[AB0647] DISEASE SUBSETS, AUTOANTIBODIES AND CLINICAL FEATURES IN 176 ADULT ITALIAN PATIENTS WITH SYSTEMIC SCLEROSIS: A CROSS SECTIONAL STUDY

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Background: Systemic sclerosis (SSc) is a chronic autoimmune disease characterized by vascular changes and progressive fibrosis of skin and various internal organs. A variety of circulating autoantibodies have been detected in SSc. Some of them are well-established tools strongly associated with SSc and useful for diagnosis and management of the disease (e.g. ANA, anti-centromere, anti-topoisomerase I, anti-RNA polymerase III – RNAP III). Other autoantibodies are less frequent and non-specific for SSc even if potentially useful to better assess clinical subsets and prognosis (e.g. anti-Th/To, anti-Ku, anti-PM/Scl).
Objectives: To investigate the prevalence of anti-Th/To, anti-Ku, anti-RNAP III and anti-PM/Scl antibodies as well as to study the associations between these antibodies and clinical features in Italian SSc patients.
Methods: Sera from 176 consecutive patients with SSc collected at two Italian sites were tested for anti-Th/To (anti-Rpp25 and anti-Rpp38), anti-Ku, anti-RNAP III, anti-PM/Scl, and anti-centromere antibodies (anti-CENP-A and B) using QUANTA Flash chemiluminescence immunoassays (CIAs) on the BIO-FLASH instrument (Inova Diagnostics, San Diego, USA). Additionally, ANA screening was performed using QUANTA Flash CTD Screen Plus which includes the most relevant nuclear/cytoplasmic antigens. The Italian cohort included: women 162 (92%), limited cutaneous (lc-SSc) subtype 137 (78%), patients with present or past history of digital ulcers 56 (32%), calcinosis 25 (14%), lung fibrosis 69 (39%), heart involvement 22 (12.5%), pulmonary arterial hypertension 11 (6%) and esophageal involvement 84 (48%).
Results: 152 (86.4%) patients were CTD Screen Plus positive, 8 (4.5%) anti-Th/To positive, 8 (4.5%) anti-Ku positive, 20 (11.4%) anti-RNAP III positive, 0 (0%) anti-PM/Scl positive, 70 (39.8%) anti-CENP-A positive, and 87 (49.3%) anti-CENP-B positive. Anti-CENP-A, anti-CENP-B and anti-Ku antibodies were significantly associated with the lc-SSc subtype ($p<0.0001$, $p<0.0001$ and $p=0.0469$, respectively). None of patients positive for anti-Ku antibodies displayed digital ulcers ($p=0.0075$). Anti-Th/To (Rpp25) antibodies were associated with the presence of calcinosis ($p=0.0399$) while anti-RNAP III antibodies with its absence ($p=0.0353$). All patients positive for anti-Th/To were positive for the Rpp25 component while only one of them was also positive for the Rpp38 component.
Conclusions: The present study points out the low prevalence of anti-RNAP III and highlights a lower prevalence of anti-Th/To in Italian SSc patients. Several known associations between autoantibodies as SSc subsets have been confirmed while others could not be verified in the study. This supports the potential utiliy of autoantibodies to stratify SSc into clinical subsets. Large international multi-center studies are required to define and validate rare autoantibodies such as anti-Th/To, anti-Ku and anti-PM/Scl antibodies.
Disclosure of Interest: None declared
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