Diagnostic criteria for dermatomyositis (DM)

Bohan and Peter's criteria
(Bohan A and Peter JB, N Engl J Med. 292:344-7, 1975; Bohan A and Peter JB, N Engl J Med. 292:403-7, 1975)

The diagnosis of DM is considered definite, probable and possible when skin rash is associated with 3, 2 or 1 muscular criteria, respectively*:

1- **Symmetric proximal muscle weakness** determined by physical examination
2- **Elevation of serum skeletal muscle enzymes**, including creatine kinase, aldolase, serum glutamate oxaloacetate and pyruvate transaminases, lactate dehydrogenase
3- The **electromyographic triad** of short, small, polyphasic motor unit potentials; fibrillations, positive sharp waves, and insertional irritability; and bizarre, high-frequency repetitive discharges
4- **Muscle biopsy** abnormalities of degeneration, regeneration, necrosis, phagocytosis, and an interstitial mononuclear infiltrate
5- **Typical skin rash of DM**, including a heliotrope rash and Gottron's sign/papules

**Exclusion criteria: central or peripheral neurologic diseases, muscular dystrophies, granulomatous and infectious myositis, metabolic and endocrine myopathies, and myasthenia gravis.*

Dalakas and Hohlfeld's criteria
(Dalakas MC and Hohlfeld R, Lancet 362:971-82, 2003)

Criterion	**Myopathic DM**		**Amyopathic DM**
	Definite	**Probable**	**Definite**
Myopathic muscle weakness	Yes	Yes	No
Electromyographic findings	Myopathic	Myopathic	Myopathic or non-specific
Serum skeletal muscle enzymes	High or normal	High	High or normal
Muscle biopsy findings	Perifascicular, perimysial or perivascular infiltrates; perifascicular atrophy		Non-specific or diagnostic for DM
Rash or calcinosis	Present	Not detected	Present

Note:
The Bohan and Peter's criteria, Dalakas and Hohlfeld's criteria and the European NeuroMuscular Centre (ENMC) classification criteria (Hoogendijk JE et al., Neuromusc Disord 14:337-45, 2004) are discussed in the following article: Iaccarino L. et al., J. Autoimmunity 48-49:122-127, 2014.