Supplementary data

Materials and methods

Additional items in the EUSTAR-online myopathy sub-study

Medical history: date of onset of muscle symptoms, patient's assessment of global muscle activity visual analog scale (VAS) (mm), proximal dysphagia, dysphonia, cardiac arrhythmias, pericarditis/myocarditis, osteoporotic fractures including vertebral collapse (excluding avascular necrosis), malignancy newly diagnosed within the last 5 years, proximal muscle weakness not explainable by other causes, mechanic's hands; physical exams: physician's assessment of global muscle activity VAS (mm), physician's assessment of global muscle damage VAS (mm), manual muscle test for 8 muscle groups (MMT-8), functional index-2 (FI-2); laboratory parameters: Jo-1, Ku, Mi-2, SRP, PL-7, PL-12, OJ, EJ, p155/140, MDAS, NXP2, Serum creatine, aldolase, aspartate aminotransferase (AST), alanine aminotransferase (ALT), lactate dehydrogenase (LDH), creatine kinase myocardial band (CK-MB), Troponin-T, Troponin-I, Phosphate, Sodium, Potassium, Calcium, thyroid-stimulating hormone (TSH), 25-OH-Vitamin D; diagnostic tools: electromyography (EMG), magnetic resonance imaging (MRI), muscle biopsy.

Inclusion criteria for myopathy sub-study

Any of the following criteria is sufficient for inclusion:

- Elevated levels of the serum muscle enzymes CK or aldolase above upper limit of normal according to local laboratory without obvious explanation (*e.g.*, extensive exercise, intramuscular injections, muscle injury)
- Proximal muscle weakness on physical examination as judged by the physician not explainable by other causes (*e.g.,* neuropathogenic, genetic, metabolic, endocrinologic, infectious disorders, drug-induced, cachexia)
- Muscle atrophy on physical examination as judged by the physician not explainable by other causes (*e.g.*, neuropathy, genetic disorders, cachexia)
- Positive myositis-associated autoantibodies (Jo-1, PM-Scl, U1-RNP, Ku, Mi-2, SRP, PL-7, PL-12, OJ, EJ, p155/140, MDA5, MXP2).

Supplementary table S1. Matching criteria for SSc patients with and SSc patients without myopathy.

Characteristics	Patients with myopathy	Patients without myopathy
Sex		
Female	43/58 (74.1)	43/58 (74.1)
Male	15/58 (25.9)	15/58 (25.9)
Age (years)	59.7 (49.0, 68.3)	59.9 (48.4, 68.6)
Extent of skin involvement		
Limited cutaneous involvement	37/58 (63.8)	38/58 (65.5)
Diffuse cutaneous involvement	18/58 (31.0)	18/58 (31.0)
Systemic sclerosis *sine* scleroderma	3/58 (5.2)	2/58 (3.4)
Disease duration		
>=5 years	35/58 (60.3)	35/58 (60.3)
<5 years	23/58 (39.7)	23/58 (39.7)
Scleroderma-antibodies[1]	48/58 (82.8)	47/58 (81.0)

For nominal variables, the absolute and relative frequencies are shown: n/total valid cases (%). Continuous variables are described as median and 1st, 3rd quartiles (Q1, Q3).
[1]Scleroderma antibodies include anticentromere, antitopoisomerase-I, and anti-RNA polymerase III.