JMEA Journal of Modern Educational Achievements
Volume 6, 2024 https://scopusacademia.org/

The relationship between systemic sclerosis and anti-fibrillarin antibodies.

Sanjarbek Istamovich Zaripov, Nargiza Mirza-Bakhtiyarkhonovna Abdurakhmanova

Tashkent Medical Academy, Tashkent, Uzbekistan.

Abstract. Systemic sclerosis, also known as scleroderma, is a complex autoimmune condition that impacts the body's connective tissues. It involves abnormal growth of connective tissue, resulting in skin and organ fibrosis, as well as vascular issues. A key aspect of systemic sclerosis is the presence of autoantibodies, like anti-nuclear antibodies (ANAs), which target cell nucleus components. Various ANA subtypes linked to systemic sclerosis include anti-centromere, anti-topoisomerase (anti-Scl-70), anti-RNA polymerase III, and anti-U3-RNP/fibrillarin antibodies. These antibodies target specific proteins involved in ribosomal RNA processing. Detecting ANAs, including anti-U3-RNP/fibrillarin antibodies, can aid in diagnosing and managing systemic sclerosis. ANA testing can also help healthcare professionals monitor disease progression and assess risks of complications like pulmonary hypertension and lung disease.

KEYWORDS: anti-fibrillarin antibodies, fibrillarin, anti-nuclear antibodies, Raynaud's phenomenon, SSc (systemic sclerosis).

INTRODUCTION.

Systemic sclerosis, also known as scleroderma, is a complex autoimmune disease characterized by fibrosis and vascular abnormalities. The presence of anti-nuclear antibodies, including anti-U3-RNP/fibrillarin antibodies, is essential for diagnosing and managing this condition. Ongoing research on these autoantibodies may enhance our understanding and treatment of systemic sclerosis in the future. Systemic sclerosis is relatively rare, with an estimated prevalence of 50-300 cases per million individuals. It typically affects adults, with the highest incidence between ages 30 and 50, although it can occur at any age. Women are affected more frequently than men, with approximately three times higher prevalence. There are geographic and ethnic variations in the disease's occurrence, with higher rates among certain populations like Native Americans and African Americans compared to those of European descent. Regional differences exist in the prevalence of specific subtypes of systemic sclerosis and associated autoantibodies. The exact cause of systemic sclerosis, or scleroderma, is not fully understood but is believed to involve a complex interplay of genetic, environmental, and immunological factors. Genetic predisposition may contribute to the disease, as evidenced by familial clustering and specific genetic markers linked to increased risk. However, no single gene has been identified as the primary cause, suggesting multiple genetic factors are involved. Environmental factors such as exposure to chemicals like silica and organic solvents, as well as viral infections, have been proposed as potential triggers for systemic sclerosis. The disease's higher prevalence in women implies hormonal and reproductive factors may also influence its development. Immunological

dysfunction is a crucial aspect of systemic sclerosis, involving an abnormal immune response leading to inflammation, fibrosis, and vascular damage.

An essential feature of systemic sclerosis is the dysregulation of the immune system, characterized by the production of autoantibodies like anti-nuclear antibodies (ANAs) that target various cellular components and are believed to play a role in the disease's development. The development of systemic sclerosis (SSc), or scleroderma, is a complex process involving vascular dysfunction, immune system dysregulation, and fibrosis. This leads to extensive tissue damage and fibrosis in the skin, internal organs, and blood vessels. While the precise sequence of events in SSc pathogenesis remains incompletely understood, several key mechanisms have been identified as driving factors in the disease progression.

Anti-U3-RNP antibodies are a type of autoantibodies commonly detected in systemic sclerosis (SSc) patients. They specifically target the U3 ribonucleoprotein (RNP) complex, which plays a role in ribosomal RNA processing and ribosome assembly. The presence of these antibodies in SSc patients has been linked to specific clinical manifestations and disease characteristics. While the precise role of anti-U3-RNP antibodies in the development of systemic sclerosis is not completely understood, it is believed that they contribute to immune system dysregulation and tissue fibrosis. These antibodies may trigger immune cell activation and the production of inflammatory and fibrotic cytokines, which can drive the chronic inflammation and fibrosis observed in SSc. Moreover, they may also be involved in the abnormal vascular changes and endothelial dysfunction associated with the disease. Patients with systemic sclerosis who test positive for anti-U3-RNP antibodies often display unique clinical features compared to those with other autoantibodies.

MATERIALS AND METHODS. The research was conducted at the Rheumatology division of Tashkent Clinical Hospital, involving 50 subjects who had been diagnosed with systemic sclerosis based on the 2013 EULAR/EUSTAR criteria. The mean age of the patients was 40.6 ± 3.2 years, with an average disease duration of 6.7 ± 2.1 years. Of the participants, 35 (70%) were women and 15 (30%) were men. Each patient underwent comprehensive clinical, laboratory, instrumental, and immunological assessments.

RESULTS. The majority of patients exhibited a range of skin manifestations, including edema in 18 individuals (36%), induration in 22 individuals (44%), digital ulcers in 10 individuals (20%), and sclerodactyly in 21 individuals (42%). Raynaud's phenomenon was universally present. Dysphagia emerged as the most prevalent internal symptom among the patients evaluated (60%). Anti-fibrillarin antibodies were found in 12 patients (24%) who developed the condition at a young age, with an average age of 34 years. Among these patients, 4 (33.3%) had limited cutaneous systemic sclerosis (lcSSc) and 8 (66.7%) had diffuse cutaneous systemic sclerosis (dcSSc). Comparative analysis revealed that individuals with antibodies targeting fibrillarin tended to have an early disease onset, a progressive systemic sclerosis course, and rapid fibrosis of the lungs and skin.

DISCUSSION. The findings of this study highlight the significant clinical manifestations and serological associations in patients with systemic sclerosis. The presence of skin changes, Raynaud's phenomenon, and internal symptoms such as dysphagia are consistent with the typical features of systemic sclerosis. The identification of anti-fibrillarin antibodies in a subset of patients, particularly those with an early disease onset, suggests a potential role in disease pathogenesis and could serve as a prognostic marker for disease progression. The association between anti-fibrillarin antibodies and a more severe disease course, characterized by rapid fibrosis of the lungs and skin, underscores the importance of identifying specific autoantibodies in systemic sclerosis patients to guide treatment strategies and monitor disease progression.

Conclusion. In conclusion, the detection of anti - fibrillarin antibodies can indicate the progression of fibrosis in the skin and lungs. Early identification of AFA allows for the recognition of patients with a more aggressive disease course, who may benefit from treatments targeting the immune system and fibrosis. This approach has the potential to improve the overall effectiveness of treatment strategies.

References:

1. Абдурахманова, Н. М., & Рахимов, С. С. (2023). Генетические аспекты резистентности к лечению метотрексатом у больных ревматоидным артритом.

2. Надирова, Ю., Жаббаров, О., Бобошарипов, Ф., Умарова, З., Сайдалиев, Р., Кодирова, Ш., ... & Жуманазаров, С. (2023). ОПТИМИЗАЦИЯ КОМБИНИРОВАННОЙ ТЕРАПИИ ПРИ БОЛЕЗНИ АРТЕРИАЛЬНОЙ ГИПЕРТЕНЗИИ БЛОКАТОРОМ КАЛЬЦИЕВЫХ КАНАЛОВ И ИНГИБИТОРОМ АПФ. Solution of social problems in management and economy, 2(2), 181-186.

3. Axmedov, X., & Raximov, S. (2023). Ankilozlovchi spondiloartritda IL-17 ning kasallik kechishidagi axamiyati.

4. Зарипов, С. И., Тураев, И. А., & Рахимов, С. С. (2022). Quality of life in patients with chronic kidney disease receiving program hemodialysis and possible ways of its correction. УЗБЕКСКИЙ МЕДИЦИНСКИЙ ЖУРНАЛ, 3(5).

5. Abdurakhmanova, N., Akhmedov, K., & Rakhimov, S. (2023). Features of the Course of Rheumatoid Arthritis Depending on The C3435t Polymorphism of The Mdr1 Gene. Texas Journal of Multidisciplinary Studies, 21, 18-24.

6. Turaev, S. Z. I., & Rakhimov, S. (2023). ASSESSMENT OF THE QUALITY OF LIFE IN PATIENTS WITH CHRONIC KIDNEY DISEASE IN THE PRACTICE OF HEMODIALYSIS. Journal of Modern Educational Achievements, 6(6), 103-109.

7. Khalmetova, F. I., Akhmedov, K. S., Buranova, S. N., Rakhimova, M. B., Rakhimov, S. S., & Abdurakhimova, L. A. (2023). Immunological Features of Reactive Arthritis of Various Etiologies. Journal of Coastal Life Medicine, 11, 1322-1325.

8. Rakhimova, M. B., Akhmedov, K. S., & Turaev, Y. A. (2021). Endothelial dysfunction as a link in the pathogenesis of ankylosing spondylitis against the background of a new coronavirus infection. ACADEMICIA: An International Multidisciplinary Research Journal, 11(3), 2493-2498.

9. Rakhimova, M., Akhmedov, K., Tagaeva, M., Sadikova, S., Razakova, F., & Khalmetova, F. (2022). Endothelin-1biomarker Features In Patients With Ankylosing Spondylitis After COVID-19. Journal of positive school psychology, 9369-9375.

10. Rakhimova, M. B., Akhmedov, K. S., Rakhimov, S. S., & Zaripov, S. I. (2023). Extrasceletal Manifestations in Patients with Ankylosing Spondylitis. Journal of Coastal Life Medicine, 11, 1315-1321.

11. Rakhimova, M., Kh, A., & Rakhimova, O. (2024). Endothelial dysfunction in patients with ankylosing spondylitis after covid-19.

12. Rakhmatov, A. M., & Jabbarov, A. A. KodirovaSh. A., Jumanazarov SB (2022). CLINICAL MANIFESTATIONS OF GOUTHY NEPHROPATHY. THEORETICAL ASPECTS IN THE FORMATION OF PEDAGOGICAL SCIENCES, 1 (6), 140–141.

13. Jabbarov, O. O., Maksudova, M. H., Mirzayeva, G. P., & Rakhmatov, A. M. (2023). The Relationship of Blood Group with Human Diseases. Web of Semantic: Universal Journal on Innovative Education, 2(3), 331-334.

14. Jumanazarov, S., Jabborov, O., Qodirova, S., & Rahmatov, A. (2022). THE ROLE OF PODOCYTIC DYSFUNCTION IN THE PROGRESSION OF CHRONIC GLOMERULONEPHRITIS.

15. Sultonov, P. I., Umarova, Z. F., Jabbarov, O. O., Khodjanova, S. I., Jumanazarov, S. B., Rahmatov, A. M., & Rahimov, I. S. (2023). EFFECT OF ARTIAGREGANT THERAPY ON KIDNEY FUNCTIONAL RESOURCES IN CHRONIC DISEASE. Theoretical aspects in the formation of pedagogical sciences, 2(5), 137-138.

16. Fayzullaevna, M. G., Otakhanovich, J. O., Tokhirovna, B. N., Mamatovich, R. A., & Bakhadirovich, J. S. (2022). Gout Therapy With Reduced Kidney Function. Central Asian Journal of Medical and Natural Science, 3(6), 198-203.