AB0784

MYOCARDIAL FIBROSIS DETECTED BY MAGNETIC RESONANCE IMAGING IN SYSTEMIC SCLEROSIS—PATHOPHYSIOLOGICAL SIGNIFICANCE

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Background: Systemic sclerosis (SSc) is characterised by chronic fibrosis in various organs such as skin, lung and heart leading to poor prognosis. Myocardial fibrosis is one of factors of pulmonary hypertension (PH) as well as pulmonary arterial hypertension or interstitial lung disease (ILD). Detection of cardiac lesions has progressed much using imaging techniques such as echocardiography (UCG) in combination with measurement of various biomarkers such as BNP. Recently, cardiac magnetic resonance imaging (CMR) has been shown to be sensitive to detect even subclinical cardiac lesions. However, it is unclear what sub-type of SSc is prone to myocardial fibrosis, when it manifests in the clinical course, or whether fibrosis occurs simultaneously in various organs.

Objectives: To clarify the pathophysiological significance of myocardial fibrosis in SSc, CMR was performed in the patients with limited (lc) and diffuse (dc) cutaneous SSc with or without PH.

Methods: Twelve patients (male 2, female 10) who fulfilled ACR/EULAR criteria (2013) for SSc were enrolled. Eight patients were diagnosed as having lcSSc and 4 dcSSc. In addition to CMR, chest CT scan, UCG and laboratory tests including serum autoantibodies specific for SSc, blood brain natriuretic peptide (BNP) and pulmonary function test (%FVC,%DLCO) were performed in all patients. Right heart catheterization was performed in patients whose systolic right ventricular pressure estimated by UCG was higher than 30 mmHg. Positivity of late gadolinium enhancement (LGE) was compared with clinical findings and these parameters. Difference between the patient groups were tested using Student's t-test.

Results: LGE was positive in 6 out of 12 patients. Patient composition of dc/lc in LGE (+) and LGE (-) group was 1/5 and 3/3, respectively. Complication of ILD was present in 3 among LGE (-) patients, while esophageal involvement in 3 among LGE (+) patients. The mean age of LGE (+) group tended to be higher than that of LGE (-) group (73.8±5.8 vs 68±6.4), duration of disease (year) tended to be shorter in LGE (+) group than those of LGE (-) group (1.8±3.0 vs 7.7±5.5), and BNP level (170.6±150.1 vs 90.1±69.0 pg/ml) and RVP (33.5±8.1 vs 29.7±8.1 mmHg) tended to be higher in LGE (+) than in LGE (-) group, although difference was not statistically significant. There was no tendency of positivity of autoantibodies and pulmonary function test, while patients with low%DLCO (<70) and without ILD were positive for LGE. LGE tended to be positive in the patients whose E/e' ratio determined by UCG or pulmonary capillary wedge pressure by RHC was elevated.

Conclusions: Since myocardial fibrosis was found rather in the cases without ILD, there might be difference in the progression of fibrosis depending on the organ, although it accelerates by ageing. If LGE is seen, PH, especially that associated with left heart disease, might occur in future. This study suggests that CMR might be useful to detect cardiac lesions from early period of clinical course, as well as in the cases with some abnormalities in biomarkers such as BNP or DLCO regardless of existence of ILD, although further study is needed to clarify the indication of CMR using more cases of SSc.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.1850

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RED CELL DISTRIBUTION WIDTH IS A PROMISING MARKER OF PULMONARY ARTERIAL HYPERTENSION IN SCLERODERMA-RELATED DISORDERS

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Background: The early identification of those Systemic Sclerosis (SSc) patients harbouring Pulmonary Arterial Hypertension (PAH) is a mainstay in the management of SSc. Novel biomarkers might improve the specificity of screening algorithms currently available.

Objectives: In this study we aimed to test the potential diagnostic value of Red cell distribution width (RDW), which has been previously described as a potential prognostic marker in thromboembolic and idiopathic PAH.

Methods: We prospectively recruited 118 patients affected by Scleroderma related disorders (Mixed connective tissue disease N. 7, 5.9%; SSc N. 93, 78.8%; Scleroderma overlap syndromes N. 11, 9.3%, undifferentiated connective tissue

diseases N. 7, 5.9%) in a PAH outpatient clinic of a University Hospital. All patients underwent an extensive clinical and laboratory evaluation and an echocardiographic examination; a right heart catheterization was performed in 13 patients according to clinical indication.

Results: According to echocardiography and right heart catheterization, 14/118 patients were diagnosed with PAH. Patients affected by PAH had a higher RDW (15 [13.4–18.9] vs. 13.9 [12.1–19.1]; p=0.005), despite similar haemoglobin levels between groups. Patients affected by PAH also showed a higher BNP plasma concentration (245.6 [81.6–494.3] vs. 60.3 [50.3–77.4]; p=0.0001) and lower platelets count [206 188–243 × 10³/μl vs 240 [198–299] × 10³/μl; p=0.04]. RDW was also directly related to ultrasound-assessed Pulmonary Artery Pressure (PAPs) (ρ=0.20; p=0.03) and DLCO corrected for haemoglobin (ρ=-0.30; p=0.001); finally, RDW was related to the mean PAP measured by right heart catheterization (ρ=0.65; p=0.015).

Conclusions: RDW is an inexpensive, potentially useful marker in the detection of SSc-related PAH; its inclusion in screening algorithms could be considered to further improve their diagnostic performance.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5756

AB0786

ROLE OF CALCIUM ANTAGONISTS IN THE CANCER OF SYSTEMIC SCLEROSIS, AN ASSOCIATION UNDER DISCUSSION

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Background: The use of calcium channel blockers (CCB) is widespread in systemic sclerosis (SSc) for the treatment of Raynaud's phenomenon, present in 95% of patients. CCB can alter apoptosis, a mechanism for the destruction of cancer cells. In SSc the risk of cancer is increased, but the role that CCB can play is not clear, with contradictory data obtained so far.^{1,2}

Objectives: To analyse the role of CCB in the appearance of cancer in a cohort of patients with systemic sclerosis.

Methods: Encoded patients under the diagnosis of SSc in our hospital from 1985 until December of 2017 were collected. Medical records were reviewed, recording clinico-epidemiological data and treatments used.

Results: 120 patients have been diagnosed of SSc (103 females, 17 males), 22 of whom (18.3%) have developed cancer. The type of cancer were (in order of frequency): breast (12 patients, 57%), gastrointestinal (4 patients, 33%), one hypernephroma, one endometrial carcinoma, one lymphoma, one skin cancer and one epidermoid of tongue. Adenocarcinoma was the most frequently histologic pattern found (16 patients, 76%). Interestingly, the diagnosis of SSc was made at an older age in those patients diagnosed of cancer (66.4±11.7 vs 54.8±17.3 years old, p=0.019); An age older than 55 years old conferred a relative risk (RR) of cancer of 1.69^{1,26-2.25} in the global cohort and a RR of 3.26^{1,18-8.99} in the female cohort. Although the association cancer and CCB did not reach statistical significance, the association CCB and female patients older than 55 years elevate de RR of cancer to 4.16 (0.96–17.95). All other characteristics (epidemiological, clinical, analytical and another treatments) analyzed did not reach statistical significant differences.

Conclusions: CCB increase in our series the risk of cancer in women over 55 years old. CCB could be implicated in the pathogenesis of cancer in SSc. Due to the broad use of this in the SSc population, it may be require to study in a large number of patients in order to explore this association.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4279