ANTI-KS AUTOANTIBODY IS ASSOCIATED WITH SICCA SYNDROME AND INTERSTITIAL LUNG DISEASE

Y. Kondo, S. Sasaki, T. Kurabayashi, Y. Koyama, S. Sato. Division of Rheumatology, Department of Internal Medicine, Tokai University School of Medicine, Kanagawa, Japan

Background: Anti-aminocarboxyl-tRNA synthetase (anti-ARS) autoantibodies have been found in patients with polymyositis/dermatomyositis (PM/DM). Anti-KS, an anti-ARS antibody, often presents with interstitial lung disease (ILD) without other clinical symptoms of connective tissue disease (CTD). However, the clinical manifestations of anti-KS-positive patients are not well studied.

Objectives: To clarify the clinical and laboratory characteristics of Japanese patients with antibodies against anti-KS antibody.

Methods: Sera from 326 patients with CTD (including PM/DM) or ILD at Tokai University were screened for anti-KS antibody using RNA and protein immunoprecipitation assays. Demographic data, clinical symptoms, laboratory findings and chest computed tomography (CT) scan results were retrospectively reviewed from medical charts.

Results: Five patients with anti-KS autoantibody were identified. All five patients were female with a mean age (±SD) of 59.4 (±13.9) years at onset, presenting respiratory symptoms without any sign of myositis. In all patients, ILD was chronic and chest CT scan revealed a non-specific interstitial pneumonia pattern in three patients and the remaining two showed a usual interstitial pneumonia pattern. Three patients (60%) had arthritis, mechanic’s hand, while one (20%) had Gottron’s sign and was diagnosed as amyopathic DM with ILD. Interestingly, three patients (60%) showed symptoms of the Sicca syndrome with presence of anti-SSA antibody and two of those were diagnosed as Sjögren’s syndrome. The frequency of Sicca syndrome in anti-KS-positive patients was significantly higher compared with other anti-ARS antibody-positive patients (80% vs. 14%, respectively, p=0.031).

Conclusions: Results highlight the presence of anti-KS antibody is associated with the Sicca syndrome as well as ILD without muscle symptoms.

REFERENCES:

Disclosure of Interest: None declared

AB0821 EARLY DETECTION OF THE CHANGES IN PULMONARY ARTERIAL PRESSURE AND VASCULAR FUNCTIONS IN SYSTEMIC SCLEROSIS: EXPLORING NON-INVASIVE CLINICAL TESTS AND UNDERLYING GENE EXPRESSIONS

Y. Koyama1, S. Fuke2, T. Ohno3, Y. Sato4, T. Higuchi5. 1.Centre for Autoimmune Diseases, Division Of Rheumatology, 2.Department of Cardiology, 3.Department of Dermatology, Japanese Red Cross Okayama Hospital, Okayama; 4.DNA Chip Research Inc, Tokyo, Japan

Background: Pulmonary arterial hypertension (PAH) is prominent as a vascular involvement in systemic sclerosis (SSc), which remains a leading cause of death in spite of current best treatments. Although recent studies focused on early diagnosis of established PAH, it is known that more than a half of the pulmonary circulation is impaired before early PAH is detected. However, there is little study about the changes of vascular functions or the underlying gene expressions during its subclinical stage.

Objectives: I. To detect the pathological changes in pulmonary arterial pressure (PAP) and vascular functions before PAH is manifested. II. To explore the changes in its underlying gene expressions of peripheral blood.

Methods: Total of 103 cases without PAH symptoms (NYHA I) with either Raynaud phenomenon (RP: n=87), skin sclerosis (n=65) or SS-related autoantibody (n=68) were enrolled. To detect the pathological change of PAP, exercise Doppler echocardiography was carried out, and exercise induced pulmonary hypertension (exPH) group was segregated from normal response group (exN) with using the definition described in R. Naeije et al.1 Vascular function was evaluated with thermography after 0°C-stress and determination of ankle-brachial index (ABI) and cardio-ankle vascular index (CAVI), Furthermore, reactive hyperemic index (RHI), augmentation index (AI) and second derivative of photoplethysmogram ageing index (SDPTGAI) were assessed using EndoPAT. Micro-vascular changes were also recorded with nailfold videocapillaroscopy. Meanwhile, genome-wide gene expression analysis was performed with using whole peripheral blood. The genes correlated with each vascular function tests were analysed by weighted gene co-expression network analysis (WGCNA) and pathway enrichment analysis (PathVisio).

Results: There were significant differences between exPH and exN group in the result of thermography after 0°C-stress test, CAVI and AI normalised to heart rate of 75bpm (AI@75bpm). As the CAVI and AI are known to correlate positively with age, careful interpretation was necessary because the mean age of exPH group was higher as compare with exN group (69.05±11.04 vs. 60.23±14.73). However, the fact that recovery of blood-flow from RP was significantly delayed in exPH group suggested the additional pathological changes of vascular and endothelial functions. Gene expression analysis revealed that several mutual pathways such as type2 interferon signalling, oxidative damage and fatty acid omega oxidation seemed to underlie some vascular changes.

Conclusions: The results of vascular function tests including thermography after 0°C-stress, CAVI and AI@75bpm were significantly different between exPH and exN group. On the other hand, gene expression analysis showed that many factors such as ageing, arteriosclerotic and immunological mechanisms were involved in the changes of these vascular functions. Although further prospective study is required to select appropriate set of the tests, it is possible that evaluation of these vascular functions may be useful as a non-invasive test to assess the pulmonary vascular disease before PAH is manifested.

REFERENCE:

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AB0822 A MEASUREMENT OF ANTI-ARS ANTIBODIES, ANTI-MI-2 ANTIBODY, ANTI-TIF1 GAMMA ANTIBODY AND ANTI-MDA5 ANTIBODY BY ENZYME-LINKED IMMUNOSORBENT ASSAY AS A DIAGNOSTIC TOOL OF IDIOPATHIC INFLAMMATORY MYOPATHY RHEUMATOLOGY DAILY PRACTICE

Y. Miyoshi1,2, T. Kise1, N. Yokogawa1, K. Shimada1, S. Sugii1. 1.Department of rheumatic diseases, Metropolitan Tama Medical Center, Fuchu, Tokyo; 2.Department of rheumatic diseases, Tama-Hokubu medical center, Higashimurayama, Tokyo, Japan

Background: Enzyme-linked immunosorbent assay (ELISA) tests of anti-ARS antibodies, anti-Mi-2 antibody, anti-TIF1γ antibody and anti-MDA5 antibody might be a potential strategy for DM.

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