AUTOIMMUNE CONNECTIVE TISSUE DISEASES

PROTEAN CLINICAL MANIFESTATIONS OF SCLERODERMA-OVERLAP SYNDROMES ASSOCIATED WITH ANTI-PM/SCL AUTOANTIBODIES - A REVIEW THROUGH THE LENS OF 2 UNIQUE CASES.

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Background: Various scleroderma overlap syndromes have been described. Among others, these rare presentations include overlapping scleroderma and polymyositis, scleroderma and rheumatoid arthritis, scleroderma and systemic lupus erythematosus, and scleroderma with vasculitis. Various autoantibodies have been associated with overlap syndromes, including anti-PM/Scl in the setting of scleroderma – polymyositis (sclerodermatomyositis). The pathogenesis of scleroderma, particularly scleroderma overlap syndromes, remains incompletely understood. This is particularly striking in the uncommon case of anti-PM/Scl antibody positivity, where clinical presentations can be strikingly variable with regard to both cutaneous and systemic involvement. As a result, appropriate diagnosis and treatment of such patients may be delayed.

Observation: Herein, we present the cases of 2 patients with positive anti-PM/Scl autoantibodies and dramatically different clinical presentations. The first patient was found to have only subtle cutaneous involvement first noted by an orthopedist despite resulting in a delayed diagnosis despite prior evaluation by rheumatology. The second patient was found to have classic cutaneous manifestations of scleroderma in association with interstitial lung disease and erosive arthritis without myositis. Neither patient demonstrated classic overlapping features of sclerodermatomyositis typically associated with anti-PM/Scl autoantibodies.

Key Message: Dermatologists and clinicians should remain mindful of the possibility of scleroderma-overlap syndromes, which may have positive anti-PM/Scl autoantibodies. Such cases can have a range of clinical manifestations and may pose a diagnostic challenge, as neither scleroderma nor myositis are uniformly present in cases of positive anti-PM/Scl. Appropriate diagnosis is necessary to ensure timely evaluation and treatment of potential cutaneous and systemic complications associated with scleroderma overlap syndromes. Attention to this matter may result in more timely diagnosis, work-up, screening, and treatment of affected patients.