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【 CASE REPORT 】

Anti-contactin 1 Antibody-associated Membranous Nephropathy in Chronic Inflammatory Demyelinating Polyneuropathy with Several Autoantibodies

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Abstract:
A 50-year-old man diagnosed with anti-contactin 1 (CNTN1) antibody-associated chronic inflammatory demyelinating polyneuropathy (CIDP) was referred to our department for the evaluation of proteinuria. A kidney biopsy revealed membranous nephropathy (MN). Immunohistochemistry for CNTN1 revealed positive granular staining along the glomerular basement membrane, confirming anti-CNTN1 antibody-associated MN. Immunofluorescence showed a full-house pattern, and several autoantibodies, such as anti-nuclear antibody, anti-double-strand DNA antibody, and anti-cardiolipin antibody, were detected in the patient's serum. Although limited autoantibodies have been investigated in some of the reported cases, a variety of autoantibodies might be produced in anti-CNTN1 antibody-associated CIDP, accompanied by MN.

Key words: contactin 1, chronic inflammatory demyelinating polyneuropathy, membranous nephropathy, systemic lupus erythematosus

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Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an immune-mediated neuropathy characterized by symmetrical involvement and proximal/distal muscle weakness. Autoantibodies to the node of Ranvier of myelin, such as neurofascin-155 (NF-155) (1, 2), contactin-1 (CNTN1) (3-5), contactin-associated protein 1 (CASPR 1) (6), and neurofascin isoforms (NF140/186) (7), have been reported to be involved in the pathogenesis of some patients with CIDP. In addition to CIDP, an anti-CNTN1 antibody has recently been suggested to induce membranous nephropathy (MN) (8).

A number of histological characteristics of MN in anti-CNTN1 antibody-associated CIDP have been reported, including granular CNTN1 antigen staining along the glomerular basement membrane (GBM) with IgG4 subclass predominance; furthermore, clinical characteristics have been reported, with a predominance observed in middle to elderly men (8-10).

We herein report a case of MN in anti-CNTN1 antibody-associated CIDP whose serum included several autoantibodies, such as anti-nuclear antibody (ANA), anti-double-strand DNA (dsDNA) antibody, and anti-cardiolipin antibody.

Case Report

A 50-year-old man was admitted to our department for the evaluation of continuous proteinuria. Proteinuria had been noted by a medical examination four years prior and had persisted until referral. He noticed weakness and numb-

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Table 1. Laboratory Date on Admission.

Parameter	Value	Parameter	Value
Complete blood cell count		Serology	
White blood cells, /μL	5,510	C3, mg/dL	84
Hemoglobin, g/dL	12.7	C4, mg/dL	14
Platelets, $\times 10^4$/μL	21.7	CH50, IU/mL	44
MCV	88	IgG, mg/dL	2,082
Lymphocyte, %	30	IgA, mg/dL	376
Lymphocyte, /μL	1,653	IgM, mg/dL	121
		Antinuclear antibody	1:160
Blood chemistry			Homogeneous, speckled
Lactate dehydrogenase, U/L	235	Anti-double stranded DNA antibody, IU/mL	36
Aspartate aminotransferase, IU/L	16	Anti Sm antibody	Negative
Alanine aminotransferase, IU/L	15	C1q, μg/mL	<1.5
Alkaline phosphatase, IU/L	264	Anti-β2-glycoprotein I antibody	<1.0
Total protein, g/dL	7.2	Anti-cardiolipin antibody, U/mL	13
Albumin, g/dL	3.6	Lupus anticoagulant	1
Triglycerides, mg/dL	219	Anti SS-A/Ro antibody	Negative
Total cholesterol, mg/dL	172	Anti SS-B/La antibody	Negative
Low-density lipoprotein cholesterol, mg/dL	110	Anti RNP antibody	Negative
Serum urea nitrogen, mg/dL	11.8	Direct antiglobulin test	Negative
Creatinine, mg/dL	0.64	HBS-Ag, IU/mL	<0.03
eGFR, mL/min/1.73 m^2	103	HCV-Ab	Negative
Uric acid, mg/dL	4.7		
Sodium, mEq/L	142	**Coagulation system**	
Potassium, mEq/L	3.8	PT-INR	0.94
Chloride, mEq/L	108	APTT, %	82
Calcium, mg/dL	8.9	D-dimer, μg/mL	1.4
Inorganic phosphate, mg/dL	4.1		
C-reactive protein, mg/dL	0.16	**Urinalysis**	
Ferritin, ng/mL	134	pH	5.5
Hemoglobin A1c, %	5	Protein, g/g・creat	0.79
Blood glucose, mg/dL	123	Red blood cells, /μL by flow cytometry	4.1 (normal range: 10.0)
		creatinine, mg/dL	101.6
		β2-microglobulin, μg/L	208
		α1-microglobulin, mg/L	12.9
		N-acetyl-β-D-glucosaminidase, U/L	8.4

C3: complement component 3, C4: complement component 4, CH50: complement activities, Ig: immunoglobulin, HBs-Ag: hepatitis B surface antigen, HCV-Ab: hepatitis C virus antibody

ness in his upper and lower limbs one year before admission. He was admitted to the Department of Neurology due to difficulty walking and frequent tumbles and was diagnosed with CIDP. Anti-NF-155 antibody, one of the most common autoantibodies found in patients with CIDP, was not detected in his serum or cerebrospinal fluid, whereas anti-CNTN1 antibody, a newly reported autoantibody in CIDP patients, was present in both his serum and cerebrospinal fluid. Therefore, the diagnosis of anti-CNTN1 antibody-associated CIDP was made.

On admission, his blood pressure was 116/89 mmHg, his pulse was regular at 90 beats/min, and his body temperature was 36.9°C. A physical examination revealed weakness of the upper and lower limb muscles, hyperesthesia, and deep sensory disorder peripheral to both the wrist and ankle joints. Laboratory data are shown in Table 1. As ANA, anti-dsDNA, and anti-cardiolipin antibodies were detected in the patient's serum, a kidney biopsy was performed to differentiate lupus nephritis, MN in anti-CNTN1 antibody-associated CIDP, and other renal diseases.

Light microscopy showed 35 glomeruli with spike formation and stippling on GBM (Fig. 1A), resulting in the diagnosis of MN. In individual glomeruli, a slight increase in the mesangial cells and matrix and double contours of GBM were observed in a focal segmental manner (Fig. 1B). Masson's trichrome staining revealed tubular atrophy and interstitial fibrosis in approximately 40% of the tubulointerstitial area. Immunohistochemistry for IgG showed enhanced, granular staining along the GBM (Fig. 1C). Immunofluorescence showed a full-house pattern (IgG, IgA, IgM, C3, C4, and C1q were all positive) along the GBM and mesangial areas (Fig. 1D-I). Electron microscopy revealed electron-dense deposits (EDDs) with some lucent areas in the subepithelial, intramembranous, and para-mesangial areas (Fig. 1J).

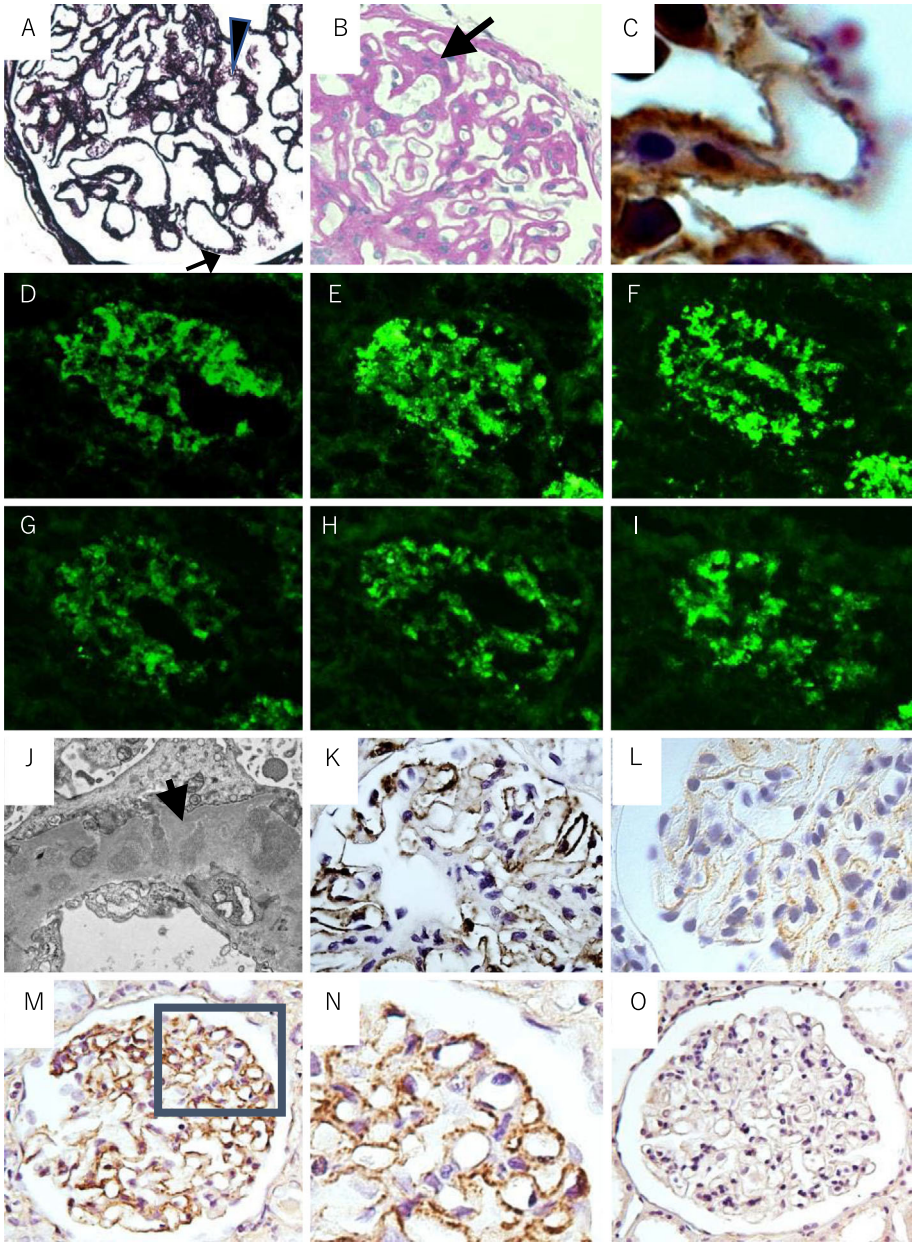

Figure 1. **Photographs of the glomeruli in the kidney. (A) Periodic acid methenamine silver staining shows spike (arrow) and bubbling formations (arrowhead). Original magnification: ×400. (B) Periodic acid-Schiff staining of the renal biopsy specimen shows slightly increased mesangial cells and matrix (arrow). Original magnification: ×400. Enhanced granular expression of CNTN1 at a higher magnification in a fragment of Fig. 1M is depicted in Fig. 1N. (C) Immunohistochemistry for IgG shows enhanced, granular staining of IgG along the GBM. Original magnification: ×1,000. Mouse anti-IgG (1:2,000, #05-4200; Thermo Fisher Scientific, Waltham, USA) was used. Immunofluorescence shows granular staining for IgG (D), IgA (E), IgM (F), C3 (G), C4 (H), and C1q (I) along the GBM. (J) Electron microscopy reveals subepithelial and intramembranous deposits in the GBM. Electron lucent deposits are also recognized in some EDDs (arrow). Original magnification: ×5,000. Immunohistochemistry for IgG1 (K) and IgG4 (L) shows granular staining along glomerular capillary walls. Original magnification: ×1,000. Anti-human IgG1 rabbit monoclonal antibody (1:1,000, #RM117; RevMAb Biosciences, Burlingame, USA) and mouse anti-IgG4 (1:500, #HP6025; Binding Site, Birmingham, UK) were used. (M-O) Immunohistochemistry for CNTN1. Renal specimens from the present case showed positive, granular staining of CNTN1 along the GBM. Original magnification: ×400 (M) or ×1,000 (N). A sample from a PLA2R-associated MN patient was used as a negative control (O). Original magnification: ×400. For immunohistochemistry of CNTN1, antigen retrieval was performed by Proteolytic-Induced Epitope Retrieval (HistoReveal®; Abcam, Cambridge, UK). Goat anti-contactin-1 (1:200, #AF904; R&D Systems, Minneapolis, USA) was used.**

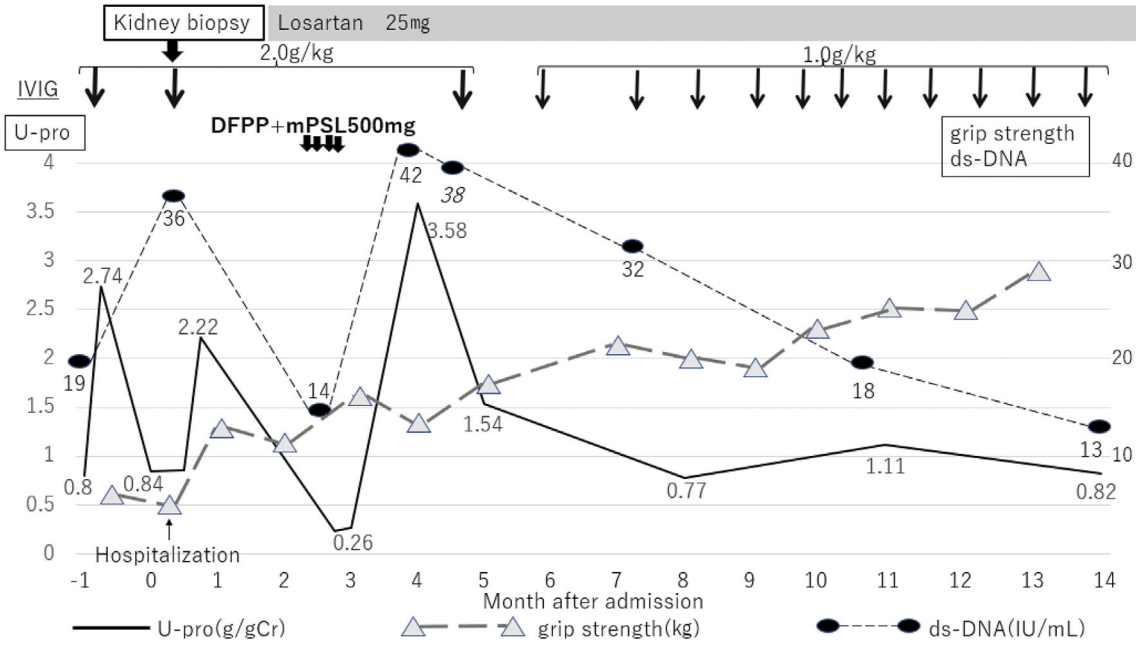

Figure 2. Clinical course after admission. The levels of grip strength, U-pro, and anti-ds DNA antibody are shown with lines. IVIG: intravenous immunoglobulin, DFPP: double filtration plasmapheresis, mPSL: methylprednisolone, U-pro: urine protein (urinary protein-creatinine ratio), ds-DNA: anti-double-strand DNA antibody

Tubuloreticular inclusion in glomerular cells and fingerprint structure in EDDs were not found. Immunohistochemistry for IgG1 (Fig. 1K) or IgG4 (Fig. 1L) showed granular staining along the GBM. Immunohistochemistry for CNTN1 revealed positive, granular staining along the GBM, confirming MN in anti-CNTN1 antibody-associated CIDP (Fig. 1M-O). Phospholipase A2 receptor (PLA2R), thrombospondin type 1 domain containing 7A, neural epidermal growth factor-like 1 and exostosin 1/exostosin 2 (EXT1/2) staining were negative (data not shown). Screening for secondary causes of MN, which included testing for anti-hepatitis B surface antigen, obtaining information on prescribed medications, and malignancies evaluated with plane computed tomography from the neck to the pelvis, did not detect any other cause of secondary MN.

The clinical course is shown in Fig. 2. The patient was treated with intravenous immunoglobulin and double filtration plasmapheresis followed by methylprednisolone 500 mg single dose (4 times) as standard therapies for anti-CNTN1 antibody-associated CIDP (8). After starting the treatment, muscle weakness and sensory disturbance gradually improved. The urinary protein level also gradually decreased to approximately 1 g/gCr at 14 months after the treatment. The anti-ds-DNA antibody level also gradually decreased in parallel with the improvement in muscle strength and the decrease in urinary protein during follow-up. Serum albumin and creatinine levels were stable. The patient had not displayed any signs indicative of autoimmune diseases at the time of printing (14 months after the treatment).

Discussion

We reported a case of anti-CNTN1 antibody-associated MN and CIDP with several autoantibodies. In the current case, the existence of anti-CNTN1 antibody in the patient's serum and the positive staining of CNTN1 along the GBM led to the diagnosis of MN in anti-CNTN1 antibody-associated CIDP. However, the presence of anti-dsDNA antibody, the immunostaining pattern in the mesangial areas and the full-house pattern of immunofluorescence findings suggest the possibility of the coexistence of anti-CNTN1 antibody-associated MN and systemic lupus erythematosus (SLE)/lupus nephritis type V.

As MN with anti-CNTN1 antibody-associated CIDP has recently been reported as a new entity of MN, only 11 cases have been reported to date, and the characteristics of MN in anti-CNTN1 antibody-associated CIDP have not been fully established. According to the summary of reported cases, as listed in Table 2, the age at presentation was approximately 60-80 years old. Ten (90.9%) patients were men. Most patients had nephrotic syndrome, with a preserved kidney function. All cases evaluated with IgG subclass staining in renal tissues have been reported to have IgG4 predominance. With regard to the treatment, corticosteroids, intravenous immunoglobulin (IVIG), rituximab, and plasma exchange were often administered. Urinary protein levels gradually decreased after treatment in parallel with the therapeutic effect on CIDP symptoms in many cases, but there were few reports on the long-term follow-up; thus, the recurrence rate and long-term renal prognosis were unclear. Regarding the

Table 2. Characteristics of Patients with MN in Anti-CNTN1 Antibody-associated CIDP.

Age (years)/ sex	Sequence of manifesta-tions	U-pro (g/day)	sAlb (g/ dL)	sCr (mg/ dL)	Treat-ment	Response of MN/ follow up period	Test for auto-antibodies in serum	Immunofluorescence	EM stage	Immunohis-tochemistry	Reference
75/Male	Concurrent or MN→CIDP	7.2	2.8	0.98	IVIG/ CS	PR	Negative*	IgG4(3+) >IgG1・IgG2(2+) >IgG3(+) C3(+), CNTN1(+) granular staining along capillary walls	1	CNTN1(+), PLA2R(-), THSD7A(-)	#9
71/Male	Concurrent or MN→CIDP	5.7	2.8	1.73	RTX	Death	LAC	IgG4(3+) >IgG3(2+) >IgG2(1+) >IgG1(-) C3(+), CNTN1(+) granular staining along capillary walls	1	PLA2R(-), THSD7A(-)	#9
58/Male	Concurrent or MN→CIDP	4.5	2.7	0.6	IVIG	PR	LAC, anti-β2-glycoprotein I antibody	IgG4・IgG1・IgG3(3+) >IgG2(-) ・C3(+), CNTN1(+) granular staining along capillary walls	2	PLA2R(-), THSD7A(-)	#9
80/Male	Concurrent or MN→CIDP	1.8	2.6	1.13	IVIG/ RTX/ PE	Death	Negative*	IgG4・C3(+), CNTN1(+) granular staining along capillary walls	1	PLA2R(-), THSD7A(-)	#9
66/Male	Concurrent or MN→CIDP	2.2	2.5	0.93	IVIG/ RTX/ CS	CR	Negative*	IgG4(3+)>IgG1・IgG2・IgG3(-) C3(+)	1	PLA2R(-), THSD7A(-)	#9
73/ Female	ND	3.6	2.9	0.8	CS/ RTX	Improved gradually	Negative for ANA, ENA, anti-dsDNA antibody, LAC, anti-cardiolipin antibody, ANCA, anti-PLA2R antibody	IgG・κ・λ・CNTN1(+), IgA・C1q(-), granular staining along capillary walls	1	PLA2R(-), THSD7A(-)	#11
61/Male	CIDP→MN	7	2.1	N/A	IVIG/ RTX	CR/3 years	ND	IgG4(+)>IgG1 C3(+), CNTN1(+) granular staining along capillary walls	1	ND	#12
57/Male	MN→CIDP	7.8	3	N/A	RTX/ CS/ IVIG	Improved	Anti-PLA2R antibody	IgG4・IgG1(2+) >IgG2(+) >IgG3(-) IgM・C3・κ・λ(+), IgA・C1q(-) granular staining along capillary walls	3	PLA2R(+), THSD7A(-)	#8
78/Male	Concurrent	4.1	0.7	N/A	CS/ IVIG	Im-proved/18 months	Anti-SS-A/Ro antibody	IgG4(+) >IgG1・IgG2・IgG3(-) C3(-) granular staining along capillary walls	2	PLA2R(±), THSD7A(-)	#10
75/Male	CIDP→MN	5.4	2.2	0.54	CS/ PE/ RTX	Improved → death	Negative for anti-PLA2R antibody	IgG4・C3(+), C1q(-), granular staining along capillary walls	ND	PLA2R(-)	#13
43/Male	CIDP→MN	22	1.5	N/A	IVIG/ CS/ CyA	PR/18 months	Negative for anti-PLA2R antibody	IgG(+)	2	CNTN1(+), PLA2R(-)	#14
50/Male	MN→CIDP	0.84	3.6	0.65	IVIG/ PE	Improved gradually/ 7 months	ANA (homoge-neous, speckled) anti-dsDNA antibody anti cardiolipin antibody	Full house pattern along capillary walls and mesangial areas	3-4	CNTN1(+), PLA2R(-), THS-D7A(-), IgG1(+), IgG4(+)	Present case

MN: membranous nephropathy, CNTN1: contactin 1, CIDP: chronic inflammatory demyelinating polyneuropathy, U-pro: urinary protein, sAlb: serum albumin, sCr: serum creatinine level, CS: corticosteroid, CyA: cyclosporin, PE: plasma exchange, RTX: rituximab, IVIG: intravenous immunoglobulin, PR: partial remission, CR: complete remission, EM: electron microscopy, PLA2R: M-type phospholipase A2 receptor, THSD7A: thrombospondin domain-containing 7A, ANA: antinuclear antibody, ENA: extractable nuclear antigen, dsDNA: double-stranded DNA, LAC: lupus anticoagulant, ANCA: anti-neutrophil cytoplasmic antibody, ND: not described

*, detailed information on autoantibodies was not described.

onset pattern, CIDP-preceding, MN-preceding, and concurrent cases have been reported (8-14).

In the current case, the patient was a man, and his kidney function was preserved. The degree of proteinuria was relatively mild compared to the reported cases. Histologically, IgG1 and IgG4 were positive along the GBM, but IgG1 seemed to be predominant. Regarding the treatment response, IVIG and plasma exchange were effective for both CIDP and MN. Urinary protein maintained incomplete remission type I, and no signs of relapse were observed at 14 months after the treatment. Regarding the onset, the present case was considered to be MN-preceding because the urinary protein had been positive three years before the diagnosis of CIDP. However, the patient had a long history of hypertension, and a renal biopsy revealed changes in nephrosclerosis. Therefore, the preceding proteinuria might have been due to atherosclerotic nephropathy.

Interestingly, several autoantibodies, such as ANA, anti-dsDNA antibody, and anti-cardiolipin antibody, were detected in the serum from the present case. There were two possibilities concerning the presence of autoantibodies, such as ANA, anti-dsDNA antibody, and anti-cardiolipin antibody. First, the present case had SLE, and anti-CNTN1 antibody might be produced in some SLE patients. Anti-dsDNA antibody is often present in SLE patients and is a marker of disease activity (15). Based on the European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) diagnostic criteria (16), the current case could be diagnosed as SLE if the renal lesion was considered lupus nephritis. A variety of autoantibodies are reportedly present in SLE patients (17-19). Nervous system-directed antibodies were detected in 13.2% (24/174) of patients with SLE, and 56.5% (13/24) of them had neurological symptoms of CIDP (17). In addition, the prevalence of anti-neutrophil cytoplasmic antibody or anti-GBM antibody has been reported in 29.1% (16/55 patients) or 8.9% (14/157 patients) of SLE patients, respectively (18, 19). These autoantibodies might contribute to the development of a variety of antibody-related symptoms in SLE patients. However, our case did not show any characteristic clinical findings of SLE, hypocomplementemia or specific pathological findings of lupus nephritis, such as fingerprint structure in EDDs or tubuloreticular inclusion in glomerular cells. Furthermore, immunohistochemistry for EXT1/2 was negative, and approximately 30% was detected along the capillary walls in lupus nephritis pure type V (20-23). However, the presence of anti-dsDNA antibody, the immunostaining pattern in the mesangial areas, and the full-house pattern of immunofluorescence findings suggest the presence of SLE/lupus nephritis type V (15, 24). Second, unclassified autoimmune diseases that produce various autoantibodies, such as SLE, might produce anti-CNTN1 antibodies, ANAs, anti-dsDNA antibodies, and anti-cardiolipin antibodies, of which only anti-CNTN1 antibodies cause organ damage, such as CIDP and MN. Although information on examined autoantibodies is limited to a few reports (9, 12-14), a variety of autoantibodies, such as anti-PLA2R antibody, lupus anticoagulant, anti-β2-glycoprotein I antibody, and anti-SS-A/Ro antibody, have been detected in sera from MN patients with anti-CNTN1 antibody-associated CIDP (Table 2) (8-10). Several reports regarding MN in anti-CNTN1 antibody-associated CIDP have described the examination of ANA, anti-DNA antibodies, and anti-cardiolipin antibodies (9, 10), but the presence of these autoantibodies has not yet been reported. The accumulation of further cases is necessary to clarify which kinds of autoantibodies are produced and how often they exist in patients with MN in anti-CNTN1 antibody-associated CIDP.

Several limitations associated with the present study warrant mention. First, anti-CNTN1 antibody in the serum after treatment was not measured; therefore, we could not evaluate whether or not the disease condition improved in parallel with the antibody titer. Second, we were able to evaluate the IgG subclasses of IgG1 and IgG4 by immunohistochemistry but not IgG2 or IgG3 due to the lack of available antibodies for IgG2 or IgG3. In addition, frozen kidney tissue was not sufficient to evaluate IgG subclasses. Therefore, we were unable to confirm which IgG subclass was predominant.

We herein reported a case of anti-CNTN1 antibody-associated MN and CIDP with several autoantibodies. It was speculated that a variety of autoantibodies might be produced in MNs in anti-CNTN1 antibody-associated CIDP. A further investigation is needed to clarify what kinds of autoantibodies are produced and how often they exist in patients with MN in anti-CNTN1 antibody-associated CIDP.

The authors state that they have no Conflict of Interest (COI).

Ryutaro Shida, Takamasa Iwakura, and Naro Ohashi contributed equally to this work.

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