



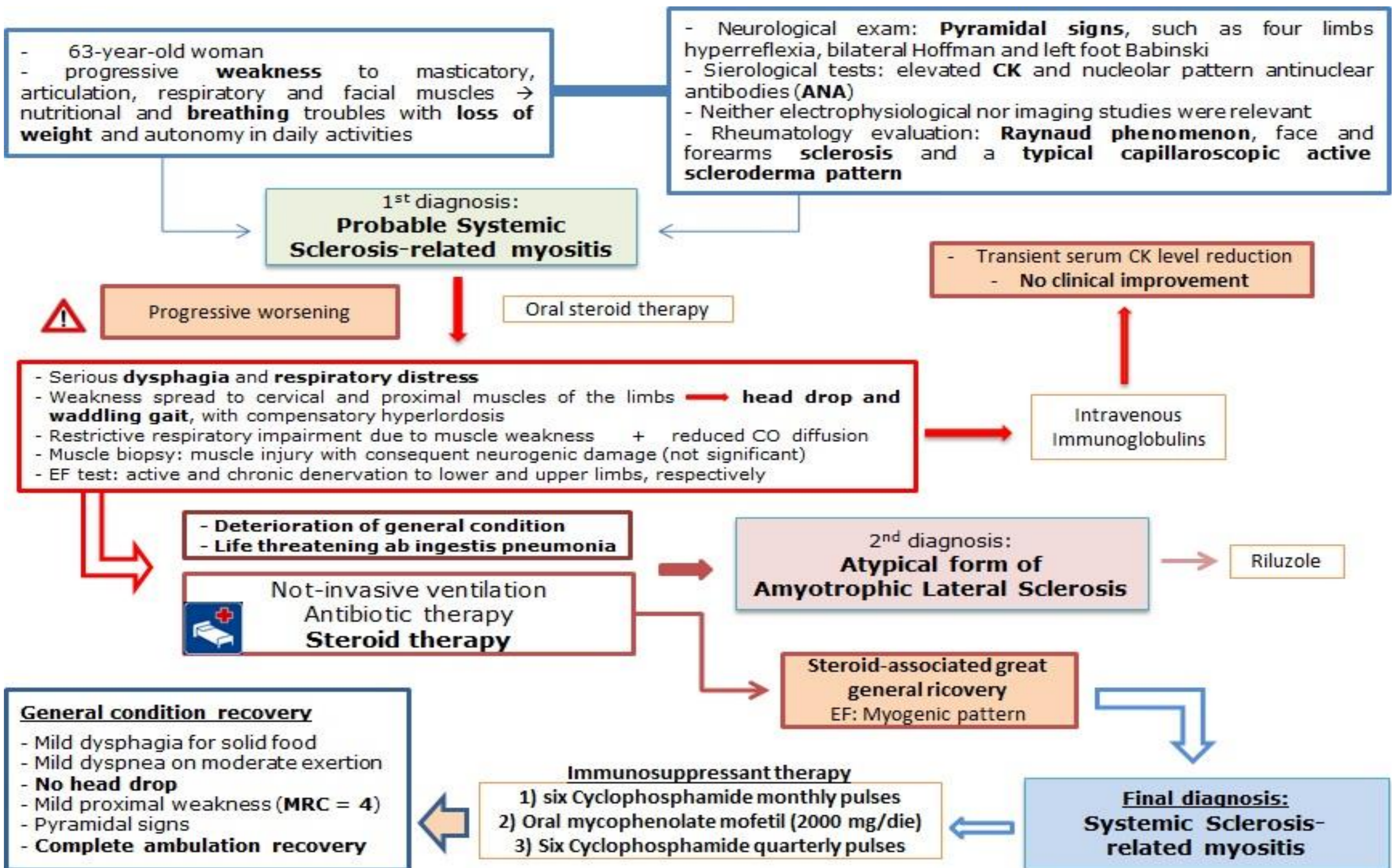
# Atypical Systemic Sclerosis-related myositis: report of a case



Massa F, Lapucci C, Caponnetto C, Mancardi GL

Department of Neurosciences, Rehabilitation, Ophthalmology, Genetics, Maternal and Child Health (DINO GMI., University of Genoa, Italy)

**Background:** Systemic Sclerosis (Scc) is a rare, autoimmune disorder, characterized by inflammation, vascular injury, and fibrosis of skin and internal organs. Recent studies reported up to 40% neurological consequences in the disease, especially involving PNS, particularly muscles. The clinical presentation of SSc-associated myopathy shares many features with polymyositis or dermatomyositis, as the weakness is symmetrical, especially in proximal muscles, and the histopathological pattern often shows inflammation, along with endomysial microangiopathy and interstitial fibrosis.



**Discussion:** The peculiarity of our case can be found in the atypical Systemic Sclerosis presentation which led to a misdiagnosis. Rare cases of scleroderma-associated Amyotrophic Lateral Sclerosis are described, even if a clear link between those diseases have not been found. The pyramidal signs which led to a Motor Neuron disease diagnosis, along with respiratory and bulbar muscles distress and electrophysiological abnormalities, could be reconducted to chronic medulla ischemia due to vascular injury or immune mechanism as some authors proposed. It has been estimated that up to 10% of patients initially diagnosed as having ALS are afterwards rediagnosed as having a disease other than ALS, included in the so called 'amyotrophic lateral sclerosis (ALS) mimic syndrome'.

**Conclusion:** In our case correct diagnosis and therapies had great impact on patient's prognosis and quality of life. As the proper diagnosis of ALS remains a complex clinical problem and brings a lot of further implications, recognizing the neurological involvement of those mimicking diseases – i.e. Systemic Sclerosis we described – has to be a critical goal.

## References

- Amaral TN, Peres FA, Lapa AT, Marques-Neto JF, Appenzeller S. **Neurologic involvement in scleroderma: a systematic review.** *Semin Arthritis Rheum.* 2013 Dec;43(3):335-47.
- Ranque B, Authier FJ, Berezne A, Guillevin L, Mouthon L. **Systemic sclerosis-associated myopathy.** *Ann N Y Acad Sci.* 2007 Jun;1108:268-82.
- Ouhabi H, Bourazza A, Rouimi A, Boutaleb N, Mosseddaq R. **Association of amyotrophic lateral sclerosis with scleroderma. Study of 2 cases.** *Rev Neurol (Paris).* 1997 Dec;153(12):790-1.
- B. J. Traynor BJ, Codd MB; Corr B, Forde C, Frost E; Hardiman O. **Amyotrophic Lateral Sclerosis Mimic Syndromes: A Population-Based Study.** *Arch Neurol.* 2000;57(1):109-113