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AB0596 INTERFERON- γ RELEASE ASSAY (T-SPOT.TB) IN THE DIAGNOSIS OF TUBERCULOSIS INFECTION IN PATIENTS WITH BEHÇET'S DISEASE: A SINGLE CENTER EXPERIENCE IN CHINA

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Background: In many aspects Behcet's disease (BD) is correlated with tuberculosis (TB). They may mimic each other on clinical manifestations. For example erythema nodosum, arthritis, gut lesions could be occurred in both disease and it is difficult for differential diagnosis. Some patients with newly-diagnosed BD were found to have active or latent or old TB and anti-TB therapy could relieve some of the symptoms. In etiology, it has been postulated that tubercle bacilli may act as a trigger of BD through the mechanism of molecular mimicking. Vice versa, defective cell-mediated immunity in BD patients may increase individual susceptibility of TB. It is extremely important to clarify TB existence or not in BD patients. Since BD patients commonly need glucocorticoid and immunosuppressant or even biologic agent such as anti-TNF α . And these medications may increase the risk of occurrence or flare of TB. But positive result of PPD shows little value in this special population due to acupuncture reaction in BD patients. In this pilot study, the values of interferon- γ release assay (T-SPOT.TB) in diagnosing active TB in BD patients were explored.

Objectives: To investigate the diagnostic value of the T-SPOT.TB in BD patients complicated with tuberculosis infection.

Methods: The clinical, radiology and laboratory data were collected and analyzed in 175 hospitalized BD patients from the Peking Union Medical College Hospital between January 2010 and March 2015. Statistical analysis was carried out using IBM SPSS version 20.

Results: Of the 175 BD patients, the positive rate of tuberculosis infection in BD patients was 34.3%. The sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) of the T-SPOT.TB test for the diagnosis of ATB were 87.5%, 73%, 36.8%, 98.3%, respectively. Positive likelihood ratio (PLR) and negative likelihood ratio (NLR) were 3.24 and 0.17. The median number of SFCs in the BD-ATB group was higher than that in the BD-LTB group and BD-OTB ($p < 0.001$ and $p = 0.012$). By ROC method, it was suggested that 70 SFCs act as a cutoff for diagnosing BD-ATB with the sensitivity, specificity, PPV, NPV, PLR and NLR were 87.5%, 85%, 24.6%, 98.5%, 5.79 and 0.15, respectively. The rates of LTb infection had no significant difference between BD patients and healthy controls. Agreement between T-SPOT.TB and TST in BD patients measured by the kappa coefficient was poor ($\kappa = 0.37$). Multiple logistic regression analysis revealed that BD patients with positive T-SPOT.TB had the highest likelihood of ATB (OR 11.93, 95% CI 2.108–67.508, $p = 0.005$).

Conclusions: T-SPOT.TB is a promising test in the diagnosis of BD complicated with tuberculosis infection, and higher number of SFCs may have a higher risk of ATB.

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AB0597 LOW-DOSE RITUXIMAB AS INDUCTION THERAPY FOR JAPANESE PATIENTS WITH ANCA-ASSOCIATED VASCULITIS

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Background: Four times of once-weekly doses of 375 mg/m² rituximab (RTX) are frequently used as remission induction therapy for ANCA-associated vasculitis (AAV). Since this regimen has been basically generated from experience of B-cell non-Hodgkins' lymphoma in the Europe and North America, appropriate dose and interval for patients with AAV in other population have been poorly investigated. Here we comprehensively analyzed the efficacy and tolerability of high or low dose regimen of RTX in Japanese patients with AAV.

Objectives: We investigated the efficacy and safety issue of low dose RTX for Japanese patients with AAV.

Methods: We retrospectively examined AAV patients who met the 2012 Chapel Hill classification from 2006 to 2016. We divided them into 2 groups, those treated with high-dose (HD) and low-dose (LD) RTX. HD RTX was the original regimen and LD RTX consisted of twice of one-weekly dose of 375 mg/m². We evaluated cumulative complete remission (CR) rate and relapse-free rate for 1.5 years. CR was defined as BVAS=0 and relapse was defined as BVAS $\geq$ 1.

Results: We evaluated 17 patients with HD and 11 patients with LD RTX. Higher percentage of elderly patients was observed in LD group ($p < 0.01$). No significant difference was found in BVAS ($p = 0.49$) and VDI ($p = 0.15$) before treatment. No significant difference was found in cumulative CR rate ($p = 0.90$) (Fig. 1A), relapse-free

rate ($p = 0.48$) (Fig. 1B), B cells counts and serious adverse events. We found patients with nasal involvement and pulmonary nodule/cavity formation had higher relapse rate in LD group than those with HD group ($p = 0.05$, and $p = 0.09$ respectively).

Figure 1

Conclusions: LD regimen of RTX, especially in elderly patients, might be effective in remission induction therapy in AAV.

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Scleroderma, myositis and related syndromes

AB0598 SERUM DEFENSIN LEVEL IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Scleroderma is an autoimmune disease characterized by fibrosis of skin and lung as well as involvement of kidney, gastrointestinal system and heart (1). Etiology and exact mechanism of disease is poorly understood. A small number of studies have examined the role of AMPs on autoimmune diseases. It has been demonstrated that the amount of alpha- and beta-2 defensin serum levels are increased in systemic lupus erythematosus patients (2,3). Likewise, the association between AMPs and other diseases such as idiopathic pulmonary fibrosis, diffuse panbronchiolitis, pulmonary alveolar proteinosis and psoriasis has been reported (4).

Objectives: No study investigated the role of AMPs on scleroderma patients. Hence, we aimed to investigate AMP serum levels and their possible association in these patients.

Methods: There were 42 patients (40 female, mean age 42 years) and 38 healthy subjects (32 female, mean age 38 years) in the study. For SSc patients, the following data were recorded at enrollment: disease subset (limited/diffuse), autoantibodies (antinuclear, anti-centromere (ACA), and anti-scl), blood tests, erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP), modified Rodnan skin score, presence and history of digital ulcers, presence and history of involvement kidney and gastrointestinal system, interstitial lung disease detected by chest HRCT and pulmonary function tests, estimated pulmonary arterial systolic pressure at echocardiography.

Results: There were 42 patients (40 female, mean age 42 years) and 38 healthy subjects (32 female, mean age 38 years) in the study. Twenty-nine of the patients had diffuse systemic sclerosis and thirteen of the patients had limited systemic sclerosis. Average disease duration is 5.5 years. Pulmonary involvement was detected in twenty patients. The levels of beta 1 and beta 2 defensin that are epithelial defensins were higher than control group but it has not reached statistical significance. (beta-1 defensin 235 $\pm$ 178 vs 185 $\pm$ 24 pg/ml, $p = 0.08$ and beta2 defensin 253 $\pm$ 453 vs 152 $\pm$ 101 pg/ml, $p = 0.1$). Alpha defensin levels in scleroderma patients were significantly higher than control group (563 $\pm$ 415 vs 377 $\pm$ 269 ng/mL, $p = 0.02$). In sub-group analysis patients with interstitial lung disease had a higher level of alpha defensin than those without involvement (684 $\pm$ 473 vs 430 $\pm$ 299 ng/ml, $p = 0.04$). There was a negative correlation between alpha defensin and Rodnan score ($r = 0.30$) and CRP ($r = 0.34$).

Conclusions: Alpha defensin levels in scleroderma patients were significantly higher than control group. There may be an increase in the level of alpha defensin in a cause of vasculopathy in scleroderma patients

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AB0599 COMBINATION OF CAPILLAROSCOPIC AND ULTRASONOGRAPHIC EVALUATIONS OF THE HAND TO DETECT SEVERE VASCULOPATHY IN SYSTEMIC SCLEROSIS: RESULTS OF A CROSS-SECTIONAL STUDY

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Background: Although micro-vessel alterations have been largely described, macrovascular involvement is also frequent in Systemic Sclerosis (SSc). Macrovascular damages specifically involve narrowing or occlusions of proper palmar digital arteries and ulnar artery. On the contrary, radial artery is rarely concerned. Ulnar artery occlusion (UAO) assessed by power doppler ultrasonography (PDUS) has proven to be predictive of the onset of new ischaemic DUs in longitudinal studies (1). PDUS could also be a reliable tool to evaluate finger pulp blood flow (FPBF). Only few studies have explored the association between macrovascular damages evaluated by PDUS and microvascular involvement assessed by nailfold capillaroscopy (NC) (2). The association between macrovascular disease and calcinosis or Acro-osteolysis is still to be determined in SSc.

Objectives: to confront microvascular damages on NC with macrovascular manifestations evaluated by PDUS in SSc patients. Micro and macro-vascular damages were confronted with the main digital manifestations of the disease: digital ulcers (DU), Acro-osteolysis and Calcinosis.

Methods: NC, hand X-Rays and PDUS were systematically performed in 64 unselected SSc patients. PDUS evaluation with assessment of Ulnar Artery Occlusion (UAO) and finger pulp blood flow (FPBF) was performed blinded for the results of X-Rays and NC.

Results: UAO and pathologic FPBF were associated with severe capillary loss (<4 capillaries/mm) on NC (respectively OR=4.04 (1.23–13.29); p<0.05 and OR=3.38 (1.03–11.05); p<0.05). UAO was significantly associated with Cutolo's late NC pattern (OR=3.80 (1.31–11.01); p<0.05). A DU history was associated with UAO (OR=10.71 (3.36–34.13); p<0.0001), pathologic FPBF (OR=7.67 (2.52–23.28); p<0.0001), late pattern (OR=6.33 (2.03–19.68) and severe capillary loss (OR=8.52 (2.15–33.78); p=0.001). Acro-osteolysis was also associated with UAO (OR=15.83 (3.95–63.54); p<0.001), pathologic FPBF (5.52 (1.71–17.90) p=0.003), late NC pattern (OR=6.86 (2.18–21.53); p=0.001) and severe capillary loss (OR=7.20 (2.16–24.02); p=0.001). Calcinosis on X-rays were associated with late NC pattern (5.41 (1.82–16.12); p=0.002), severe capillary loss (OR=12.69 (3.14–51.26); p<0.0001) and UAO (3.19 (1.14–8.92); 0.025) but not with pathologic FPBF. Combination of UAO and severe capillary loss in a same patient was especially associated with DU history (OR=18.60 (2.24–154.34); p<0.001) and Acro-osteolysis (OR=10.83 (2.56–45.88); p=0.001).

Conclusions: The combination of macro and microvascular evaluations by PDUS and NC may help to detect patients with a more severe vasculopathy.

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AB0600 CLINICAL RELEVANCE OF AUTOANTIBODY PROFILES IN SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a connective tissue disease accompanied by immune abnormalities. A number of autoantibodies such as anti-centromere, anti-topoisomerase I, anti-RNA polymerase III and anti-U3 (fibrillar) antibodies were proven to be of great diagnostic and prognostic factors in patients with scleroderma. Certain studies have reported the presence of

antinuclear, anti-SSA (Ro) and/or anti-SSB (La), anti-Pm/Scl, anti-endothelial cell and anti-nucleosome antibodies in patients with systemic sclerosis. However, the clinical relevance of these autoantibodies is yet to be fully elucidated.

Objectives: Our aim was to assess clinical features, capillaroscopic abnormalities and autoantibody titers in patients with systemic sclerosis as well as analyze relationships between these parameters.

Methods: We conducted a prospective observational study on 36 adult patients who satisfied ACR/EULAR 2013 criteria for systemic sclerosis. We recorded disease duration, current symptoms and classified patients as limited cutaneous (lc) SSc and diffuse cutaneous (dc) SSc. Skin involvement was assessed using the modified Rodnan skin score (mRSS). We performed nailfold videocapillaroscopy using a FEDMED Digitale 100N at a magnification of 200X. Thoracic X-rays were done to establish the presence of pulmonary fibrosis. Ultrasounds were performed by a single examiner to evaluate pulmonary artery pressure. Blood samples were drawn to measure anti-topoisomerase I, anti-centromere, anti-SSA (Ro), anti-SSB (La), anti-U1RNP and anti-nucleosome antibody titers (ELISA). Patient characteristics were included in a database and analyzed using IBM SPSS Statistics v20.

Results: Our study group was composed of 20 dc SSc (55.6%) and 16 lc SSc (44.4%) patients. Severity of capillary changes correlated with mRSS values (p<0.01), anti-topoisomerase I (p=0.006), anti-SSA (p=0.01) and anti-nucleosome antibodies (p=0.02). We found positive associations between the presence of dysphagia, anti-centromere (p=0.009) and anti-SSA (p<0.01) titers in patients with lc SSc. Anti-centromere antibody positivity also correlated with pulmonary hypertension (p=0.011) and pulmonary fibrosis (p=0.04) in these patients. Anti-SSA antibodies correlated with pulmonary hypertension (p<0.01) and capillaroscopic changes (p=0.024) in dc SSc.

Conclusions: Our findings support the relationship between autoantibody titers, systemic involvement and microvascular changes in scleroderma patients. Non-specific autoantibodies such as anti-nucleosome and anti-SSA antibodies were associated with microvascular changes in our study group. Further studies in this field may provide new information on SSc pathogenesis and possibly novel targets for treatment in scleroderma patients.

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AB0601 ANATOMIC AND FUNCTIONAL ASSESSMENT OF PERIPHERAL PERFUSION IN PATIENTS WITH SYSTEMIC SCLEROSIS. IS THERE ANY CORRELATION BETWEEN CAPILLAROSCOPIC FINDINGS AND ERGOSPIROMETRY?

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Background: Microvasculopathy of systemic circulation in patients with Systemic Sclerosis (SS) is widely assessed by digital capillaroscopy (DC), a method that evaluates the architecture of capillary network and reveals changes of vascular anatomy. On the other hand, ergospirometry (ERG) reveals functional impairment of microcirculation by assessing indirect measures of peripheral tissue ischemia (most suitable VE/VCO₂). Since today, there have been studies correlating DC findings with spirometric parameters (respiratory volumes)^{1,2}; nevertheless, no reports have been published correlating the same DC findings with ERG parameters (functional microvascular perfusion).

Objectives: To propose the use of DC as a screening tool for impaired functional microvasculopathy by investigating correlations between patterns of capillaroscopic findings with ergospirometric values of peripheral tissue blood perfusion.

Methods: 11 patients (11 women mean age 43±12 ys) with SS were evaluated contemporarily with High Resolution Computed Tomography of the chest, ERG, and DC. Parameters were correlated with multiple regression analysis. Statistic significance was considered p<0.05.

Results: Patient data are shown in the table.

Patients with late pattern in capillaroscopy had less endurance in exercise test (P<0.05) but no correlation was found between capillaroscopic pattern and VE/VCO₂ (P>0.05). Age was a crucial confounding factor.

Conclusions: The correlation of late pattern capillaroscopy findings with reduced ergospirometric endurance in 11 patients indicates that in a larger cohort, specific parameter associations between DC and ERG are probable to emerge.