

Myopathy Associated With Antibodies to Signal Recognition Particle

Disease Progression and Neurological Outcome

Shigeaki Suzuki, MD, PhD; Yukiko K. Hayashi, MD, PhD; Masataka Kuwana, MD, PhD; Rie Tsuburaya, MD, PhD; Norihiro Suzuki, MD, PhD; Ichizo Nishino, MD, PhD

Objective: To characterize the clinical course of myopathy associated with antibodies to signal recognition particle (SRP), or anti-SRP myopathy.

Design: Case series.

Setting: Keio University Hospitals and National Institute of Neuroscience, National Center of Neurology and Psychiatry, Tokyo, Japan.

Patients: We reviewed clinical features of 27 patients with anti-SRP myopathy and analyzed disease progression and neurological outcome.

Main Outcome Measures: Anti-SRP antibodies in se-

rum were detected by RNA immunoprecipitation assay using extracts of K562 cells.

Results: Of the 27 patients, 5 (19%) showed chronic progressive muscle weakness as well as atrophy of limbs and trunk muscles from a younger age with more severe neurological outcomes compared with the other 22 patients (81%) with the subacute form.

Conclusion: A subset of patients with anti-SRP myopathy can show a chronic progressive form associated with severe clinical deficits.

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AUTOANTIBODIES AGAINST signal recognition particle (SRP) were first found in the serum of a patient with polymyositis and were listed as myositis-specific antibodies.¹ Myopathy associated with antibodies to SRP (anti-SRP myopathy) has recently been regarded as an immune-mediated necrotizing myopathy based on histological findings and has been clinically characterized by severe muscle weakness, marked elevation of serum creatine kinase (CK) levels, and poor response to corticosteroid therapy.²⁻⁷ These observations were gathered mainly from patients with a clinical diagnosis of inflammatory myopathies. However, the clinical spectrum of anti-SRP myopathy may be broader.

The rapid progression of weakness is a characteristic clinical feature of anti-SRP myopathy.²⁻⁷ The mean interval from its onset to diagnosis is 3 to 4 months, and clinical symptoms are usually progressive for 5 to 6 months.³⁻⁵ In contrast, Dimitri et al¹⁸ first described a 31-year-old man in whom weakness progressed for more than 3 years. Before the anti-SRP anti-

body was detected, he was diagnosed as having limb-girdle muscular atrophy. We also described a 32-year-old man with childhood-onset myopathy whose diagnosis alternated between inflammatory myopathy and muscular dystrophy for 21 years.⁹ These results suggested that patients with anti-SRP myopathy can show chronic progression indistinguishable from muscular dystrophy. Herein, we analyzed the disease course and neurological outcomes in patients with anti-SRP myopathy.

METHODS

We chose 27 patients with myopathy with the anti-SRP antibody, including 10 previously reported cases.^{9,10} The diagnosis of anti-SRP myopathy was based on clinical, electrophysiological, histopathological, and serological findings. Muscle weakness was assessed by manual muscle strength (Medical Research Council scale grade), and severe weakness was defined as grade 3 or lower. Muscle biopsy was performed in all 27 patients and showed fiber size variation as well as fiber necrosis and regeneration with or without lymphocyte infiltration. No patients had taken statins.

Author Affiliations: Department of Neurology (Drs S. Suzuki and N. Suzuki) and Division of Rheumatology, Department of Internal Medicine (Dr Kuwana), Keio University School of Medicine, and Department of Neuromuscular Research, National Institute of Neuroscience, National Center of Neurology and Psychiatry (Drs Hayashi, Tsuburaya, and Nishino), Tokyo, Japan.

Anti-SRP antibodies were detected by RNA immunoprecipitation assay using extracts of K562 cells as previously described.¹¹ Briefly, 10 μ L of serum was mixed with 2 mg of Protein A Sepharose CL-4B (Pharmacia Biotech AB) in 500 μ L of immunoprecipitation buffer (10mM TRIS hydrochloride, pH 8.0, 500mM sodium chloride, 0.1% Nonidet P40) and incubated for 2 hours. After washing 3 times with immunoprecipitation buffer, antigen-bound Sepharose beads were mixed with 100 μ L of K562 cell extract (6×10^6 cell equivalents per sample) for 2 hours, and 30 μ L of 3M sodium acetate, 30 μ L of 10% sodium dodecyl sulfate, and 300 μ L of phenol:chloroform:isoamyl alcohol (50:50:1, containing 0.1% 8-hydroxyquinoline) were added to extract bound RNA. After ethanol precipitation, the RNA was resolved by using a 7M urea–8% polyacrylamide gel, and the gel was silver stained (Bio-Rad). Immunoprecipitated RNA located in the 7SL-RNA lesion was regarded as anti-SRP antibody. Other myositis-specific and myositis-associated autoantibodies were also detected by the RNA immunoprecipitation assay.

Neurological outcomes were assessed using the modified Rankin Scale (mRS).¹² This scale was principally used for evaluating function of patients with stroke; however, it was also applied to patients with myositis.¹³ Neurological outcomes were divided into 3 groups: recovered, mild deficit, and severe deficit. Patients who responded optimally to the treatment and returned to their jobs (mRS score of 0–1) were defined as recovered. Patients who responded partially to treatment and resumed most activities of daily living (mRS score of 2–3) were defined as having a mild deficit. Patients who showed re-worsening muscle weakness or re-elevation of serum CK levels after the treatment were also included in this group. Patients who responded minimally to the treatment and required support in daily activities (mRS score of 4) were defined as having a severe deficit.

This study was approved by the institutional review boards at Keio University and the National Center of Neurology and Psychiatry. Statistical analyses were performed using Stat-View version 5.0 statistical software (SAS Institute, Inc).

RESULTS

Figure 1 shows the distribution of periods between disease onset and the first examination. We divided 27 patients with anti-SRP myopathy into 2 subtypes (subacute and chronic forms) based on the clinical course. Of the 27 patients with anti-SRP myopathy in our study, 5 (19%) were considered to have the chronic form. The patients' demographic and clinical features are compared between those with the subacute and chronic forms (**Table 1**). Disease onset occurred at a younger age in those with the chronic form than in those with the subacute form (mean age, 15.4 vs 52.4 years, respectively; $P < .001$). No patients with the chronic form had a clear clinical history of antecedent infection, whereas 3 patients (14%) with the subacute form had antecedent infection. Despite a previous report,⁵ seasonal occurrence was not clear in our series. Disease progression of the subacute form was usually rapid, and the mean duration between disease onset and the first examination was 3.1 months. In particular, 3 patients showed rapid disease progression in 2 to 3 weeks. In contrast, patients with the chronic form showed significantly slower progression, and the mean duration between disease onset and the first examination was 10.2 months ($P = .001$).

In our series, asymmetrical muscle involvement was seen in 2 patients, whereas the other 25 patients showed proximal-dominant symmetrical limb muscle weak-

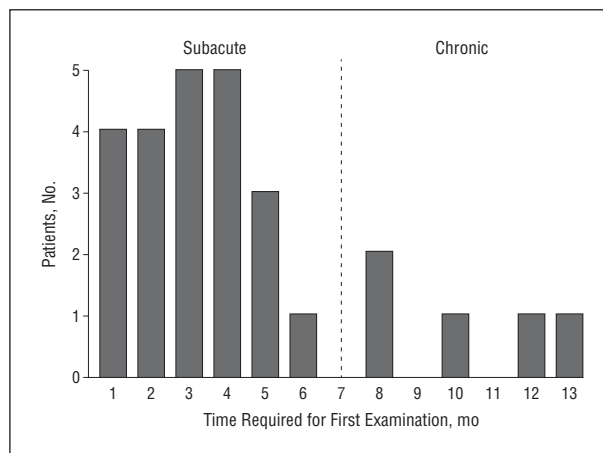


Figure 1. Period between disease onset and the first examination in 27 patients with anti-signal recognition particle myopathy. They were divided into 22 patients with the subacute form and 5 patients with the chronic form based on the clinical course.

ness. Lower limbs were more severely affected than upper limbs. All 5 patients with the chronic form and about half of the patients with the subacute form showed severe muscle weakness and atrophy at the first examination. Several reports emphasized that dysphagia, but not dysarthria, was observed at a high frequency in 43% to 75% of patients with anti-SRP myopathy.^{3,5,7} In our series, 7 patients (26%) had dysphagia and 3 (11%) reported it as the initial symptom. Previous reports also showed a high frequency of cardiac involvement,^{2,5} while only 1 patient in our series had arrhythmias, which did not require treatment. Respiratory muscle involvement was detected in 3 patients. Myalgia was noted in 9 patients (36%) and tended to precede muscle weakness. Extramuscular manifestations were observed only in patients with the subacute form. Skin rash and interstitial lung disease, which were clinically suggestive of dermatomyositis, were observed in 2 and 4 patients, respectively. Serum CK levels were markedly elevated to more than 1000 IU/L (to convert to microkatal per liter, multiply by 0.0167) in all 27 patients; however, there was no difference between the subacute and chronic forms. Other autoantibodies were found in 6 patients with the subacute form, including Ro/SSA (3 patients), Th/To (1 patient), ribosome (1 patient), and U1RNP (1 patient).

All 27 patients were treated with oral prednisolone (1 mg/kg/d). Half of the patients were treated with additional immunosuppressive agents, including methotrexate ($n = 5$), azathioprine ($n = 4$), tacrolimus ($n = 2$), cyclophosphamide ($n = 1$), and cyclosporine ($n = 1$), or with intravenous immunoglobulin ($n = 6$). Although some patients required 2 to 3 months to respond to treatment, the patients with anti-SRP myopathy did not always respond poorly. The combination of oral prednisolone and intravenous immunoglobulin appears to be most effective for patients with the subacute form as the initial treatment. The neurological outcomes showed that 10 patients (45%) with the subacute form recovered. In contrast, all 5 patients with the chronic form had more severe neurological outcomes compared with the 22 patients with the subacute form ($P = .008$) (**Figure 2**).

Table 1. Comparison of Clinical Features Between Subacute and Chronic Forms of Anti-Signal Recognition Particle Myopathy

Clinical Feature	Patients, No. (%)		P Value
	Subacute (n = 22)	Chronic (n = 5)	
Age at onset, mean (range), y	52.4 (14-82)	15.4 (5-32)	<.001 ^a
Female	12 (55)	3 (60)	.78 ^b
Antecedent infection	3 (14)	0	.93 ^b
Time required for first examination, mean (range), mo	3.1 (1-6)	10.2 (8-13)	.001 ^a
Muscle weakness			
Arms < legs	16 (73)	3 (60)	.98 ^b
Arms > legs	6 (27)	2 (40)	.98 ^b
Severe involvement	11 (50)	5 (100)	.12 ^b
Laterality	1 (5)	1 (20)	.80 ^b
Facial muscle involvement	1 (5)	1 (20)	.80 ^b
Bulbar sign	6 (27)	1 (20)	.81 ^b
Cardiac involvement	1 (5)	0	.80 ^b
Respiratory failure	3 (14)	1 (20)	.73 ^b
Neck weakness	9 (41)	4 (80)	.27 ^b
Muscle atrophy	10 (45)	5 (100)	.08 ^b
Myalgia	8 (36)	1 (20)	.86 ^b
Extramuscular involvement			
Fever	4 (18)	0	.73 ^b
Skin rash	2 (9)	0	.80 ^b
Arthritis	1 (5)	0	.80 ^b
Raynaud phenomenon	1 (5)	0	.80 ^b
Interstitial lung disease	4 (18)	0	.73 ^b
Associated disorder			
Cancer	1 (9)	0	.80 ^b
Rheumatic disorder	1 (9)	0	.80 ^b
Serum creatine kinase, mean (range), IU/L	6101 (1149-15 585)	4190 (2465-5725)	.08 ^a

SI conversion factor: To convert serum creatine kinase to microkatal per liter, multiply by 0.0167.

^aStatistical analysis by *t* test.

^bStatistical analysis by χ^2 test.

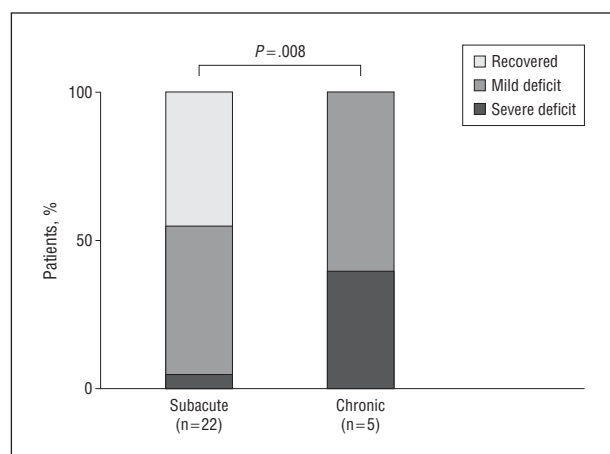


Figure 2. Neurological outcomes were assessed using the modified Rankin Scale¹² with some modifications and were compared between subacute and chronic forms of anti-signal recognition particle myopathy. The neurological outcomes were divided into recovered, mild deficit, and severe deficit. Differences between the groups were analyzed with the Mann-Whitney test. Five patients with the chronic form showed more severe outcomes than 22 patients with the subacute form ($P = .008$).

Detailed clinical features of 5 patients with the chronic form are summarized in **Table 2**. All patients had severe muscle weakness and marked atrophy in all 4 limbs and the trunk. Two patients (patients 2 and 5) noticed arm muscle weakness as the initial symptom. Importantly, scapular winging was noted in 2 patients (pa-

tients 2 and 3) at the first examination and was suspected to involve facioscapulohumeral muscular dystrophy. The serum CK level was decreased after treatment in patients with the chronic form, but muscle weakness gradually progressed and recovery of muscle strength was delayed. Three patients (patients 1, 2, and 3) became unable to walk independently, and 1 (patient 3) required mechanical ventilation. Because muscle biopsies were not suggestive of inflammatory myopathy, 1 patient (patient 3) was treated for only 3 months and 2 (patients 1 and 2) were treated after the detection of anti-SRP antibody. Of these younger patients, 2 (patients 2 and 3) became severely disabled, whereas the other 2 (patients 4 and 5) were treated soon after the muscle biopsy and responded partially to treatment.

COMMENT

There are 2 methods for detecting anti-SRP antibodies: the RNA immunoprecipitation assay we used and an immunoassay using the signal peptide-binding 54-kDa subunit of SRP (SRP54) as the antigen. Because SRP54 is regarded as the main antibody target, the immunoassay using SRP54 is easily conducted and the antibody level is also available.^{1,2,14} However, epitopes of anti-SRP antibodies may also be located in other subunits of SRP proteins or 7SL-RNA.^{7,15} In contrast, RNA immunoprecipitation assay, the standard method for detection of

Table 2. Clinical Features of 5 Patients With the Chronic Type of Anti-Signal Recognition Particle Myopathy

Feature	Patient No.				
	1 ^a	2 ^a	3 ^a	4	5
Sex	F	F	M	F	M
Age at onset	5 y 9 mo	9 y 8 mo	10 y 2 mo	20 y 10 mo	32 y 9 mo
Initial symptoms	Frequent falls	Difficulty raising arm	Difficulty running fast	Difficulty climbing stairs	Difficulty raising his child
Weakness and atrophy	Proximal limbs (U < L); trunk	Proximal limbs (U > L); trunk; scapular winging; left dominant; myalgia	Proximal limbs (U < L); trunk; scapular winging; facial, bulbar; respiratory	Proximal limbs (U < L); trunk	Proximal limbs (U > L); trunk; sternocleidomastoideus
Serum creatine kinase, IU/L	4629	2467	4180	3951	5725
Muscle images	Atrophy in proximal limbs and trunk	Left-dominant atrophy and edematous change in proximal limbs and trunk	Atrophy and edematous change in proximal limbs and trunk	Atrophy and edematous change in proximal limbs and trunk	Atrophy and edematous change in proximal limbs and trunk
Age at muscle biopsy	6 y 5 mo	10 y 4 mo	11 y 3 mo, 16 y 6 mo	21 y 8 mo	33 y 9 mo
Muscle biopsy					
Variation in fiber size	Scattered	Scattered	Marked	Marked	Marked
Fiber necrosis and regeneration	Moderate	Marked	Marked	Scattered	Marked
Lymphocyte infiltration	None	None	None	None	Perivascular
Endomysial fibrosis	Minimal	Mild	Marked	Minimal	Mild
Age at anti-SRP antibody detection	7 y 4 mo	10 y 9 mo	32 y 6 mo	21 y 10 mo	34 y 3 mo
Age at treatment start	7 y 4 mo	10 y 9 mo	11 y 6 mo	21 y 8 mo	33 y 9 mo
Treatment	PSL, MTX, MPR	PSL, MTX, IVCY, AZA, tacrolimus	PSL (3 mo)	PSL, MPR	PSL, MTX, IVIg, tacrolimus
Age at final follow-up	9 y 3 mo	13 y 10 mo	34 y 8 mo	23 y 3 mo	35 y 6 mo
Response and neurological outcome	Partial response; progression for 2 y; relapse; MMT grade 4; Gowers sign	Minimal response; progression for 2 y; MMT grade 2-3; walking 20 m; difficulty in holding dishes	No response; progression for 3 y; recovered from mechanical ventilation; MMT grade 2-3; wheelchair use	Partial response; progression for 1 y; MMT grade 4	Partial response; progression for 1.5 y; weakness recovered; relapse

Abbreviations: AZA, azathioprine; IVCY, intravenous cyclophosphamide; IVIg, intravenous immunoglobulin; L, lower; MMT, manual muscle strength; MPR, high-dose methylprednisolone sodium succinate; MTX, methotrexate; PSL, prednisolone; SRP, signal recognition particle; U, upper.

SI conversion factor: To convert serum creatine kinase to microkatal per liter, multiply by 0.0167.

^aThese patients were previously described.^{9,10}

anti-SRP antibodies, has advantages in sensitivity and specificity.^{1,2,4,6,9,11} The RNA immunoprecipitation assay can recognize the conformational epitopes of SRP, although the titer of antibodies is not available. Many studies showed that anti-SRP antibodies were principally specific to myositis or necrotizing myopathy except in a few patients with systemic sclerosis or rheumatoid arthritis.^{1,2,4,6,9,11} In regard to myopathies, we demonstrated that anti-SRP antibody was not detected in patients with various types of muscular dystrophy, and it was useful for the differential diagnosis of myopathies using RNA immunoprecipitation assay.⁹

Anti-SRP myopathy can show a wider variety of clinical symptoms than was previously considered. When weakness progresses rapidly, within 2 to 3 weeks, with extremely high serum CK levels (>10 000 IU/L), acute rhabdomyolysis should be differentiated.⁸ When patients experience progressive weakness within 2 to 6 months²⁻⁷ accompanied by interstitial lung disease, skin rash, or associated rheumatic disorders, polymyositis or dermatomyositis should be considered. Because skin rash is observed in approximately 10% of cases of anti-SRP

myopathy in the present and previous studies,⁵ anti-SRP antibodies may be also detected in patients clinically diagnosed as having dermatomyositis. In fact, Hama-guchi et al¹⁶ reported that anti-SRP antibodies were detected in 7 of 376 patients (2%) with dermatomyositis using a similar detection method.

In our series, 5 of 27 patients with anti-SRP myopathy (19%) showed chronic progressive muscle involvement. The mean age at onset in these 5 patients was significantly younger than that of the patients with the subacute form, and patients with the chronic form showed severe weakness and atrophy in limbs and trunk muscles as well as poorer outcomes. It was speculated that the poor outcome may be partially ascribed to the delay of the first examination or anti-SRP antibodies detection. Importantly, these clinical features may indicate the possibility of muscular dystrophy rather than inflammatory myopathy,⁸⁻¹⁰ although the disease progression was faster than occurs in muscular dystrophy. In fact, facioscapulohumeral muscular dystrophy was initially suspected in 2 patients owing to prominent shoulder-girdle weakness.^{9,10}

It is well known that anti-SRP myopathy is usually resistant to treatment, resulting in severe disability.^{2-4,6,7} However, our observation suggested that patients with the subacute form had relatively good neurological outcomes. Early diagnosis by screening for anti-SRP antibodies is important for choosing intensive immunotherapy, which might contribute to better outcomes. In this regard, Hengstman et al⁵ reported that the response to treatment for patients with anti-SRP myopathy did not differ significantly from that of myositis without anti-SRP antibodies. They reported that 75% of patients with anti-SRP myopathy could walk without any assistance after treatment. The severe outcomes of anti-SRP myopathy described in the previous studies may be attributable partly to results for patients with the chronic form. Rituximab therapy is potentially effective for patients with the chronic form.⁷ Based on these findings, it may be useful to divide patients by disease progression to predict the neurological outcome.

An apparent question about the relationship between anti-SRP antibodies and muscle involvement is whether the anti-SRP antibodies themselves have any pathogenic effect against muscle. This hypothesis may be supported by several lines of data: (1) anti-SRP antibodies purified from patients' serum samples can inhibit the in vitro translocation of secretory proteins into endoplasmic reticulum¹⁷; (2) the levels of anti-SRP54 autoantibodies are closely associated with the levels of myolysis¹⁴; and (3) the removal of anti-SRP antibodies by plasma exchange improves muscle strength.^{14,18} Nevertheless, the causal relationship between anti-SRP antibodies and muscle involvement is still not established, and further experiments such as passive transfer to animals are necessary to elucidate the pathogenesis of anti-SRP antibodies.

In conclusion, anti-SRP myopathy can show quite variable disease progression and neurological outcomes.

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Correspondence: Shigeaki Suzuki, MD, PhD, Department of Neurology, Keio University School of Medicine, 35 Shinanomachi, Shinjuku-ku, Tokyo 160-8582, Japan (sgsuzuki@z3.keio.jp).

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