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CORRESPONDENCE

Myocarditis in a patient with anti-OJ and Th/To autoantibody-positive overlap syndrome

Anti-OJ and anti-Th/To antibodies are rare and specific with polymyositis/dermatomyositis and systemic sclerosis (SSc), respectively.[1,2] We report a patient with SSc who subsequently developed polymyositis and fulminant cardiomyopathy, and had both anti-OJ and anti-Th/To antibodies.

A 48-year-old woman with SSc was diagnosed with fulminant myocarditis 6 months prior and developed muscle weakness and dysphagia 3 months prior to her initial visit.

The power of both deltoids, biceps, triceps, brachioradialis, gluteus maximus, and quadriceps muscles were all 3/5. She had a 10-year history of Raynaud's phenomenon and finger-to-forearm skin sclerosis. Muscle cramps, Gottron's sign, mechanic's hands, heliotrope and periungual erythema, nail fold bleeding, and digital ulcer were all absent. The 2013 American College of Rheumatology/European League Against Rheumatism classification criteria for SSc scored 12 points.

Laboratory values were as follows: AST 75 IU/L, ALT 115 IU/L, LDH 303 IU/L, γ-GTP 151 IU/L, CK 1217 IU/L, aldolase 18.7 IU/L (2.1-6.1 IU/L), CK-MB 34 IU/L(<5.0 IU/L), troponin T 0.20 pg/ml (<0.014 pg/ml), CRP, 0.19 mg/dl, ESR 43 mm/h, KL-6 198 IU/L, NT-proBNP 1399 pg/ml, and ANA 320x (nucleolar pattern). Anti-OJ and anti-Th/To antibodies were identified using immunoprecipitation. No autoantibodies such as anti-ARS, MDA-5, and TIF1-γ antibodies, were detected. A skin biopsy of the forearm showed increased collagen fibers and a sweat gland secreting area located at the center of the dermis; these features were suggestive of scleroderma (Figure 1A). Electromyography indicated myogenic changes, and a left bicep muscle biopsy showed lymphocytic infiltration in the perivascular and peri-bronchial areas with necrosis and atrophy of muscle fibers (Figure 1B,C). The lymphocytes that infiltrated the muscle tissue were CD-8-dominant cells (Figure 1D). Chest computed tomography showed no features of interstitial pneumonia. An electrocardiogram showed frequent ventricular extrasystoles. Cardiac ultrasound revealed a reduced ejection fraction of 44%, and there was no pulmonary hypertension. Cardiac contrast-enhanced MRI was not suggestive of myocarditis or myocardial fibrosis. Myocardial biopsy showed no lymphocytic infiltration, myonecrosis, or myocardium fibrosis. Overlap syndrome of limited cutaneous SSc and polymyositis was diagnosed. She was also diagnosed with chronic myocarditis because she had abnormal CK-MB, and troponin T levels, although there were no findings on MRI that were suggestive of a heart pathology.

After bolus doses of glucocorticoids, multiple immunosuppressive agents, and intravenous immunoglobulins were administered, the muscle weakness improved, and CK-MB levels normalized. Troponin T level decreased to 0.031 pg/ml but was not yet normalized 7 years after the commencement of treatment. Since the CK-MB and troponin T levels improved with immunosuppressive therapy, we considered that the chronic myocarditis in the patient might be an autoimmune myocarditis associated with polymyositis or scleroderma.[3,4]

Generally, anti-OJ antibody-positive dermatomyositis/polymyositis is often associated with muscle symptoms and interstitial pneumonia but responds well to steroids.[1] Anti-Th/To antibody-positive scleroderma is associated with severe pulmonary hypertension, interstitial pneumonia, and renal crisis, but is usually not complicated by myositis.[2]

Although the patient had refractory myositis and myocarditis, she did not develop pulmonary hypertension and interstitial pneumonia. This was the first report that both autoantibodies were detected together, and the patient had unique clinical symptoms.

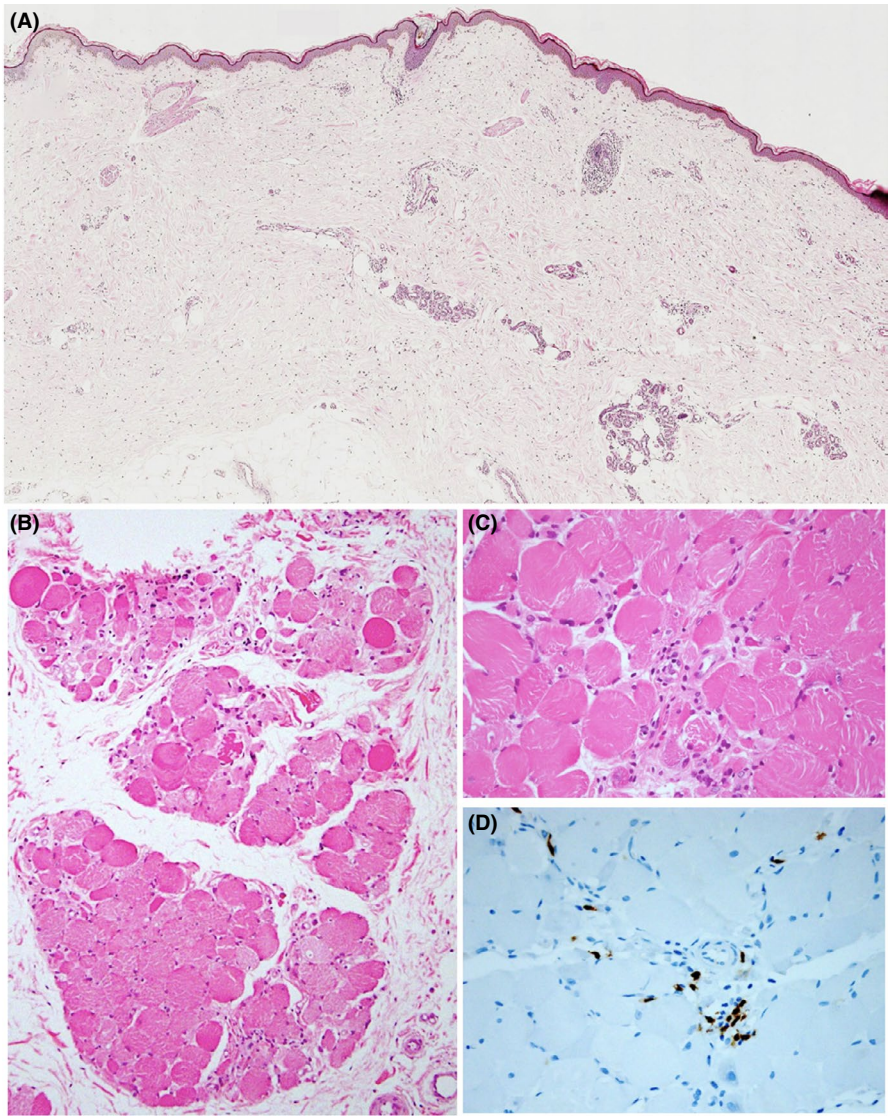

FIGURE 1 Histological findings of the skin samples taken from the forearm and the biceps tendon. (A) Increased collagen fibers and eccrine sweat gland secreting area located in the dermis. (B) Necrosis is seen in the muscle fibers. (C) Lymphocytic infiltration is seen around the muscle fibers and the vessels. (D) CD-8-positive infiltrating lymphocytes [Color figure can be viewed at wileyonlinelibrary.com]

DECLARATION SECTION

Approval of the research protocol: This manuscript is a case report and has not been submitted for approval as a research.
Informed Consent: Informed consent was obtained from the patient.
Registry and the Registration No.: N/A.
Animal Studies: N/A.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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