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2015 Persian Gulf Criteria for Early Diagnosis of Polymyositis/Dermatomyositis

Iraj Salehi-Abari

Rheumatology Research Center, Amir Alam Hospital, Tehran University of Medical Sciences, Tehran, Iran

Email address

salehiabari@sina.tums.ac.ir

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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 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 thigs.

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 Criterias 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) [a].*

I. Clinical criteria	
• Symmetric proximal muscle weakness [b]	1 point
• First-degree family history of muscle weakness[c]	-1 point
II. Laboratory criteria	
• Elevated muscle enzymes in serum:	1 point
Including CPK and/or aldolase	
• Myositis specific autoantibodies (MSA)[d]:	2 points
Including Anti-Jo-1 and/or Anti-SRP and/or Anti-Mi-2	
• Electromyograohy (EMG): Myogenic pattern	1 point
• Electromyograohy (EMG): Neurogenic pattern	-1 point
• Muscle inflammation:	Up to 1 point
Inflammation signaling in muscle MRI or	1 point
Muscle pathology showing lymphocyte infiltration and/or vasculitis	1 point

a: We need at least 4 points including one point from domain I for definite diagnosis of Polymyositis. With 3 points including one point from domain I, the diagnosis of PM is highly suggestive and with 2 points including one point from domain I, it is probable.

b: No other etiology including drugs, metabolic/endocrine disease, muscular dystrophies can better explain this weakness upon history and physical examination.

c: Positive family history of muscle weakness is in favor of muscle dystrophies and against polymyositis.

d: They are not seen in muscular dystrophies and other myopathies.

Table B. *2015 Persian Gulf Criteria for early diagnosis of Dermatomyositis (DM) [a,b].*

I. Cutaneous criteria:	
• Gottron's papules or sign	2 points
• Heliotrope rash	2 points
• Holster sign	1 point
• Shawl sign	1 point
• V sign	1 point
• Mechanics' hands	1 point
• Periungual capillary abnormality	1 point
II. Muscular criteria:	
as same as polymyositis (PM)	

a: For definite diagnosis of DM, we need at least 4 points including at least 2 points from cutaneous criteria and at least one point from muscular criteria.

b: The presence of Gottron's papule (or sign) and/or Heliotrope rash without any muscular criteria is called Amyopathic Dermatomyositis that is suggested after 6 months and it is confirmed after 2 years.

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 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

[1] Hoogendijk JE, Amato AA, Lecky BR, et al. 119th ENMC international workshop: trial design in adult idiopathic inflammatory myopathies, with the exception of inclusion body myositis, 10-12 October 2003, Naarden, The Netherlands. NeuromusculDisord 2004; 14:337.

[2] Denbow CE, Lie JT, Tancredi RG, Bunch TW. Cardiac involvement in polymyositis: a clinicopathologic study of 20 autopsied patients. Arthritis Rheum 1979; 22:1088.

[3] Lundberg IE. The heart in dermatomyositis and polymyositis. Rheumatology (Oxford) 2006; 45 Suppl 4: iv18.

[4] deMerieux P, Verity MA, Clements PJ, Paulus HE. Esophageal abnormalities and dysphagia in polymyositis and dermatomyositis. Arthritis Rheum 1983; 26:961.

[5] Katzap E, Barilla-LaBarca ML, Marder G. Antisynthetase syndrome. Curr Rheumatol Rep 2011; 13:175.

[6] Hochberg MC, Feldman D, Stevens MB. Adult onset polymyositis/dermatomyositis: an analysis of clinical and laboratory features and survival in 76 patients with a review of the literature. Semin Arthritis Rheum 1986; 15: 168.

[7] Targoff IN. Myositis specific autoantibodies. Curr Rheumatol Rep 2006; 8:196.

[8] Gunawardena H, Betteridge ZE, McHugh NJ. Myositis-specific autoantibodies: their clinical and pathogenic significance in disease expression. Rheumatology (Oxford) 2009; 48:607.

[9] Love LA, Leff RL, Fraser DD, et al. A new approach to the classification of idiopathic inflammatory myopathy: myositis-specific autoantibodies define useful homogeneous patient groups. Medicine (Baltimore) 1991; 70:360.

[10] Reichlin M, Arnett FC Jr. Multiplicity of antibodies in myositis sera. Arthritis Rheum 1984; 27:1150.

[11] May DA, Disler DG, Jones EA, et al. Abnormal signal intensity in skeletal muscle at MR imaging: patterns, pearls, and pitfalls. Radiographics 2000; 20 Spec No:S295.

[12] Callen JP. Cutaneous manifestations of dermatomyositis and their management. Curr Rheumatol Rep 2010; 12:192.

[13] Marvi U, Chung L, Fiorentino DF. Clinical presentation and evaluation of dermatomyositis. Indian J Dermatol. 2012; 57(5): 375-81.

[14] Gunawardena H, Wedderburn LR, Chinoy H, et al. Autoantibodies to a 140-kd protein in juvenile dermatomyositisare associated with calcinosis. Arthritis Rheum 2009; 60:1807.

[15] Drake LA, Dinehart SM, Farmer ER, et al. Guidelines of care for dermatomyositis. American Academy of Dermatology. J Am Acad Dermatol 1996; 34:824.

[16] Krathen MS, Fiorentino D, Werth VP. Dermatomyositis. Curr Dir Autoimmun. 2008; 10: 313-32.

[17] Bohan A, Peter JB. Polymyositis and dermatomyositis (second of two parts). N Engl J Med 1975; 292:403.

[18] Salehi I, Khazaeli S, Khak M. Early diagnosis of rheumatoid arthritis: an introduction to the newly designed Iran Criteria for Rheumatoid Arthritis. Rheumatol Int. 2013; 33(1): 45-50

[19] Salehi-Abari I, Khazaeli S, Khak M. Early diagnosis of ankylosing spondylitis: an introduction to the newly designed Iran criteria for ankylosing spondylitis. Rheumatol Int. 2013; 33(5): 1303-8

[20] Salehi-Abari I, Khazaeli S, Khak M, Motesaddi M, Hasibi M. Early diagnosis of granulomatosiswith polyangiitis: an introduction to the newly designed Iran criteria for early diagnosis of granulomatosiswith polyangiitis. Indian Journal of Rheumatology. 2013

[21] Salehi-Abari I. 2015 ACR/SLICC Revised Criteria for Diagnosis of Systemic Lupus Erythematosus. Autoimmune Dis Ther Approaches Open Access. 2015; 2:114

[22] Salehi-Abari I, Khazaeli S, Niksirat A. Chondromalacia Patella and New Diagnostic Criteria. Open Science Journal of Clinical Medicine. 2015; 3(4):126-8.