pain in 7(18.4%). Gastrointestinal (GI) symptoms comprised of regurgitation in 31 (81.5%) and dysphagia in 14 (36.8%). Interstitial lung disease was present in 30 (78.9%) patients, with higher prevalence in diffuse scleroderma (100%) than in limited scleroderma (70%) ($p=0.01$). Pulmonary hypertension was present in 18 patients, with significantly higher prevalence in diffuse disease (57.1%), that was secondary to interstitial lung disease and in limited disease it was found in (11.8%) ($p<0.01$). Thirty (78.9%) patients were found to have restricted disease on pulmonary function tests.

Obstetric history showed a higher prevalence of primary infertility in at least 6 (15.8%) patients, with significantly higher prevalence in limited systemic sclerosis disease as compared to diffuse disease (23.5% vs 9.5%, and $p=0.05$). Fibromyalgia diagnosed as per ACR criteria was present in 7 (18.4%) patients, and depression assessed by Hospital Anxiety and depression (HADS) score was present in 10 (26.3%) patients.

Anti nucleic acid antibody (ANA) was found positive in 30 (78.9%) patients. Anti Scl-70 antibodies were in 24 (63.2%) patients, with significant association with diffuse disease (85% vs 35.3% and $p<0.01$), while anti centromere antibodies were present in 20 (52.6%) patients; significantly higher in limited disease (94.2% vs 19.0%, and $p<0.01$).

Conclusion: Scleroderma is a very important, autoimmune multisystem disease. It has female preponderance. Raynaud phenomenon is the most initial clinical feature followed by other manifestations of variable course and disease severity. Interstitial lung disease and pulmonary hypertension were the most important complication found in our patients which has poor prognosis. So, it is imperative to early diagnose and treat the disease manifestations to prevent future complications.

Keywords: Scleroderma, Systemic sclerosis.

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AB0094

INCREASE OF ENDOTHELIAL PROGENITOR CELLS IN SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE

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Background: Endothelial progenitor cells (EPC), involved in vasculogenesis and endothelial tissue repair, have been described as relevant players in vascular and connective tissue diseases [1-2]. In this regard, a previous study of our group disclosed that the degree of EPC frequency may help to identify the presence of interstitial lung disease (ILD) in rheumatoid arthritis patients [3]. Given that ILD is the main cause of mortality in patients with systemic sclerosis (SSc) [1, 4-6], the understanding of the role of EPC in the mechanism of SSc-ILD⁺ vasculopathy is crucial.

Objectives: To assess the potential role of EPC on vascular dysfunction associated with the presence of ILD in patients with SSc.

Methods: Peripheral venous blood was collected from a total of 39 patients with SSc, 20 with ILD (SSc-ILD⁺) and 19 without ILD (SSc-ILD⁻). All subjects were recruited from the Rheumatology and Pneumology departments of Hospital Universitario Marqués de Valdecilla, Santander, Spain. Quantification of EPC was analyzed by flow cytometry. EPC were considered as CD34⁺, CD45^{Low}, CD309⁺ and CD133⁺.

Results: Statistically significant differences in EPC frequency between patients with SSc-ILD⁺ and patients with SSc-ILD⁻ were disclosed. Specifically, an increase of EPC frequency was observed in SSc-ILD⁺ patients when compared to patients with SSc-ILD⁻ (mean $\pm$ standard deviation: 0.033 ± 0.012 versus 0.021 ± 0.017 , respectively, $p=0.012$).

Conclusion: Our results suggest a potential role of EPC on vascular damage associated with the manifestation of ILD in patients with SSc.

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AB0095

EXPRESSION AND PATHOGENIC ROLES OF INTEGRIN FAMILY GENE IN SYSTEMIC SCLEROSIS

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Background: Emerging evidence have shown that some integrin members are associated with inflammation and fibrosis in systemic sclerosis (SSc) patients^[1,2]. However, the expression patterns and pathogenic significance of the whole integrin family in SSc are still unclear.

Objectives: This study aimed at evaluating the integrin family gene expression in skin lesion from SSc patients and exploring its potential pathogenic mechanism.

Methods: We utilized the public datasets of SSc skin tissue from Gene Expression Omnibus (GEO) database to analyze the expression and clinical significance of integrin family genes in SSc. In addition, functional enrichment and pathway analysis were also conducted.

Results: Compared with healthy controls, ITGA5, ITGA7, ITGA8, ITGB2, ITGB5, ITGAE and ITGB3BP were abnormally overexpressed in the skin of SSc. Further analysis indicated that ITGA5, ITGA7, ITGA8, ITGB2 and ITGB5 were positively correlated with modified Rodnan skin thickness score (mRSS), while ITGAE and ITGB3BP were negatively correlated with mRSS in SSc. Increased ITGB5 expression was associated with positive of anti-centromere antibody (ACA). Functional enrichment and pathway analysis showed that integrin members had multiple functions in SSc. Among them, ITGA5, ITGB2 and ITGB5 might synergistically promote SSc through affecting extracellular matrix (ECM) turn over, ECM-receptor interaction, focal adhesion and leukocyte trans-endothelial migration. ITGA5 and ITGB5 also affected angiogenesis and endothelial cell function. In addition, ITGA5 was uniquely enriched for actin organization, ITGB5 was uniquely enriched for TGF- β signaling, and ITGB2 was uniquely associated with immune cells activation.

Conclusion: Our results implied that integrins, especially ITGA5, ITGB5, ITGB2 participated in the process of inflammation, vasculopathy and fibrosis in SSc. Together, they might render important therapeutic targets for SSc.

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Vasculitis - aetiology, pathogenesis and animal models

AB0096

IGA VASCULITIS AND IGA NEPHROPATHY SHARE A SIMILAR IL17A ASSOCIATION PATTERN

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Background: IgA vasculitis (IgAV) and IgA nephropathy (IgAN) are inflammatory conditions that share pathogenic and molecular mechanisms [1] and may represent different outcomes of a continuous spectrum of disease [2]. *Interleukin (IL)17A* has been identified as a common genetic risk locus for several immune-mediated diseases [3, 4].

Objectives: To determine whether IgAV and IgAN exhibit a different *IL17A* association pattern.

Methods: Five *IL17A* tag polymorphisms (rs4711998, rs8193036, rs3819024, rs2275913 and rs7747909) were genotyped in 388 Caucasian patients with IgAV, 99 patients with IgAN and 1,003 sex and ethnically matched healthy controls.

Results: No statistically significant differences between patients with IgAV and healthy controls and between patients with IgAN and healthy controls were observed when each *IL17A* genetic variant was analyzed independently (Table 1). Similarly, IgAV patients exhibited similar genotype and allele *IL17A* frequencies than those with IgAN (Table 1). Moreover, no genotype or allele differences between IgAV patients who developed nephritis and patients with IgAN were detected. Furthermore, haplotype frequencies were similar in patients with IgAV, IgAV and nephritis and those with IgAN.

Table 1. Genotype and allele frequencies of *IL17A* gene in patients with IgA vasculitis, patients with IgA nephropathy and healthy controls.

Polymorphism	Change	Data set	1/1	1/2	2/2	1	2
rs4711998	G/A	IgAV	53.4 (207)	38.9 (151)	7.7 (30)	72.8 (565)	27.2 (211)
		IgAN	49.0 (48)	42.9 (42)	8.2 (8)	70.4 (138)	29.6 (58)
		Controls	52.7 (529)	41.2 (413)	6.1 (61)	73.3 (1471)	26.7 (535)
rs8193036	T/C	IgAV	57.0 (221)	38.4 (149)	4.6 (18)	76.2 (591)	23.8 (185)
		IgAN	64.3 (63)	31.6 (31)	4.1 (4)	80.1 (157)	19.9 (39)
		Controls	60.3 (605)	35.2 (353)	4.5 (45)	77.9 (1563)	22.1 (443)
rs3819024	A/G	IgAV	44.1 (171)	43.3 (168)	12.6 (49)	65.7 (510)	34.3 (266)
		IgAN	39.4 (39)	54.5 (54)	6.1 (6)	66.7 (132)	33.3 (66)
		Controls	45.6 (457)	44.6 (447)	9.9 (99)	67.8 (1361)	32.2 (645)
rs2275913	G/A	IgAV	44.6 (172)	43.3 (167)	12.2 (47)	66.2 (511)	33.8 (261)
		IgAN	39.8 (39)	53.1 (52)	7.1 (7)	66.3 (130)	33.7 (66)
		Controls	44.8 (449)	44.2 (443)	11.1 (111)	66.8 (1341)	33.2 (665)
rs7747909	G/A	IgAV	53.9 (209)	39.4 (153)	6.7 (26)	73.6 (571)	26.4 (205)
		IgAN	41.1 (39)	54.7 (52)	4.2 (4)	68.4 (130)	31.6 (60)
		Controls	53.0 (532)	39.4 (395)	7.6 (76)	72.7 (1459)	27.3 (547)

Conclusion: Our results revealed that IgAV and IgAN share a similar *IL17A* association pattern.

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AB0097

DIAGNOSTIC ACCURACY OF SERUM MARKERS IN LARGE VESSEL VASCULITIS AND CORRELATION WITH PET IMAGING

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Background: The onset of symptoms in large-vessel vasculitis (LVV) tends to be subacute, which often leads to a delay in diagnosis, during which time vascular disease may start and progress to become symptomatic. PET/CT has been recognised since the 2003s as a promising tool in evaluating of LVV. There is no gold standard diagnostic laboratory tests for this patient group, novel markers for active LVV is needed.

Objectives: to investigate the association between vascular inflammation, as detected by PET imaging and interleukin-6 (IL-6), pentraxin3 (PTX3), and B-cell-activating-factor (BAFF) in subjects with LVV.

Methods: The study included 67 patients patients with newly diagnosed GCA (n= 27) or TA (n=9) and healthy control (n= 31) who had been referred to the Rheumatology Unit at Gazi University, between December 2017 and August 2020. PET images obtained from an 29 patients (22 with GCA and 7 with TA) who had not received any corticosteroid treatment prior to PET imaging and blood sampling. IL-6, PTX3, and BAFF levels were determined quantitatively by enzyme-linked immunosorbent assay (ELISA) kits.

Results: 36 patients with LVV (20 females, 16 males; age 64.5±16.6) and 31 HC (14 females, 17 males; age 37.1±9.6) were analysed. Serum levels of IL-6, PTX3, BAFF, ESR and CRP are increased in patients with newly diagnosed LVV compared with those in control subjects. In a ROC analysis, serum IL-6 provided excellent discrimination of newly diagnosed LVV patients from HC, as indicated by AUCs>0.90. Serum BAFF also accurately distinguished newly diagnosed LVV patients from HC with AUCs>0.80. Serum PTX3 did not provide an AUC>0.80. In this study, we correlate vascular inflammation, as detected by PET imaging in newly diagnosed LVV patients, with the ESR, CRP, PTX3, IL-6 and BAFF. As a result, none of these markers has been associated with vascular inflammation as measured using PET.

Conclusion: In conclusion, our study shows that serum levels of PTX3, IL-6 and BAFF are increased in most LVV patients. The diagnostic value of BAFF and IL-6, both separately and in combination, should be further evaluated in larger cohorts of LVV patients, as well as in patients with infections or other inflammatory conditions. However, none of these markers has been associated with vascular inflammation as measured using PET.

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