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 (IMI). Anti-aminocarboxyl-tRNA synthetase (anti-ARS) autoantibodies have been identified highly specific for IMI 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 rapid progression ILD (RP-ILD) and upper lung field lesions in CT scans were significantly higher in anti-Jo1 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-Jo1 positive patients had poor ILD prognosis than non-Jo1 patients with the 5-year 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-Jo-1 antibody positivity associated with relatively poor prognosis and RP-ILD.

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ACKNOWLEDGMENT
The authors would particularly like to thank Etsuko Iwata and Masui for their assistance.

Disclosure of Interests: None declared.


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 was described in a single-center cohort.

Disclosure of Interests: Maria Grazia Lazzaroni: None declared, Cristian Cammi: None declared, Eunegia Bertolino: None declared, Franco Franceschini: None declared, Angela Tincani Consultant for: UCB, Pfizer, Abbvie, BMS, Sanofi, Roche, GSK, Alph Sigma, Lilly, Jannsen, Cellgene, Novartis, Paolo Airo: None declared.

patients were included as having active DU or pulmonary arterial hypertension who had active DU in the last 3 months. A DU was defined as an area with visually discernable depth and a loss of continuity of epithelial coverage, which could be denuded or covered by a scab or necrotic tissue. If the area was denuded, the ulcer was designated active. If denudation could not be judged because of the presence of overlying scab or necrotic tissue, ulcers presenting with features, including underlying pain, based on investigator clinical judgment to be consistent with loss of epithelialization, epidermis, or dermis, and requiring treatment, were designated as active. The effectiveness and tolerability of bosentan was monitored within 24 weeks of study treatment, while healing of DU, largest diameter and the new incidence were also assessed up to week 24.

Results: The primary objective of this study was to assess the efficacy of bosentan in reducing DU including the complete healing rate. Secondary endpoints included development of new DU, assessment of discontinuation, measures of nailfold capillaroscopic (NFC) change. Thirty-five SSc patients were enrolled and seventeen patients were included in the NFC analysis. Mean age was 50.5 year-old, 68.6% was woman, 68.6% were diffuse type of SSc; mean mRSS was 15.4 and mean disease duration was 77.9 months. Among 35 patients, 20 patients was finished 24 weeks treatment duration. The main reason for discontinuation is when the patient wants to stop because of the full effect of the drug (n=12, 80%). The side effects of the drug occurred in 4 cases during the treatment period. The mean number of DU's before treatment was 2.4 and was statistically decreased to 1.2 (p<0.001). From the 4th week of bosentan treatment, improvement of DU number was observed in 65.7% and complete healing was observed in 55% at 24 weeks. However, the mean size of largest DU diameter, remained at the end of treatment, was slightly increased from 4.1 to 5.2 mm, with no statistical significance. There was no statistically significant difference in the semi-quantitative changes of NFC before and after bosentan treatment.

Conclusion: Administration of bosentan over 24 weeks was statistically associated with complete healing and significant reduction in net number of DU in Korean patients with SSc. However, the size of the remaining DU that did not respond to bosentan, has rather increased.

Disclosure of Interests: None declared


AB0670

INTESTINAL ANGIODISPLASIAS IN THE PATIENT WITH SYSTEMIC SCLEROSIS

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Background: Gastric angiodysplasia and the typical "watermelon stomach" aspect is one of the most characteristic vascular manifestations in systemic sclerosis (SS). These angiodysplasias can affect the intestine occasionally as well as the stomach.

Objectives: Description of a series of 5 patients with systemic sclerosis and presence of intestinal angiodysplasia (IA).

Methods: Retrospective unicentric observational study in a tertiary hospital. Demographic data, comorbidities, manifestations of the disease, serological profile, concomitant treatments and bleeding manifestations, as well as endoscopic findings and received treatment were collected. Descriptive statistics was used to present the data.

Results: We reviewed the medical records of the 88 patients with a diagnosis of SS followed up in the Rheumatology Service. Five cases (5.6%) of IA were identified (4 women and one male), with a mean age of 73 years (SD 13.71). In one case, the diagnosis of SS and IA was simultaneous. In the remaining 4 cases, the mean time from the diagnosis of ES to the diagnosis of the IA was 7.75 years (SD 2.6). Regarding the clinical presentation, 4 of the cases presented as anemia with iron deficiency. In these cases IA were found in the endoscopic studies. In one case, IA were a finding in the context of the study of diarrhea. In 4 of the cases, the IA were in the small intestine, so the diagnosis was made using an endoscopic capsule. In one case they were found in the colon, so the diagnosis was by colonoscopy. Only one of the cases of angiodysplasias in the small intestine required specific treatment with argon laser by enteroscopy. One case required a transfusion of packed red blood cells. In the remaining cases, treatment with oral or intravenous iron therapy was enough. In all 5 cases, it was a limited form of the SS with the presence of antitrombore antibodies. All cases presented Raynaud’s phenomenon, 3 of them with digital ulcers. Three of the cases associated primary biliary cholangitis, 3 cases pulmonary hypertension, 3 cases peristaltic involvement, 1 case interstitial lung disease and 1 case restrictive cardiomyopathy. As comorbidities, we identified 1 case of essential mixed cryoglobulinemia associated with hepatitis C virus, 2 cases of valvular heart disease, 2 cases of atrial fibrillation.

Conclusion: The extragastric localization of the angiodysplasias should be taken into account in the patient with SS and iron deficiency, especially in those with a limited form of the illness and positive antitrombore antibodies.

Disclosure of Interests: Carolina Merino Argumánez: None declared, Olga Rusinovich: None declared, María Espinosa: None declared, Consuelo Ramos Giráldez Speakers bureau: Sanofi, Carmen Barbadillo Mateos: None declared, José Luis Andrés Sanchez: None declared, Hilda Godoy: None declared


AB0671

DISCORDANCE IN ANTI-TIF1-γ ANTIBODIES DETECTION THROUGH COMMERCIAL KIT IN COMPARISON WITH HOMEMADE IMMUNOBLOT: CLINICAL EVALUATION

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Background: The antibody against the transcriptional intermediary factor 1γ (Anti-TIF1-γ or Anti-p155), is considered specific for dermatomyositis (DM) and its a marker of cancer risk. Our center carries out the determination of Anti-TIF1-γ by a homemade immunoblot (IB) technique with a 155 kDa TIF1γ human recombinant protein (OriGene). Currently there is