SBR 2019
36º CONGRESSO BRASILEIRO DE REUMATOLOGIA
CENTRO DE EVENTOS DO CEARÁ
04 A 07 DE SETEMBRO
PROMOÇÃO
70 anos Sociedade Brasileira de Reumatologia
REALIZAÇÃO
SCR SOCIEDADE CEARENSE DE REUMATOLOGIA

PROGRESSION TO SYSTEMIC SCLEROSIS IN A GROUP OF SECONDARY RAYNAUD PHENOMENON WITHOUT SKIN INVOLVEMENT: FOLLOW-UP IN A LARGE SINGLE BRAZILIAN COHORT.

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BACKGROUND

With the growing need of early diagnosis of systemic sclerosis (SSc), many strategies were developed in the last decade to predict patients with Raynaud phenomenon (RP) who will develop SSc. The objective of this study was to analyze predictors for development of SSc in a cohort of patients with secondary RP without skin involvement.

MATERIALS AND METHODS

This is a single-center retrospective analysis of consecutive patients presenting RP without skin involvement and at least one SSc manifestation (specific SSc autoantibody, specific SSc visceral involvement or SD pattern at nailfold capillaroscopy) who attended a scleroderma outpatient clinic between 2010 and 2019. Data were obtained from an electronic register database. Statistical significance was set up as $p < 0.05$.

RESULTS

Among the 215 patients who were considered for investigation and follow up, 151(70.2%) were classified as SSc according to ACR/EULAR classification criteria after a median (interquartile range) of 4(2-7) years. Among SSc patients, 37(24.5%)

already fulfilled SSc classification criteria after the initial investigation with a median time from RP to diagnosis of 2(0-4) years. When compared to the group who did not fulfill the SSc classification criteria (n=64; 29.8%), SSc patients presented significantly

higher frequency of puffy fingers (57%vs.4%; p<0.0001) and lower frequency of esophageal dysmotility disorder (3.3%vs.28.1%; p<0.0001) as first non-Raynaud symptoms. In contrast, arthritis (6% vs.12.5%), interstitial lung disease (ILD)(16.6% vs.17.2%) and pulmonary hypertension (PH)(5.3%vs.1.6%) were found in similar frequencies among the groups (p>0.05), despite of comparable median follow up in both groups [4(2-7) vs.4(2-7); p=0.81]. Moreover, SSc patients had more SD pattern (83.4%vs.56.3%; p<0.0001), anti-Scl70 antibody (16.6%vs.3.1%; p=0.006), and nucleolar ANA pattern (22.5%vs.10.9%; p=0.048). During a comparable median follow

up [4(2-7)vs.4(2-7); p=0.81], patients with SSc referred more frequently: pitting scars (85.4%vs.6.3%, p<0.0001), digital ulcers (23.8%vs.3.1%, p<0.0001), telangiectasias (51%vs.9.4%, p<0.0001), esophageal dysmotility disorder (58.9%vs.42.2%, p=0.024),

and ILD (66.2%vs.34.4%,p<0.0001), in comparison to the secondary RP patients without SSc.

CONCLUSION

A significant number of patients presenting secondary RP without skin involvement fulfilled the 2013 ACR/EULAR classification criteria for SSc in the first 4 years of follow up (¼ of them in the first year). At presentation of disease, puffy fingers, SD pattern, anti-Scl70 antibody, and nucleolar ANA pattern were predictors of SSc, while esophageal dysmotility disorder was protective, reinforcing the strength of the 2013 SSc classification criteria in the characterization of SSc patients without skin involvement.