LETTER TO EDITOR

Role of Autoantibodies in Scleroderma

Dear Editor,

It is a well-known fact the immune system plays a protective role in disease prevention. However, when the immune system acts against its tissues and produces antibodies against the self-components of the body, it leads to the production of autoantibodies. There is usually a close association between a particular autoantibody and a particular form of disease or its clinical symptoms. Levels of a specific type of autoantibody in blood can reflect disease activity. Scientists continue to look for and identify different types of these autoantibodies as potential biomarkers for specific diseases. We have been working on the role of autoantibodies in many diseases including scleroderma.

Scleroderma, or Systemic Sclerosis or SSc, is a fibrotic, rare heterogeneous autoimmune disease in which immune and circulatory system abnormalities contribute to various dermatological and systemic abnormalities.1 The heterogeneity and wide variation in organ involvement, disease severity, rate of disease progression, and survival between patients are prominent characteristics of SSc. Additionally, when compared to other rheumatic diseases, SSc is linked to the greatest loss of life expectancy(more than 30 years for female patients and 16 years for male patients).2 Scleroderma is diagnosed using various diagnostic criteria, the most prevalent of which is the American College of Rheumatology (ACR) criteria. The patterns of organ involvement and prognosis are well predicted by the SSc-specific autoantibodies like antinuclear antibodies (ANA) subtypes associated with various clinical presentations, disease phenotypes, and prognosis.3 Antinuclear antibodies (ANA) may be present in more than 90% of cases with systemic sclerosis, and up to 70% of cases may also contain at least one of the more targeted autoantibodies (anti-centromere, anti-topo-I, or anti-RNA polymerase III).4Autoantibody status in SSc appears to be influenced by a variety of ethnicity-related variables, including genetic and environmental factors. Therefore, data from various ethnic groups would provide light on the precise association between the disease's immunogenetic background and the associated autoantibody response. The authors Sakeena et al.5 have also conducted a study on the association of autoantibodies with clinical characteristics in limited and diffuse subsets of Scleroderma in the Kashmiri population. About 92.5% of SSc patients were positive for ANA after autoantibody testing. Of these ANA-positive patients, 46.25% were of the dcSSc subset and 53.75% were of the lcSSc subset of Scleroderma. 68.75% of patients were anti-topo I (ATA) positive and 31. re found anti-centromere antibody (ACA) positive. When using autoantibodies to assess clinical symptoms, it was found that pulmonary fibrosis was more common in the ATA group of patients. At the same time, calcinosis, arthralgia, and telangiectasia were more common in the ACA group. There was a significant relationship between autoantibodies and disease characteristics such as gangrene, swollen fingers, and pulmonary fibrosis.70% of individuals with pulmonary fibrosis tested positive for ATA ($p<0.01$). ATA positivity was associated with a higher probability of interstitial lung disease and vascular abnormalities.

Anti-topo I positivity was associated with a higher frequency of interstitial lung disease (ILD). Perera et al.6

Correspondence:
Dr. Zafar A Shah, Phd
Professor & Head ,
Department of Immunology & MM, SKIMS
email: zaffaramin@gmail.com

Access this article online

Website: www.jmsskims.org

DOI: https://doi.org/10.33883/jms.v27i2.1378

How to cite this article: Ayub S, Shah Z, Rasool R, Dangroo MA, Sofi FA. Role of Autoantibodies in Scleroderma. JMS 2024: 27 (2)43-44

Received: 2024-06-08 Accepted: 2024-06-15

This is an open-access article distributed under the terms of the Creative Commons Attribution License (CC BY 4.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

reported that anti-topo I autoantibody was associated with rapidly progressive diffuse skin involvement and ILD. Current findings confirmed the results of others showing certain autoantibodies are linked to the severity of SSc.7, 8 The type of autoantibody a patient carries rarely changes over time, although clinical characteristics and disease-specific autoantibodies differ across the globe and among ethnic groups. Significant correlations exist between the kind of antibody, the pattern of organ dysfunction, and survival in SSc patients. Autoantibodies have long been thought to be better disease prognosis and organ involvement indicators. Clinical practice routinely tests for the aforementioned disease-specific autoantibodies, which aid in SSc diagnostic confirmation and risk stratification. Autoantibodies are now widely recognized as superior markers of disease outcome and organ involvement due to the strong correlations between antibody type and the pattern of organ dysfunction and survival in SSc patients. Because of this intimate connection, antibody testing is necessary to diagnose diseases.

Thus autoantibodies provide important prognostic data for SSc and help identify the clinical subtypes of the disease. Overall, pairing the antibodies with a specific clinical presentation, risk of complications, and prognosis should be introduced as a clinical standard and another step in the advancement of the management of patients with systemic rheumatic diseases. Research in the field of antibodies and their relationship to specific treatment plans can eventually completely change our understanding of this and many other autoimmune diseases.

REFERENCES

1. Steen VD, Medsger TA. Changes in causes of death in systemic sclerosis, 1972-2002. Ann Rheum Dis. 2007;66(7):940–4.
2. Mok CC, Kwok CL, et al. Life expectancy, standardized mortality ratios, and causes of death in six rheumatic diseases in Hong Kong, China. Arthritis Rheum. 2011;63(5):1182–9.
3. LeRoy EC, Black C, Fleischmajer R, et al. Scleroderma (systemic sclerosis): classification, subsets and pathogenesis. J Rheumatol. 1988;15(2):202–5.
4. Gleicher N, Barad D, Weghofer A. Functional autoantibodies, a new paradigm in autoimmunity? Autoimmun Rev. 2007;7(1):42–5.
5. Sakeena A, Zafar AS, Tabasum S, et al. Molecular typing of HLA-class II alleles reveals an association with autoantibodies and disease subsets of systemic sclerosis in a North Indian (Kashmir) population. The Egyptian Rheu.2023; 45: 247-253.
6. Perera LMB, Sekiguchi A, Uchiyama A, et al. The regulation of skin fibrosis in systemic sclerosis by extracellular ATP via P2Y2 purinergic receptor. J Invest Dermatol 2019;139(4):890–9.
7. Walker UA, Tyndall A, Czirják L, et al. Clinical risk assessment of organ manifestations in systemic sclerosis: a report from the EULAR scleroderma trials and research group database. Ann Rheum Dis2007;66(6):754-63.
8. Dick T, Mierau R, Bartz-Bazzanella P, et al. Coexistence of antitopoisomerase I and anticentromereantibodies in patients with systemic sclerosis. Ann Rheum Dis 2002;61:121–7.

Sakeena Ayub[1], Zafar A Shah[2], Roohi Rasool[3], Mushtaq A Dangroo[4], Fayaz A Sofi[5]

[1-3] Department of Immunology & Molecular Medicine SKIMS

[4, 5] Division of Rheumatology, Department of Internal Medicine, SKIMS