pain in 71.8%). Gastrointestinal (GI) symptoms comprised of regurgitation in 31 (81.5%) and dysphagia in 14 (36.8%). Intestinal 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 intestinal 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 preponderence. Raynaud phenomenon is the most initial clinical feature followed by other manifestations of variable course and disease severity. Intestinal 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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AB0005 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 [1-5]. 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, ITGB4, ITGB5, ITGA8 and ITGB3BP were abnormally overexpressed in the skin of SSc. Further analysis indicated that ITGA5, ITGA7, ITGB4, ITGB5 and ITGB8 were positively correlated with modified Rodnan skin thickness score (mRSS), while ITGA5 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) turnover, 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, ITGB2 and ITGB5 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

AB0008 IGA VASCULITIS AND IGA NEPHROPATHY SHARE A SIMILAR IL17A ASSOCIATION PATTER


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