ULNAR OCCLUSION IS A MARKER OF GLOBAL VASCULAR DAMAGE IN SYSTEMIC SCLEROSIS: RESULTS FROM A MONOCENTRIC PROSPECTIVE STUDY OF 99 PATIENTS

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Background: Macrovascular damages of systemic sclerosis (SSc) were poorly investigated, and the link between macrovasculopathy and microvasculopathy of SSc, cardiovascular disease, and mortality remain unclear.

Objectives: To evaluate if macrovascular damage in SSc predics the occurrence of new digital ulcers (DU), cardiovascular events and mortality, and to further assess the relationship between micro and macrovascular damages in SSc.

Methods: All consecutive SSc patients followed in our SSc National Reference Centre, who underwent an arterial doppler ultrasonography (aDUS) of the upper limbs, were included and prospectively followed up until October 2017. Inclusion criteria were: 1) adults; 2) a diagnosis of SSc according to 2013 ACR/EULAR criteria; 3) aDUS performed in our vascular exploration department.

Results: Ninety-nine SSc patients were included. Median follow-up duration was 35 (IQR, 21 to 39) months. Macrovascular damages mainly affected ulnar arteries, with ulnar artery occlusion (UAO) in 28 (28.3%) patients (bilateral 60.7%). New DU occurred in 25 (27.1%) patients, new cardiovascular event in 10 (10.4%) patients, and 11 patients died during the study period. Interestingly, UAO was not associated with traditional cardiovascular risk factors (except dyslipidemia) nor with history of cardiovascular diseases, and was not predictive of new cardiovascular events. Conversely, UAO was associated with markers of microvascular damages, such as late nailfold capillaroscopy pattern (33.3% vs 6.8%; OR=6.88, 95%CI=2.54–18.8; HR=2.23, 95%CI=1.02 to 4.86; p=0.037), pleading for a SSc specific vasculopathy.

Conclusions: Our study confirms that macrovascular damages are frequent in SSc patients and mainly affect ulnar arteries. Interestingly, UAO was associated with markers of microvascular damages, but not with markers of cardiovascular diseases.

Disclosure of Interest: None declared


RACIAL DIFFERENCES IN SSc DISEASE PRESENTATION: A EUROPEAN SCLERODERMA TRIALS AND RESEARCH GROUP STUDY

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Background: Genetic and environmental factors play a significant role in systemic sclerosis (SSc). African Americans are known for a higher SSc incidence, an earlier age of onset, and in black patients, a higher prevalence of limited SSc disease (LSSc) compared to whites. Data on blacks mostly stem from African American studies on SSc in Asians are mostly from outside Asia and lack direct comparison with other racial groups.

Objectives: We aimed to evaluate differences of SSc presentations between Asian, black and white patients.

Methods: Characteristics of self-reported white, Asians or black SSc patients from the EUSTAR cohort were compared across racial groups; survival and multi-ple logistic regression analyses were used to adjust for age, sex, disease duration and antibody status.

Results: 9162 white, 341 Asian and 161 black patients were included. Of the Asian patients, 208 stem from within Asia and 133 from 34 centres outside Asia; of the black patients 65 stem from within Africa and 116 from 35 centres outside Africa. Asian and black patients were on average 10 years younger than white patients (p<0.001). Black patients developed the first non-Raynaud’s phenomenon (RP) feature of SSc faster than Asian and white patients (all p<0.01; figure 1) also after adjustment for hazard ratio (HR)[blacks] 1.4, p=0.001; HR[Asians] 1.1, p=0.13 vs whites).

Among ANA specificities, ACA predominated in white patients (whites: 40%; Asians: 16%; blacks: 10%; p<0.001) and Sc-70 in Asian patients (whites: 34%; Asians: 46%; blacks: 32%; p<0.001). The prevalence of diffuse skin involvement was similar in Asian (28%) and white patients (27%), but more common in black patients univariately (59%; p<0.001) ; however in multivariable analysis Asian patients were less likely to have diffuse SSc than white patients (OR 0.6,p<0.001) while black patients were more likely (OR 2.9, p<0.001).

The prevalence of PH (defined as PAPsys >40 mmHg as estimated by echocardiography) was similar in the three groups (whites: 13%; Asians: 17%; blacks: 14%; p=0.10) ; however multivariably, Asians were more likely to have PH (OR [Asians] 4.0, p<0.001, OR[blacks] 2.5, p=0.001 vs whites). Asians had a higher prevalence of an impaired diffusing capacity for carbon monoxide (DLCO<80% of predicted; 84%) than black (72%) or white patients (69%, p<0.001) also in multivariable analysis (OR[Asians] 3.0, p<0.001, OR[blacks] 1.2, p=0.36 vs whites). Both, Asians (43%) and black patients (58%), had a higher prevalence of a reduced forced vital capacity (FVC<80% of predicted) compared to white patients (23%, p<0.001) univariably and multivariably (OR[Asians] 2.4, p<0.001, OR[blacks] 4.0, p<0.001 vs whites).