Refractory myositis in a patient of Sjogren's syndrome having only anti-SS-A (60 kDa) antibody.

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CORRESPONDENCE

Refractory myositis in a patient of Sjögren's syndrome having only anti-SS-A (60 kDa) antibody

Dear Editor,

A 46-year-old Japanese female patient had noticed asymptomatic eruptions on her legs several years ago and visited our hospital in 2015. Physical examination showed purpuras with mild induration and scattered pigmentation (Figure 1A, B). Manual muscle test revealed muscle weakness with a score of 4 on her upper and lower extremities. Laboratory investigation showed an increase in serum creatine kinase (CK; 450 U/L), aldolase (9.0 IU/L), immunoglobulin G (2315 mg/dL), and the presence of cryoglobulin, antinuclear antibody (×640; speckle pattern), and autoantibodies against SS-A/Ro (>1200 U/mL). Other autoantibodies were not detected by enzyme-linked immunosorbent assay. A biopsy specimen from a purpura on her leg showed leukocytoclastic vasculitis in the superficial dermis (Figure 1C, D). Direct immunofluorescence examination demonstrated the deposition of C3 on the vessels in the superficial dermis. (Figure 1E). A deltoid muscle biopsy showed mild myositis (Figure 1F). A biopsy specimen from minor salivary gland on the lip showed more than 50 mononuclear cells infiltration (Figure 1G). Immunoprecipitation assays for RNA (left) using nonradiolabeled K562 cell extracts and for protein (right) using 35S-methionine-labeled K562 cell extracts in our case revealed anti-SS-A (60 kDa) antibody (Figure 1H). We diagnosed her with primary Sjögren's syndrome with myositis and hypergammaglobulinemic purpura. Regarding differential diagnosis, myopathy associated with

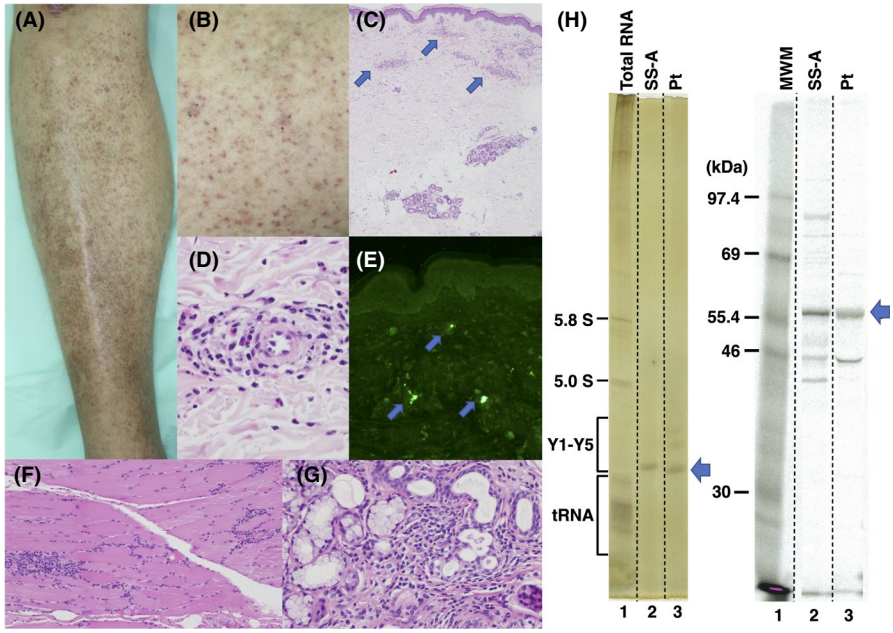

FIGURE 1 (A and B) Clinical presentation at first visit. Physical examination showed a lot of purpura with mild induration and scattered pigmentation. (C and D) Histopathological findings of the cutaneous biopsy specimen from a purpura on her leg showed leukocytoclastic vasculitis in the superficial dermis [hematoxylin–eosin (H&E) staining; original magnification, ×40 (C) and ×400 (D)]. (E) Direct immunofluorescence examination demonstrated the deposition of C3 on the vessels in the superficial dermis. (F) A deltoid muscle biopsy showed mild myositis (H&E staining; original magnification, ×100). (G) A biopsy specimen for minor salivary gland on the lip showed more than 50 mononuclear cells infiltration around the gland (H&E staining; original magnification, ×200). (H) Immunoprecipitation assays for RNA (left) using nonradiolabeled K562 cell extracts and for protein (right) using 35S-methionine-labeled K562 cell extracts in our case revealed anti-SS-A (60 kDa) antibody

cryoglobulinemic vasculitis was included. While it is usually accompanied by arthralgia, peripheral neuropathy, and haematuria,[1] she had no symptoms of them. To improve myositis, oral prednisolone of 20 mg/day was not enough and 45 mg (1 mg/kg) was needed. We reduced the dosage after 4 weeks; then, myositis recurred in 2016. We additionally administered azathioprine (100 mg/day), intravenous immunoglobulin (IVIG; 400 mg/kg/day, 5 days, 3 times) in 2017, tacrolimus (2-3 mg/day) in 2018, or methotrexate (4-6 mg/week) in 2019, which could not improve myositis completely.

Myositis in Sjögren's syndrome usually responds well to small or moderate doses of prednisolone without a recurrence.[2] However, our case needed high doses of prednisolone and various additional treatments for recurrences. Autoantibodies are divided into myositis-specific antibody (MSA) such as antimelanoma differentiation-associated gene 5 (MDA5) and myositis-associated antibody (MAA) such as SS-B/La and SS-A/Ro. Whereas the concurrent presence of more than one MSA in one patient is rare, MAAs are frequently observed with MSAs. Two different targets of proteins, Ro52 and Ro60, were identified regarding SS-A. In general adult population having anti-SS-A antibodies, the profile of Ro52-Ro60+ was found in 24%.[3] However, the profile of Ro52-Ro60+ seems to be extremely rare in Japanese patients with polymyositis/dermatomyositis (PM/DM),[4] and Sjögren's syndrome with myositis having only anti-SS-A antibody is also rare.[2] Anti-SS-A antibody could induce PM/DM through activating interferon-α production.[5] Recent studies reported that anti-SS-A/Ro52 antibody was a risk factor for relapse in PM/DM[6] and associated with poor prognosis in anti-MDA5-antibody-positive PM/DM.[7] Anti-SS-A/Ro60 antibody can function as a Y RNA-free protein,[8] but the details are still unclear. These studies mentioned not about anti-SS-A/Ro60, but only about anti-SS-A/Ro52. We suggest a possibility that not only anti-SS-A/Ro52 but also anti-SS-A/Ro60 may contribute to the resistance in the same way.

KEYWORDS

azathioprine, intravenous immunoglobulin, methotrexate, myositis, Sjögren's syndrome, tacrolimus

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None.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

DECLARATION SECTION

Approval of the research protocol: N/A.
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Registry and the Registration No. of the study/trial: N/A.
Animal Studies: N/A.

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