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Clinical phyenotypic characterization of dermatomyositis with interstitial lung disease presence

PABLO ARTURO OLIVO PALLO (HOSPITAL DAS CLÍNICAS FMUSP, Sao Paulo, SP, Brasil), LEONARDO HOFF (HOSPITAL DAS CLÍNICAS, Sao Paulo, SP, Brasil), SAMUEL KATSUYUKI SHINJO (HOSPITAL DAS CLÍNICAS FMUSP, Sao Paulo, SP, Brasil)

BACKGROUND

Dermatomyositis (DM) is a rare systemic autoimmune myopathy with variety of phenotypic presentations, including lung involvement. Herein, we initially analyzed the prevalence of the interstitial lung disease (ILD) in patients with DM. We also characterized the predictive factor associated with this ILD.

MATERIALS AND METHODS

This is a retrospective, single-center study that included 70 consecutive adult patients with defined classical DM and clinically amyopathic DM (EULAR/ACR classification criteria 2017), and was conducted from 2003 to 2019. All analyzed data were collected from electronic medical records, with pre-standardized and parameterized information. The ILD was based on altered pulmonary images obtained by computer tomography at the disease's onset.

RESULTS

The patients' mean age was 42.2±11.5 years, and they were predominantly female (64.2%) and white (92.9%). The ILD was identified in 35 (50%) patients: 25 (45.4%) out of 55 in classical DM and 10 (66.6%) out of 15 in clinically amyopathic DM. The following parameters were equally distributed between the patients with versus without interstitial lung disease: kind of disease (classical or clinically amyopathic DM), mean age at the disease's onset, constitutional symptoms (baseline), cutaneous cumulative lesions (heliotrope rash, Gottron's sign, Gottron' papules, facial rash, Shawl's sign, V-neck' sign, periungual alterations, photosensitivity, calcinosis, ulcers, mechanic's hands), muscle involvement (myalgia and limb muscle weakness), joint involvement (arthralgia or arthritis), gastrointestinal tract involvement (vasculitis or dysphagia), and serum level of muscle enzymes. In contrast, patients with ILD had higher frequencies of cutaneous vasculitis than patients without ILD (odds ratio of 3.20, confidential interval of 95%: 1.05-9.69, P=0.03).

CONCLUSION

This study shows a high prevalence of ILD in patients with DM. The ILD is associated with cutaneous vasculitis at disease's onset. Further studies are needed to corroborate to our data, include analysis of myositis-specific autoantibodies.