AB0734 SERUM IL-35 LEVELS IN SYSTEMIC SCLEROSIS AND RELATIONSHIP WITH CLINICAL FEATURES

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Background: Interleukin (IL)–35 is a member of the IL-12 family. Studies show that IL-35 is an important anti-inflammatory cytokine and suppresses effector T cell activity. Systemic sclerosis (SSc) is a chronic disease characterised by vascular damage, autoimmunity and fibrosis.

Objectives: In this study, we aimed to evaluate serum IL-35 level in SSc patients and its potential relation with disease findings.

Methods: Fifty-five SSc patients and 25 healthy controls were included in the study. All patients were diagnosed according to 2013 ACR/EULAR systemic sclerosis classification criteria. Serum IL-35 was measured using a commercial ELISA kit (Cloud-Clone Corp, Wuhan, China).

Results: SSc patients and healthy controls had similar ages and genders. The mean of serum IL-35 level was significantly higher in SSc patients (8.2±1.3 pg/ml) than in healthy controls (6.9±0.7 pg/ml) ($p < 0.001$). Serum IL-35 levels were similar in lcSSc (n=44) and dcSSc (n=11) groups (8.3±1.2 and 8±0.1, 1.6 pg/ml, respectively). There were no significant relationships between serum IL-35 level and digital ulcer, interstitial lung disease, pulmonary hypertension, tendon friction rub, gastrointestinal motility disorder, cardiac involvement and Anti-SCL70, anti-CENP-B positivities (table 1). There was not any correlation between IL-35 level and disease duration, modified Rodnan skin score, Valentini disease activity score, Medsger disease severity score, erythrocyte sedimentation rate and C-reactive protein (CRP) levels. Serum IL-35 levels between patients treated with or not vasodilating agents such as calcium channel blockers, endothelin receptor antagonists and prostaglandin analogues, and also immunosuppressive drugs did not differ significantly.

Abstract AB0734 – Table 1. Relationship between clinical and laboratory features and IL-35

		IL-35 (pg/ml)	P
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Digital ulcer	- (n=32) + (n=23)	8.3±1.4 8.1±1	0.5
Gastrointestinal involvement	- (n=43) + (n=12)	8.3±1.2 8±1.5	0.512
Cardiac involvement	- (n=52) + (n=3)	8.1±1.3 9.5±0.8	0.073
Interstitial pulmonary disease	- (n=34) + (n=21)	8.3±1.3 8.1±1.3	0.593
Pulmonary arterial hypertension	- (n=51) + (n=4)	8.2±1.3 7.9±1.2	0.617
Arthritis	- (n=36) + (n=19)	8.4±1.3 7.9±1.2	0.160
Tendon friction rub	- (n=47) + (n=8)	8.2±1.3 8.3±1	0.798
Anti-Scl 70	- (n=34) + (n=20)	8.3±1.3 8.2±1.2	0.793
Anti-CENP B	- (n=33) + (n=21)	8.1±1.3 8.4±1.3	0.442

*mean ±std, + and – denote presence and absence, respectively.

Conclusions: It has been reported that serum IL-35 level is higher SSc patients than healthy volunteers and is associated with interstitial lung disease. In our study, the level of IL-35 was higher SSc patients than healthy controls. There is no relationship between clinical parameters and IL-35 levels, this may be due to small sample size. Our results suggest that IL-35 may have a role on the pathogenesis of SSc. Further investigations are needed in large sample size of SSc patients.

Disclosure of Interest: None declared

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AB0735 NAILFOLD CAPILLARY COUNTS CORRELATE MOST WITH COMPLICATIONS OF SYSTEMIC SCLERODEMA IN JAPANESE PATIENTS

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Background: Connective tissue diseases (CTD) patients have various clinical manifestation including pulmonary hypertension (PH) and digital ulcer (DU). Especially, Raynaud's phenomenon (RP) is one of the common symptoms. On the other hand, "abnormal nailfold capillaries" are important as the findings that suggest RP associated with CTD. They are included in one of the items of classification criteria for Scleroderma (SSc) in 2013 Classification Criteria for Systemic Sclerosis (ACR/EULAR) ¹. Recently, some reports presented the usefulness of nailfold capillaroscopy (NFC) in SSc ². NFC findings showed enlarged/giant capillaries, fresh or old haemorrhages, avascular areas, ramified/bushy capillaries. These findings are known as "scleroderma pattern" features. Additionally, there are some reports that nailfold capillary density in SSc related with DU, PH and interstitial pneumonia (IP).

In this study, we clarify the association between nailfold capillary density and clinical manifestation by analysis of the case in our institution.

Objectives: To clarify the association between nailfold capillary density and clinical manifestation.

Methods: We enrolled Japanese SSc patients from May 2016 to May 2017 in our institution.

We measured total nailfold capillary count per 1 μm from second to fifth finger by NFC "OptiPIX Capillaroscopy Clinic 1.7.x". In addition, we investigated relationship with NFC findings and clinical manifestation such as current and previous DU, PH and IP.

Results: We enrolled 42 SSc patients. Total nailfold capillary counts significantly decreased, especially in SSc patients with, previous DU ($p = -0.4122$, $p = 0.0054$), and PH ($p = -0.3226$, $p = 0.0023$). In addition, there is significant difference between DU and existence of giant capillaries ($p = 0.3019$, $p = 0.0464$), existence of avascular areas ($p = 0.4057$, $p = 0.0063$). On the other hand, there is no significant difference between IP, PH and other "scleroderma pattern" features.

Conclusions: Our study revealed that total nailfold capillary counts were associated with PH and DU in SSc patients. We suggest that nailfold capillary density may most predict complications with SSc patients.

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AB0736 SYSTEMIC SCLEROSIS SINE SCLERODERMA: CHARACTERISTICS OF A SOUTH INDIAN COHORT FROM A TERTIARY CARE CENTRE

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Background: Systemic Sclerosis is a complex disorder characterised by autoimmunity, vasculopathy and fibrosis. The hallmark of Systemic Sclerosis is skin thickening. Systemic Sclerosis sine scleroderma is a variant of Systemic sclerosis which shares visceral, serological and vascular manifestations but lacks skin thickening. Systemic Sclerosis sine scleroderma still remains a rarely reported subtype of Systemic Sclerosis unlike commonly reported limited cutaneous and diffuse cutaneous types.

Objectives: Aim of this study was to analyse the characteristics of patients with Systemic Sclerosis sine scleroderma from a cohort of Systemic Sclerosis patients.

Methods: This study was done at Institute of Rheumatology, Madras Medical College, Chennai, India. It was a retrospective observational study (January 2006 to November 2016). Patients satisfying ACR/EULAR 2013 Classification criteria for Systemic Sclerosis were included in this study. Patients with Overlap syndrome, MCTD, UCTD, children and pregnant women were excluded. Poormoghim criteria was used for diagnosis of Systemic Sclerosis sine scleroderma: Absent skin

thickening +Raynaud's Phenomenon or peripheral vascular involvement +Positive ANAs+One visceral organ involvement typical of SSc +Absence of another defined connective tissue disease. Demographic, clinical and laboratory profile of all patients were analysed.

Results: Among the 249 patients included in this study, patients with limited cutaneous systemic sclerosis were 59.4% (n=148) and diffuse cutaneous systemic sclerosis were 31.3% (n=78). 9.2% (n=23) of patients satisfied poormoghim criteria for Systemic Sclerosis sine scleroderma. The following characteristics of Systemic Sclerosis sine scleroderma were observed in our cohort. Dyspnoea (82.6%) was the most common presenting complaint and Raynauds (100%) the most common sign observed. Most common internal organ involvement was oesophageal dysfunction (78%) followed by lung involvement in the form of interstitial lung disease (52.1%) and pulmonary hypertension (47.8%). Renal involvement was not observed in any of the patients. Most common autoantibody detected among patients with Systemic Sclerosis sine scleroderma was anti-centromere antibody (78.2%) followed by anti-scl70 antibody (21.8%). Nine patients (39.1%) received pulse cyclophosphamide therapy and 3 patients received rituximab therapy in view of interstitial lung disease.

Abstract AB0736 – Figure 1

Conclusions: The most common pattern of Systemic Sclerosis sine scleroderma observed in our cohort was Oesophageal dysfunction +Raynauds Phenomenon +Anti centromere antibody positivity +Interstitial lung disease with or without pulmonary hypertension which are usually the characteristics of limited cutaneous type of Systemic Sclerosis. Hence Systemic Sclerosis sine scleroderma could be a subset of limited cutaneous type without the phenotypic skin involvement.

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AB0737

AMINAPHTONE INCREASES SKIN BLOOD PERFUSION AND IMPROVES CLINICAL SYMPTOMS IN PATIENTS WITH RAYNAUD'S PHENOMENON INDEPENDENTLY FROM DIFFERENT TREATMENT BACKGROUNDS

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Background: Aminaphtone is a vasoactive drug that was recently demonstrated to improve both peripheral blood perfusion (BP) and clinical symptoms of Raynaud's phenomenon (RP) in patients with either primary or secondary RP to systemic sclerosis (SSc).¹⁻²

Objectives: The aim of this study was to evaluate possible interferences of different treatment backgrounds on both skin BP and RP-related clinical symptoms in patients treated with aminaphtone, during a six-month follow-up.

Methods: Forty-six patients with active RP were enrolled during routine clinical assessment after informed consent (11 primary RP, mean age 49±19 SD years, mean RP duration 6±3 years; and 35 secondary RP to systemic sclerosis, mean age 61±17 years, mean RP duration 11±9 years). Aminaphtone was orally administered 75 mg twice daily in addition to current treatments, and all patients were on a stable drug regimen for at least two months, which remained unmodified during the follow-up. All patients were taking cardioaspirin. Six groups of treatment backgrounds were identified: 1) no further treatments (12 patients); 2) hydroxy-chloroquine (2 patients); 3) colchicine (5 patients); 4) methotrexate (3 patients); 5) cyclosporine A (6 patients); 6) mycophenolate (6 patients); 7) proton-pump inhibitors (12 patients). Blood perfusion was measured by Laser Speckle Contrast Analysis (LASCA)³ at the level of fingertip, periungual areas, dorsum and palm of hands, and face at baseline (T0), after one (T1), four (T4), twelve (T12) and twenty-four (T24) weeks of treatment. Raynaud's condition score (RCS) and both frequency and duration of Raynaud's attacks were assessed at the same time. Statistical analysis was performed by non-parametric tests.

Results: During aminaphtone treatment, a progressive statistically significant increase of blood perfusion, as well as an improvement of RP clinical symptoms (decrease of RCS, frequency and duration of RP attacks/day), were observed in all above reported seven groups of RP patients with different treatments backgrounds from T0 to T12 in all skin areas (p<0.01). There were no statistically significant difference between the seven groups of patients concerning skin BP at different times (p=0.60). The results were similar in both primary and secondary (SSc) RP patients (p=0.40). Aminaphtone administration had to be stopped in 2 patients due to headache, and one patient was lost during follow-up.

Conclusions: This study demonstrates that the increase of skin blood perfusion and the improvement of RP clinical symptoms is not influenced by different treatment backgrounds in RP patients treated with aminaphtone. These preliminary results should be further confirmed by a randomised blind clinical trial.

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AB0738

IMPROVEMENT OF DERMAL THICKNESS IN SYSTEMIC SCLEROSIS PATIENTS TREATED WITH ENDOTHELIN RECEPTOR ANTAGONIST: LONG TERM STUDY BY HIGH FREQUENCY ULTRASOUND

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Background: Systemic sclerosis (SSc) is a connective tissue disorder characterised by skin involvement, which may be evaluated by both modified Rodnan skin score (mRSS) and skin high frequency ultrasound (US).¹⁻⁴ Endothelin-1 (ET-1) seems implicated in the development of dermal fibrosis in SSc.⁵ Bosentan, a dual ET-1 receptor antagonist seems effective in reducing skin fibrosis in SSc patients.^{6,7}

Objectives: The aim of this study was to evaluate by US the long-term effects of bosentan on dermal thickness (DT) in SSc patients, in combination with long-term cyclic intravenous iloprost versus iloprost monotherapy.

Methods: Thirty-eight SSc patients were enrolled during their standard treatment for digital ischemia. At baseline (T0), 19 patients already receiving cyclic intravenous infusion of iloprost (5 continuous days, average 80 mcg/day, every two