AB0667

SPECIFICITY OF ANTI-TRNA SYNTHETASE AUTOANTIBODIES CORRELATED WITH CLINICAL COURSE AND PROGNOSIS OF MYOSITIS-ASSOCIATED INTERSTITIAL LUNG DISEASE

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Background: Interstitial lung disease (ILD) is associated with mortality among patients with idiopathic inflammatory myopathy (IIM). Anti-aminoacyl-tRNA synthetase (anti-ARS) autoantibodies have been identified highly specific for IIM and coincidence of ILD. It has been identified that non-Jo1 positive anti-ARS patients may have decreased survival rate. However, differences of characteristics and prognosis in myositis-associated ILD with an individual anti-ARS autoantibody have not been clarified.

Objectives: To compare the characteristics and cumulative survival of myositis-associated ILD with different positivity of anti-ARS antibodies.

Methods: We investigated retrospectively all consecutive patients with myositis-associated ILD evaluated from 1999-2017. All autoantibodies were screened by using RNA and protein immunoprecipitation assays. Demographic data, laboratory findings and chest computed tomography (CT) images were obtained.

Results: Among 105 patients with myositis-associated ILD, enrolled, 47 had anti-Jo1 antibody and 58 had non-Jo1 antibody: anti-EJ (n=20), anti-PL12 (n=12), anti-PL7 (n=11), anti-KS (n=8) and anti-OJ (n=7). The diagnosis at first visit were polymyositis (PM; n=28), dermatomyositis (DM; n=42), clinically amyopathic dermatomyositis (CADM; n=3) and antisynthetase syndrome (ASS; n=31). Most common causes of death in this study were exacerbation of ILD (n=10, 71.4%). Anti-Jo1 positive patients had better ILD prognosis than non-Jo1 patients (log rank test, p = 0.03). The prevalence of rapidly progressive ILD (RP-ILD) and upper lung field lesions in CT scans were significantly higher in anti-EJ positive patients (p=0.02 and p=0.05, respectively). Although the prognosis of ILD was not significantly different among 6 anti-ARS antibodies, anti-EJ positive patients had poor ILD prognosis than anti-Jo1 patients with the 5-year unadjusted cumulative survival was 73% and 95%, respectively.

Conclusion: Our data confirm that the characteristics and outcomes of myositis-associated ILD are distinct by anti-ARS antibodies and highlighted that anti-EJ antibody positivity associated with relatively poor prognosis and RP-ILD.

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AB0668

ASSOCIATION OF ANTI-RNA POLYMERASE III ANTIBODY AND BREAST IMPLANTS RUPTURE IN ITALIAN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Several epidemiological studies have investigated the link between silicone breast implants (SBI) and Systemic Sclerosis (SSc). Although discordant data were reported, a recent analysis of SBI followed by United States Food and Drug Administration post approval studies, including nearly 100,000 individuals, described an association of SBI with a higher rate of SSc (Standardized incidence ratio 7.00), compared with normative data¹. The analysis of clinical associations in patients with SSc is complicated by the heterogeneity of the disease, both on immunological and clinical terms. Interestingly, a specific association of anti-RNA polymerase III antibody (anti-RNAP3) and SBI in Japanese patients with

SSc was described in a single-center cohort². Moreover, an association of anti-RNAP3 with breast cancer, particularly when synchronous with SSc onset, was also demonstrated^{3,4}.

Objectives: To evaluate the association of SBI with SSc in Italian patients classified according to their SSc-related autoantibodies.

Methods: 562 consecutive women with SSc, classified according the 2013 ACR/EULAR criteria, were evaluated for the presence of SSc-specific autoantibodies (anti-Topoisomerase-I (anti-Topo-I), anticentromere (ACA), and anti-RNAP3) in two Italian University centres. For each patient, history of breast cancer and SBI were recorded.

Results: Anti-RNAP3 antibodies were found in 4.6%, anti-Topo-I in 22.2% and ACA in 57.3% of total SSc women (Table 1). Comparing anti-RNAP3+ vs anti-RNAP3- patients, a significantly higher frequency of breast cancer (15.3% vs 4.1%; p=0.026), SBI (11.5% vs 1.1%; p=0.006) and SBI rupture (11.5% vs 0.2%; p=0.0003) was found. Notably a higher rate of SBI rupture without breast cancer in anti-RNAP3+ vs anti-RNAP3- was also noted (2/22 (9.1%) vs 0/514 (0%); p=0.0016).

Conclusion: In this large Italian cohort, a higher prevalence of SBI and SBI rupture in anti-RNAP3+ compared to anti-RNAP3- SSc patients was confirmed, regardless of the history of breast cancer. This preliminary observation should be confirmed in multicentre cohorts, particularly regarding the connection between SSc and SBI rupture.

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Table 1. Frequency of SBI, SBI rupture and breast cancer in 562 SSc patients, divided according to the autoantibody status.

	Patients	SBI	SBI rupture	Breast cancer
ACA	322	4	1	16
Anti-TopoI	125	0	0	2
Anti-RNAP3	26	3	3	4
Others	89	2	0	4
Total	562	9	4	26

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AB0669

THE EFFECTS OF BOSENTAN FOR TREATMENT OF DIGITAL ULCER IN KOREAN PATIENTS WITH SYSTEMIC SCLEROSIS: PROSPECTIVE, MULTICENTER, OPEN-LABEL TRIAL

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Background: Bosentan is an orally administered dual endothelin-1 (ET-1) receptor antagonist approved in the EU for reducing the number of new digital ulcers in patients with systemic sclerosis (SSc) and ongoing digital ulcer (DU). Oral bosentan therapy was beneficial and generally well tolerated in patients with DU associated with SSc. A total of 3 RCTs were identified. The first two were high quality (RAPIDS-1 and RAPIDS-2) with 122 and 188 patients and treatment durations of 16 and 24 weeks, respectively. Both studies showed that, compared with placebo, bosentan significantly reduced the number of new ulcers; however, it did not have an effect on the healing of preexisting ulcers and pain.

Objectives: This study evaluated the efficacy and safety of bosentan in DU secondary to SSc in Korean patients.

Methods: From December 2013 to December 2017, a prospective, multicenter, open-label trial was performed to investigate the complete healing of DU and tolerability of bosentan in Korean patients with SSc. The SSc