patients with at least one image per finger. For each patient, we examined number of capillaries (mean number of capillaries per mm in the distal row), enlarged and giant capillaries, micro-hemorrhages, avascular areas, ramified capillaries, and the presence of a scleroderma (SSc)-like pattern, according to Manfredi et al. Finally, we correlated NVC features with clinical and serological findings of ASSD patients.
Results: The NVC of 54 ASSD patients were analyzed (males/females 1/6.8, mean age 55.79, CI95% 51.9–59.9 years, mean disease duration 59.4, CI95% 27.9–90.9 months). Raynaud's phenomenon (RP) was recorded in 51.9% of patients, arthritis in 79.6%, myositis in 53.7%, and ILD in 92.6%. NVC alterations were observed in 53.7% of AASD patients. Nineteen patients (35.2%) showed a SSc-like pattern; the main features were disarrangement of hairpin and angiogenetic aspects (42.6%), avascular areas (38.9%), giant capillaries (27.6%), and microhemorrhages (20.4%). Finally, the mean number of capillaries was reduced (7.8±2/mm). No significant association was recorded between SSc-like pattern and the presence of arthritis, myositis, and ILD, nor with RP. Among other NVC features, angiogenesis was significantly associated to female gender (p=0.031), while microhemorrhages were inversely associated to the presence of arthritis (0.033). No association was observed between NVC features and autoantibodies profile. Of interest, in 58% of patients with ILD we observed at least a NVC alteration vs no patients without ILD (p=0.04). Finally, in patients with RP NVC alterations were recorded in 15/28 patients (53.6%) and a SSc-like pattern in 11/28 (39.3%), while only 57.9% of patients with SSc-like pattern had a clinically manifest Raynaud's phenomenon.
Conclusions: Despite preliminary, the present is the first study concerning NVC in AASD patients. Regardless of the presence of Raynaud's phenomenon, NVC alterations are frequently observed; in particular, a SSc-like pattern is recorded in more than 1/3 of patients. NVC should be performed in all ASSD patients at diagnosis regardless of the presence of RP in the patient history and during follow-up. ASSD should be always considered in the screening of RP. A prospective multicenter study has been planned to identify specific patterns and possible associations between NVC findings and clinical and serological features of ASSD.
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AB0657 SMALL INTESTINAL BACTERIAL OVERGROWTH IN RELATION TO GASTROINTESTINAL SYMPTOMS IN SYSTEMIC SCLEROSIS

M.D.P. Cruz-Dominguez[1], G. Garcia-Collinot[1], E.O. Madrigal-Santillan[2], A. Carranza-Muleiro[1], L.J. Jara[3], D.H. Montes-Cortes[4], G. Medina-Garcia[1], O. Vera-Lastra[5]. *[1]Research Division, Instituto Mexicano del Seguro Social, Hospital de Especialidades CMN la Raza; [2]Posgrade, IPN, ESM; [3]Research and Education Direction, Instituto Mexicano del Seguro Social, Hospital de Especialidades CMN la Raza; [4]Emergency Room, Hospital General CMN la Raza; [5]Internal Medicine, Instituto Mexicano del Seguro Social, Hospital de Especialidades CMN la Raza, Mexico, Mexico*

Background: Autonomic dysfunction, smooth muscle fibrosis and vascular damage lead to small intestinal bacterial overgrowth (SIBO) in Systemic Sclerosis (SSc). SIBO is characterized by diarrhea, abdominal pain, bloating, malabsorption and malnutrition.
Objectives: To evaluate by NIH PROMIS® gastrointestinal symptoms scales and SIBO by hydrogen breath test (HBT) in patients with SSc.
Methods: We include 68 patients with SSc (ACR-EULAR 2015) who signed informed consent. NIH PROMIS®questionarie was applied to evaluated gastrointestinal symptoms and classified in not symptomatic, least, mildy, moderately and most symptomatic. Glucose HBT was applied after 14 hours fast, oral hygiene and 30 days free of antibiotics. Patients who has a negative HBT with symptoms associated to glucose ingestion we repeat test with lactulose.
Results: We applied questionnaire to 58 SSc patients, age 52 (26–75)years, 65 (96%) female and 3 (4%) males, disease duration 13 (1–40) years, limited SSc 41 (59%) and diffuse 27 (41%), body mass index 24 (12–39).
They are using prednisone (28%), micofenolate (14%), methotrexate (19%), azatioprin (5%), amlodipine or nifedipine (33%). Patients had continuous and very high increase of parts per millon (ppm) of exhaled Hydrogen: min0: 13 ppm (5–21), min15:17 ppm (5–43), min30:17 (3–49), min45:18ppm (7–103), min 60:22ppm (8–145), min90:18ppm (2–250), min120:25ppm (3–212), min150:71ppm (3–235). Normal values: <10 ppm during total test (Figure1).
Frequency of gastrointestinal symptoms were flatulence (87.5vs81.2%), nausea/vomiting (72.7vs37.6%), constipation (65.6vs40%), diarrhea (45.2vs33.4%), abdominal pain %) and incontinence (39.4vs31.3%) respectively between SCB (+) positive and negative.
Hyperproduction of hydrogen in breath had a direct correlation to severity of their symptoms ($p \leq 0.05$). The severity of diarrhea was in close relation to the severity of its rectal incontinence (r=0.73,p=0.001), and greater abdominal pain with flatulence (r=72, p=0.001).
Conclusions: Gastrointestinal symptoms are common in SSc regardless of whether they have SIBO. However, a higher Row Score SGI or moderate severe status (NIH PROMIS) correlates with high H scores from the 30th minute, therefore, the questionnaire is useful within the SSc assessment.

Spearman Correlation Row Score(NIH PROMIS) with ppm H+ Test of SIBO		overgrowth (SIBO)				
		Negative		Positive		
		n=24	%	n=44	%	p
Nausea and vomiting r=0.28*min45,0.29*min60,0.43*min90,0.49*min120,0.46*min150	Asymptomatic	5	9.8%	2	3.9%	
	Least	2	3.9%	2	3.9%	
	Mild	3	5.9%	6	11.8%	0.13
	Moderate	3	5.9%	7	13.7%	
	Severe	4	7.8%	17	33.3%	
Diarrhea, r=0.43*min120	Asymptomatic	3	5.9%	10	19.6%	
	Least	3	5.9%	3	5.9%	
	Mild	5	9.8%	6	11.8%	0.34
	Moderate	5	9.8%	7	13.7%	
	Severe	1	2.0%	8	15.7%	
Constipation r=0.41*min90, 0.47*min120, 0.043*min150	Asymptomatic	0	0.0%	4	8.2%	
	Least	2	4.1%	1	2.0%	
	Mild	8	16.3%	6	12.2%	0.05
	Moderate	2	4.1%	11	22.4%	
	Severe	4	8.2%	11	22.4%	
Abdominal pain r=0.32*m120	Asymptomatic	1	2.0%	0	0.0%	
	Least	7	13.7%	9	17.6%	
	Mild	6	11.8%	8	15.7%	0.16
	Moderate	1	2.0%	4	7.8%	
	Severe	2	3.9%	13	25.5%	
Distension r=0.36*m0,0.35*min15, 0.3*min45,0.37*min60, 0.386*min90,0.36*min120	Asymptomatic	1	2.0%	1	2.0%	
	Least	2	4.0%	1	2.0%	
	Mild	1	2.0%	2	4.0%	0.08
	Moderate	8	16.0%	6	12.0%	
	Severe	5	10.0%	23	46.0%	
Incontinence r=0.28*min45, 0.37*min120	Asymptomatic	8	15.7%	17	33.3%	
	Least	0	0.0%	0	0.0%	
	Mild	5	9.8%	3	5.9%	0.24
	Moderate	2	3.9%	8	15.7%	
	Severe	2	3.9%	6	11.8%	
* p<0.05						

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AB0658 FACTORS INFLUENCING ELAPSED TIME TO A DEFINITIVE DIAGNOSIS OF SYSTEMIC SCLEROSIS AND THEIR RELATIONSHIP WITH THE NUMBER OF ATTENDING PHYSICIANS AND THEIR MEDICAL SPECIALIZATION

Y. Erez[1], H. Yarkan Tugsal[2], G. Kenar[2], B. Zengin[2], D. Solmaz[3], M. Ozmen[3], G. Can[2], N. Akkoc[2], F. Onen[2], M. Birlik[2]. *[1]Internal Medicine; [2]Rheumatology, Dokuz Eylul University School of Medicine; [3]Rheumatology, Katip Celebi University School of Medicine, Izmir, Turkey*

Background: The diagnosis of Systemic Sclerosis (SSc) can be difficult due to its rarity and heterogeneity. In addition, not all physicians are expert in the identification of patients with early SSc features.
Objectives: The aim of this study was to investigate factors influencing the time elapsed since a proper diagnosis of SSc and their relationship with the number of attending physicians and their specialization, among Turkish patients. In Turkish healthcare system,each patient may directly go to the hospitals and choose appropriate care facility and physician.
Methods: The study covers 240 SSc patients who were diagnosed according to 1980 ACR criteria in the rheumatology database at the University of Dokuz Eylul, Izmir. Data included demographics, SSc duration, SSc subtype, physical examinations (mRodnan Score-mRS), and a face-to-face structured interview. Cases were excluded if they couldn't recall date of initial symptoms and all the prior attending physicians including their specialization until a definitive diagnosis of SSc. As clinical characteristics are different than pure SSc patients, overlap syndromes were excluded. Mann-Whitney U test was used to assess the difference between dependent and independent variables. The association between mRS and dependent variables was evaluated by Spearman correlation test.
Results: Remaining 135 patients (F:88.8%), mean age±SD (52.14±11.58 years) identified. 55 (41%) patients had diffuse and 80 (59%) patients had limited disease. 119 (88%) patients SSc diagnosed by a rheumatologist however, 48 patients (35%) were first seen by an internist. Median time to diagnosis was 36 months (1–588) from the onset of Raynaud Phenomenon (RF) and 11 months (0–397) from the onset of the first non-RF symptom for our cases. Relation between some parameters and time to diagnosis were shown in Table.1. Mean number of attending physicians until SSc diagnosis was calculated as 3,54 (SD±1,66). None of the demographic and clinical parameteres (SSc subtype, mRS, symptom duration) were related with the number of attending physician. There was no correlation detected between mRS and time to diagnosis and number of attending physicians. Nevertheless it was established that median number of physicians was less for patients who were referred to a proper clinician by their familier healthcare practitioners (p: 0,019).

Table 1. Time to diagnosis from Raynaud's and first non-Raynaud's symptom onset stratified clinical and demographic parametres

	Time to diagnosis from RF median-months	p	Time to diagnosis from first non-RF median-months	p
Sex				
Female	19 (4-299)	>0,05	11 (0-397)	>0,05
Male	39 (1-588)		15 (0-277)	
Education situation				
Less than highschool	44(0-588)	>0,05	11(0-397)	>0,05
Highschool and more	20(0-288)		14(0-297)	
Scl subtype				
Diffuse	23 (1-299)	***0.013****	11(0-297)	>0,05
Limited	45 (1-588)		11.5(0-397)	
Referred by healthcare practitioner				
Positive	24 (1-349)	>0,05	11(0-191)	>0,05
Negative	42 (1-588)		11(0-397)	
Initial symptom				
RF	49 (0-588)	***0.001***		
RF-non-RF together	21 (0-470)			

RF-non-RF together: less than one year between onset of RF and first non-RF symptom

Conclusions: Time to diagnosis from onset of RF is significantly shorter in diffuse SSc. Although mRS is typically higher in diffuse SSc, no correlation has been found between mRS and time to diagnosis. Therefore, duration of diagnosis may be influenced by internal organ involvement and other complications rather than skin changes due to lack of awareness of physicians about SSc related early skin changes. Referral to a proper physician by a familier healthcare practitioner decreased the number of physicians attending till the diagnosis, although it did not affect the time to diagnosis.

Disclosure of Interest: None declared

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AB0659 ASSOCIATION BETWEEN -12518A/G GENE POLYMORPHISM ENCODING MONOCYTE CHEMOATTRACTANT PROTEIN 1 (MCP-1) AND SERUM LEVEL OF C-REACTIVE PROTEIN IN DIFFERENT CLINICAL AND SEROLOGICAL PHENOTYPES OF SYSTEMIC SCLEROSIS IN THE RUSSIAN COHORT OF PATIENTS

M. Krylov, L. Ananieva, O. Koneva, M. Starovoytova, O. Desinova, O. Ovsaynnikova, E. Samarkina, A. Novikov, E. Aleksandrova. *V. A. Nasonova Research Institute of Rheumatology, Moscow, Russian Federation*

Background: Immune system activation with associated up-regulation in the production of extra-cellular matrix proteins by fibroblasts are known specific features in the pathogenesis of systemic scleroderma (SSc). Most recent data indicate that MCP-1 and MCP-3 chemokines from the family of monocyte chemoattractant proteins are also involved into SSc pathogenetic process. C-reactive protein (CRP) is known as the marker of acute-phase inflammation. The association between increased CRP levels and SSc clinical and serological parameters has been reported recently.

Objectives: To study the association between -2518 A/G gene polymorphism, encoding *MCP-1*, and CRP levels in different clinical SSc phenotypes in the Russian cohort of pts.

Methods: PCR-RFLP method was used to identify *MCP-1* genotype in 81 SSc pts aged 49,4±12,6 years, with mean SSc duration 11,1±9,0 years. CRP concentrations were measured with highly sensitive immunoturbidimertry method.

Results: CRP levels were correlated with *MCP-1* genotypes in pts with limited (lcSSc) and diffuse (dcSSc) phenotypes, with interstitial lung disease (ILD+), with SSc duration >3 years, with increased CRP level (>5 mg/L), with positive antibody titers to DNA topoisomerase I (ATA+) and antibody to centromeres (ACA+). A total cohort analysis showed that carriers of -2518AA genotype had higher mean CRP level versus G allele carriers (12,6±7,5 mg/L vs 4,9±4,8 mg/L, respectively, p=0,009), although similar trend was found in dcSSc phenotype (16,4±19,5 mg/L vs 6,1±4,4 mg/L, respectively, p=0,040). In pts with -2518AA genotype and SSc duration >3 years mean CRP level was significantly higher than in G allele carriers (1,1±16,7 mg/L vs 4,5±4,4 mg/L, respectively, p=0,025). In (ILD+) and (ATA+) subgroup pts with -2518AA genotype demonstrated higher mean CRP levels as compared to G allele carriers (12,4±15,6 mg/L vs 5,5±5,1 mg/L, respectively, p=0,018; and 17,6±20,9 mg/L vs 5,5±5,5 mg/L, respectively, p=0,010). CRP levels (>5 mg/L) were found in 31 (38%) pts and were significantly different between AA genotype carriers and G allele carriers (27,4±19,2 mg/L vs 10,4±3,8 mg/L, respectively, p=0,003). No associations between genetic variations in the *MCP-1* gene and CRP levels in lcSSc phenotype, SSc duration <3 years, CRP levels <5mg/L and (ACA+) pts were established.

Conclusions: Our data demonstrate that -2518A/G *MCP-1* gene polymorphism is closely associated with CRP levels, thus, it can be considered as a new marker, reflecting the severity of the disease and unfavorable SSc prognosis.

Disclosure of Interest: None declared

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AB0660 CHARACTERISTICS OF AL-I LIGHT-CHAIN AND AMYLOID A DEPOSITION IN PROGRESSIVE SYSTEMIC SCLEROSIS – A COMPARATIVE POSTMORTEM CLINICOPATHOLOGIC STUDY OF 12 PATIENTS

M. Bély[1], Á. Apáthy[2]. *[1]epartment of Pathology, Policlinic of the Order of the Brothers of Saint John of God; [2]Department of Rheumatology, St. Margaret Clinic Budapest, Budapest, Hungary*

Background: Different types of amyloid deposits may be present in systemic sclerosis (SSc), as consequences (complications) of basic or associated diseases.

Objectives: The aim of this study was to determine the type, prevalence and extent of amyloid deposits on different tissue structures in various organs in SSc.

Methods: We studied histopathologically 12 SSc patients (females 11, average age: 54.82 years, range 66–32, onset of SSc: 48.86, average disease duration: 6.43 years; one male, age 65.0 years at death, onset of SSc and average disease duration not known, who died at the National Institute of Rheumatology. SSc was diagnosed clinically according to the criteria of the ACR [1]. In 1 (8.0% of 12) 67 year old female patient (onset of SSc: 66 years, disease duration: 1 year) SSc was accompanied by B-cell lymphoma and complicated by systemic AL-l light-chain amyloidosis. In 1 (8.0% of 12) 53 year old female patient (onset of SSc: 41 years, disease duration: 12 years) SSc was complicated by systemic amyloid A (aA) deposition.

Amyloid deposits on different tissue structures [arteriole, small artery, medium size artery, venule, small vein, medium size vein, interstitial collagen fiber, reticulin fiber (collagen IV), and nerve] of 6 organs [heart, lungs, kidney, gastrointestinal tract, skin and brain] were determined histologically. The extent of amyloid deposition was evaluated by semi-quantitative, visual estimation on a 0 to 3 plus scale, based on the number of involved tissue structures per light microscopic field [2].

The prevalence and extent of amyloid-l light-chain and amyloid A deposits on different tissue structures were compared by Student (Welch) t-probe.

Results: The involvement of different tissue structures (prevalence in %) and the average extent of AL-l light-chain and amyloid A deposits (absolute value) are summarized in Table 1.

Table 1

Tissue structures	SSc-l Prevalence in %	SSc-aA Prevalence in %	p<	SSc-l Average extent	SSc-aA Average extent	p<
Arteriole	83,33	66,67	0,2749	2,00	1,33	0,1709
Small artery	83,33	66,67	0,1709	2,17	1,00	0,0414
Medium size artery	83,33	66,67	0,1520	2,33	0,67	0,0090
Interstitial collagen	66,67	50,00	0,2998	0,83	0,83	0,5000
Medium size vein	50,00	50,00	0,2735	1,17	0,50	0,1476
Small vein	50,00	50,00	0,5000	0,58	0,50	0,4088
Venule	33,33	33,33	0,5000	0,33	0,33	0,5000
Collagen IV	33,33	16,67	0,2749	0,50	0,33	0,3671
Nerve	0,00	0,00		0,00	0,00	
Average/Structure	53,70	44,44	0,232	1,10	0,61	0,075

Conclusions: In SSc patients the prevalence and extent of l light-chain and amyloid A deposits on different tissue structures changed parallel.

The higher prevalence and extent of l light-chain deposits in contrast to amyloid A may be explained with qualitative differences of l light-chain and amyloid A; l light-chain seems to have greater affinity for tissues than amyloid A protein.

Infiltration of the vessel walls – regarding the amount of l light-chain and amyloid A deposits in arterioles and arteries in contrast to the veins – showed a converse tendency in SSc patients with AL-l or AAa. This may be related to sluggish blood flow or stasis (backward congestion and accumulation of circulating precursors) in both diseases.

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AB0661 ORAL HEALTH-RELATED QUALITY OF LIFE MEASURED WITH OHIP 49 HIGHLY CORRELATES WITH DISEASE ACTIVITY AND SEVERITY IN SYSTEMIC SCLEROSIS PATIENTS

K. Parat[1], M. Radić[2], K. Borić[2], D. Perković[2], D. Biočina Lukenda[1], D. Martinović Kaliterna[2]. *[1]Department of Oral Medicine and Periodontology, Study of Dental Medicine, School of Medicine, University of Split; [2]Division of Rheumatology and Clinical Immunology, Center of excellence for Systemic Sclerosis in Croatia, University Hospital Split, Split, Croatia*

Background: Systemic sclerosis (SSc) is associated with decreased saliva production and interincisal distance, more missing teeth, and periodontal disease. Orofacial manifestations of SSc contribute greatly to overall disease burden and still are regularly overlooked and under-treated. Previous studies did not confirm correlation between disease severity and oral health-related quality of life in SSc patients.