon extensor surface of hand and/or foot joints especially PIPS, DIPs and MCPs. Gottron’s sign is the erythematous macules/patches or papules on the extensor aspect of hands, elbows, knees and feet.

Heliotrope rash is a violaceous edematous lesion on the upper eyelids with or without erythema spreading toward other facial regions.

Shawl sign is an erythematous eruption upon the upper back.

V sign is an erythematous rash within the V area of the neck and upper chest.

Holster sign is a poikiloderma upon the lateral surfaces of the thighs.

Erythroderma is an erythematous rash involving entire the body skin.

Periungual abnormalities is nail fold telangiectasia (as same as scleroderma and SLE) along with cuticular overgrowth.

Mechanic’s hand is a hand presented with hyperkeratotic and dirty fissured palmar/fingers skin similar to the hand of mechanics (13).

Calcinosis cutis or calcium deposition within the skin is uncommon in adult with dermatomyositis whereas it is common in juvenile DM (14). The Gottron’s papule and Heliotrope rash among all skin features of DM are more common and more characteristic (13). Common pathologic findings of PM/DM regarding muscle involvement are muscle fiber necrosis, degeneration, regeneration and inflammatory cell infiltration (15).

Perifascicular and perivascular infiltration of B-cells can be seen in DM whereas intrafascicular infiltration of cytotoxic CD8+ T-cells can be seen in PM (16).

As same as other systemic inflammatory Rheumatic diseases the diagnosis of PM/DM can be established by clinical/laboratory judgment of an expert rheumatologist.

Bohan and Peter classification criteria for PM/DM has been used since 1975 (17). This criteria are including:

- Symmetric proximal muscle weakness
- Increased serum muscle enzymes
- Myogenic pattern in EMG
- Compatible muscle pathology
- Characteristic skin rash (Gottron’s papules and/or heliotrope rash)

For definite diagnosis of PM, we need all first four criteria whereas the combination of characteristic skin rash and at least three criteria out of four first criteria means definite DM.

We know that, Bohan and Peter criteria can not exactly differentiate the polymyositis from muscular dystrophies and many other myopathies. In Bohan and Peter criteria we need to pathologic evaluation for diagnosis in all the cases of polymyositis and it is not including MSA and muscle MRI.

So, Bohan and Peter criteria is not a good instrument for early diagnosis of polymyositis and it is not accurate for this detection.

3. Conclusion

In this letter we do not want to discuss about sensitivity/specificity and accuracy of Bohan and Peter criteria. On the other hand, corresponding author as the creator of Iran Criteria for early diagnosis of Rheumatoid arthritis, Ankylosing spondylitis and Granulomatosis with polyangiitis, 2015 ACR/SLICC revised criteria for diagnosis of Systemic lupus erythematosus and Chondromalacia Patella (18,19,20,21,22) wants to deliver a new diagnostic criteria (Persian Gulf Criteria) for early detection of PM/DM along with a guideline for approaching to diagnosis of PM/DM presented in table A, B and C respectively.

I would like to ask the members of ACR, EULAR, APLAR and all rheumatologists in the world to evaluate this new criteria. You have to be informed that we could not evaluate this criteria due to funding problems.
Table A. 2015 Persian Gulf Criteria for early diagnosis of Polymyositis (PM) 

<table>
<thead>
<tr>
<th>I. Clinical criteria</th>
<th>II. Laboratory criteria</th>
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<tbody>
<tr>
<td>Symmetric proximal muscle weakness</td>
<td>Elevated muscle enzymes in serum:</td>
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<tr>
<td>First-degree family history of muscle weakness</td>
<td>Including CPK and/or aldolase</td>
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Table B. 2015 Persian Gulf Criteria for early diagnosis of Dermatomyositis (DM) 

<table>
<thead>
<tr>
<th>I. Cutaneous criteria:</th>
<th>II. Muscular criteria:</th>
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</thead>
<tbody>
<tr>
<td>Gottron’s papules or sign</td>
<td>as same as polymyositis (PM)</td>
</tr>
<tr>
<td>Heliotrope rash</td>
<td></td>
</tr>
<tr>
<td>Holster sign</td>
<td></td>
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<tr>
<td>Shawl sign</td>
<td></td>
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<tr>
<td>V sign</td>
<td></td>
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<tr>
<td>Mechanics’ hands</td>
<td></td>
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<tr>
<td>Periungual capillary abnormality</td>
<td></td>
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<td>2 points</td>
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Table C. Amir Alam Hospital Guideline for approaching to diagnosis of PM/DM.

**Step I:**
- History and physical examination by an expert Rheumatologist in cooperation with a Neurologist and/or Dermatologist if it is necessary
- CBC, ESR, BUN/Cr, U/A, Ca/P, Alk-ph, serum Vitamine D level
- Muscle enzymes: CPK, LDH, Aldolase, AST, ALT
- Anti-jo-1, Anti-SRP, Anti-Mi-2
- PA-CXR
- If there is a overlap syndrome: check MAA including: ANA, RF, Anti-RNP, Anti-Sm and so on

**Step II:**
- EMG of proximal muscles
- MRI of skeletal muscles

**Step III:**
- Proximal muscle biopsy for evaluation of pathology

In each step, the diagnosis of PM/DM by using new diagnostic criteria can be confirmed or ruled out, the approach to diagnosis must be stopped.
References


