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Case Report

Anti-SRP positive immune-mediated necrotic myopathy with two ulnar fingers on the right hand: a case report

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Abstract: A 52-year-old female presented with progressive limb weakness, dysphagia, and dyspnea, with poor prognosis. Onset of symptoms was weakness in the two ulnar fingers of the right hand, with no other accompanying symptoms. The diagnosis was "ulnar nerve damage", according to the local hospital, and was considered appropriate. The symptoms progressed laterally to the contralateral limb, not reported previously. Results for anti-SRP (signal recognition particle) antibody were positive. Therapy was administered with a combination of methylprednisolone, IVIG, methotrexate, and tacrolimus, with poor prognosis.

Keywords: Immune, necrotizing myopathy, signal recognition particle, type of onset

Introduction

IMNM (immune-mediated necrotic myopathy), a type of IIMs (idiopathic-inflammatory myopathy), was first reported in 1969. Previously, it was considered a subgroup of PM (polymyositis). Now, however, it is known as an independent disease [1]. It varies from other classifications of IIMs [2-4], including PM (polymyositis), DM (dermatomyositis), sIBM (sporadic inclusion body myositis), ASS (anti-synthetase syndrome), and OS (overlap syndrome). Clinical features of acute or progressive onset, symmetrical proximal limb weakness, CK (creatine kinase) elevation, electromyography myogenic damage, and histopathology may be characterized by muscle fiber necrosis and regeneration, as well as lymphocyte infiltration of individual muscle cells [5]. The present study reports a case of patient with distal weakness onset of two ulnar fingers of the right upper limb, gradually progressing to the proximal extremity of the same side and the contralateral limb.

Case report

A 52-year-old female presented with progressive limb weakness, dysphagia, and dyspnea, with a poor prognosis.

History and examination

A 45-day history before admission demonstrated the onset of symptoms, including weakness of the two ulnar fingers of the right hand. This condition was diagnosed as "ulnar nerve damage" and was not considered serious. Weakness was progressive (**Table 1**) in the right forearm, upper arm, and right lower limbs, distal and proximal. Tests found that levels of muscle enzymes were > 9000 IU/L. To further clarify the diagnosis, several hospitals were used for referrals. The symptoms continued to progress to the left upper and lower limbs. Subsequently, the patient was admitted to PLA (Chinese People's Liberation Army) 309 Hospital because of progressive limb weakness.

Furthermore, no evidence of muscle pain or numbness, cutaneous rash, ocular symptoms, oral ulceration, arthralgias, or nail changes were observed. Past medical history was noted with respect to insomnia (half a year), hypertension (half a year), and hyperlipidemia (10 days), but did not consider the statins.

Clinical examinations revealed right upper and lower limb proximal and distal muscle weakness, with a clinical power grade of 3/5. The left limb was weak, with a clinical power grade

Table 1. Disease evolution schedule for diagnosis and treatment

Past medical history: insomnia, hypertension, hyperlipidemia, denying the history of statins, and family history. Current history: IMNM, insomnia								
Time	Muscle weakness part (Muscle strength grade)	Laboratory Biomarkers						Medications
		ALT	AST	CK	LDH	CK-MB	TnI	
2017-05-10	Two ulnar fingers on the right hand 4	Cervical MRI, vertebral disc (C5/6, C6/7) is slightly protruding						No
2017-05-25	Right forearm-upper arm, 4			> 9000				No
2017-05-27	Right lower limb distal, 4	Head MRI, normal						No
2017-05-29	Right lower limb proximal, 4	223	386	9780	1367	914		No
2017-06-15	Left forearm-upper arm, 4	Chest CT, tiny nodules in both lungs						No
2017-06-18	Left lower limb distal, 4	Abdominal and pelvic CT, left adrenal gland tiny nodules, adenoma possible						No
2017-06-25	Left lower limb proximal, 4	254	498.5	11232	1422	> 298	0.25	No
2017-06-27	Right hemiplegia, 3	240	467	9487	1239	> 298	0.21	No
2017-06-29	Limbs Muscle atrophy, weight loss	258	417	8482	1293	> 298	0.18	No
2017-07-03	Neck extensor, 1	277	494	10139	1420	> 298	0.27	No
2017-07-06	Intercostal muscle and diaphragm, 4	253	432	9261	1283	> 298	0.27	No
2017-07-11	Unable to wash the face and walk	244	377	7718	1282	> 298	0.27	Methylprednisolone 1 g × 3 d
2017-07-14	Bilateral limbs muscle, 3	ECG, sinus rhythm, abnormal Q wave, ST-T change. Cardiac ultrasound, lower left ventricular diastolic function. Tumor marker: NSE 32.35 ng/mL (0-16.3), $\beta_2$microglobulin 3.18 μg/mL (9-89), iron protein 111.5 ng/mL (0-3.04).						Methylprednisolone 0.5 g × 3 d
2017-07-17	Bilateral limbs muscle, 2	Whole body PET-CT, total muscular metabolism increase, Lumbar puncture, 200 mmH_2O, microprotein 0.32 g/L, Anti-nuclear antibody 1:320 cytoplasmic granule type, SRP antibody was positive in immunoblotting method.						Methylprednisolone 240 mg × 3 d
2017-07-20	Bilateral limbs muscle, 1	EMG[1], thigh MRI[2], Muscle biopsy[3], IHC[4] ([1,2,3,4]see text)						Methylprednisolone 120 mg × 3 d
2017-07-24	Dysphagia, choking	235	192	2815	822	> 298	0.36	Iv-Ig 0.4 g/kg × 5 days Prednisolone 30 mg × 3 days
2017-07-31	Bilateral limbs, 1	135	138	1151	569	> 298	0.51	Prednisolone 7.5 mg × 3 days
2017-08-05	Intercostal muscle and diaphragm, 4	103	101	766	655	207	0.62	Prednisolone withdrawal. Methotrexate 10 mg × 1/week
2017-08-07	Neck extensor, 2	87.2	80.2	681.8	519.3	212.43	0.57	Methotrexate 10 mg × 1/week
2017-09-08	Limbs proximal, 3; limbs distal, 4; neck extensor, 4	Discharged with an improved health condition						Methotrexate 10 mg × 1/week
2017-09-23	Limbs proximal, 2. limbs distal, 4	52.9	164.7	5774	788			Relapse
2017-10-01	Neck extensor, 3 Dysphagia, choking, dyspnea							Iv-Ig 0.4 g/kg × 5 days Tacrolimus 1 mg × 1/day
2017-10-05	Limbs proximal,2; limbs distal, 3	Blood concentration of FK506, 2.3 ng/mL						Tacrolimus 2 mg × 1/day
2017-10-16	Limbs proximal, 2; limbs distal, 3	64.5	190	5442	870	FK506, 4.2 ng/mL		Tacrolimus 2 mg × 2/day
2017-10-20	Neck extensor, 3 Dysphagia, choking, dyspnea	61.3	168.2	4618	871	PCO_2, 55 mmHg PO_2 50 mmHg		Non-invasive ventilator

2017-10-27	Upper limbs proximal, 1; distal 3 Lower limbs proximal, 0; distal 1; dysphagia, choking, dyspnea	Plasma exchange qod × 3 Methylprednisolone 250 mg Reduced by half every 3 days
2017-10-31	Upper limbs proximal, 2; distal 4; lower limbs proximal, 0; distal 1; dysphagia, choking, dyspnea	Prednisone 10 mg × 1/day Methotrexate 10 mg × 1/week

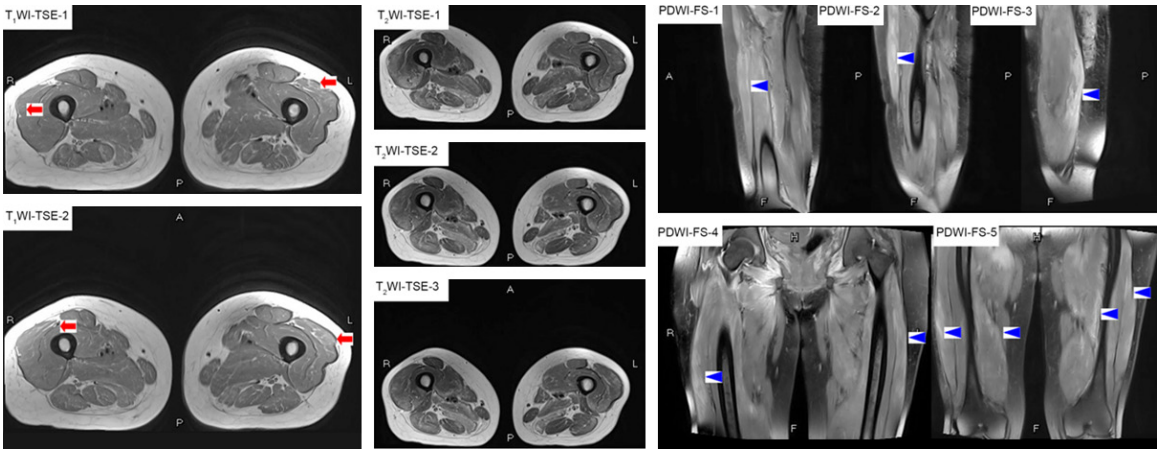

Figure 1. MRI of the bilateral thigh. (T_1WI-TSE 1-2), vastus lateralis muscle shows abnormal signal intensity on the T_1WI-TSE image, suggesting mild atrophy and fat infiltration (arrows). (T_2WI-TSE 1-3), semimembranosus and vastus lateralis muscle shows high signal intensity on the T_2WI-TSE image, suggesting muscle edema. (PDWI-FS 1-5) semimembranosus and vastus lateralis muscle shows high signal intensity on the PDWI-FS image, suggesting muscle edema (arrowheads).

of 4/5. The neck extensor was also weak, with a clinical power grade of 1/5 (**Table 1**).

Initial laboratory investigations

Whole-body PET-CT (Positron Emission Tomography-Computed Tomography) showed increased muscle metabolism in the field of vision.

Needle electromyography showed extremely relaxed pretibial muscle with appropriate spontaneous potential (spontaneous fibrillation potential +, spontaneous positive sharp wave ++), right biceps (spontaneous fibrillation potential +, spontaneous positive sharp wave +), quadriceps muscle, and double small amounts of spontaneous positive sharp wave (+), with no beam fibrillation potential. In the case of contraction, the mean time limit of the right quadriceps, biceps, and tibialis anterior CMAP (complex muscle action potential) was narrowed. In severe contraction, the right brachial biceps and tibial anterior myocardial disturbance phase, left quadriceps mixed phase, and right quadriceps muscle disturbance phase were observed. Nerve conduction velocity, F wave, and H reflex were normal. Serious damage was observed to muscle fibers. The thigh MRI showed a bilateral symmetric long T_2 signal, the vastus lateralis muscle, and semi-membrane muscle (**Figure 1**).

Tumor full set (female) was as follows: NSE (neuron-specific enolase) 32.35 ng/mL, 2 microglobulin 3.18 g/mL, and iron protein 111.5 ng/mL. T lymphocyte subgroup detection revealed a lymphocyte absolute value of 1260 (800-4000)/μL, CD_4^+/CD_8^+ 1.91 (1.0-2.16)%, cytotoxic/suppressor T-cell absolute value of 262 (330-910)/μL, positive for T-cell absolute value 6 (0-80)/μL, 61.77 (58.0-86.0)% of the total T-cell percentage, and total T-cells for the absolute value of 778 (1000-2470).

Anti-nuclear antibody (1:320) was used for immunodetection of SRP (positive), while other myositis-related and specific antibodies were negative.

Enzyme histochemistry results of the muscle biopsy (**Figure 2**) were assessed by HE (hematoxylin-eosin). Mild connective tissue hyperplasia, muscle small vessel wall thickening, and luminal stenosis were detected. No abnormal material deposition and no inflammatory cell infiltration around the blood vessels were observed. Muscle fibers were within the size range, the atrophy of muscle fibers that were primarily circular or angular showed scattered distribution, and visible compensatory hypertrophy of the muscle fibers was seen, along with visible degeneration of muscle fibers, necrosis, devouring, visible regeneration of the muscle fibers, no inflammatory cell infiltration, and suspicious inflammatory cells invasion of a non-necrotic muscle fibers. NADH-TR (reduced form of nicotinamide-adenine dinucleotide) and SDH (succinodehydrogenase) staining demonstrated the uneven distribution of oxidase in a

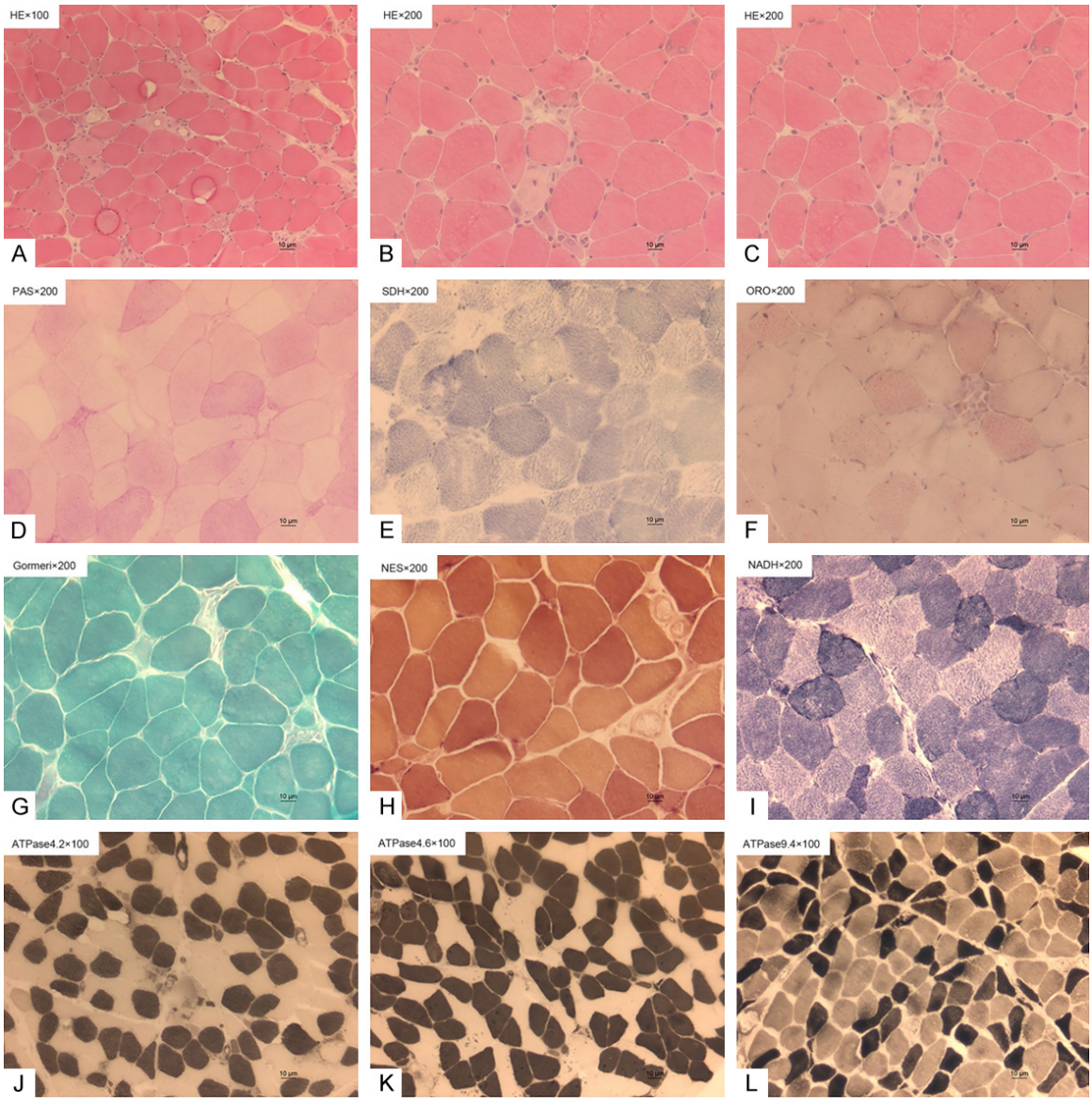

Figure 2. Enzyme histochemistry of muscle biopsy on frozen section. (A) H&E, 10 ×. (B, C) H&E, 20 ×. (G) Gomeri, 20 ×, connective tissue mild hyperplasia, small vessel wall thickening, luminal stenosis, no abnormal material deposition, and no inflammatory cell infiltration around blood vessels. Atrophy muscle fibers were primarily circular or angular with scattered distribution, visible compensatory hypertrophy of the muscle fibers, visible degeneration of the muscle fibers, necrosis, devouring, regeneration, and no inflammatory cell infiltration. (D) PAS 20 × showed no obvious abnormality. (E) SDH 20 ×, the distribution of oxidase in a few muscle fibers was unevenly distributed. (I) NADH-TR 20 × and (E) SDH 20 ×, the distribution of oxidase in a few muscle fibers was non-uniform. Thus, RBF was observed. (H) NSE 20 ×, part of the angular atrophic muscle fibers was intensely stained. (F) ORO 20 × and (J-L) ATPase 10 × (pH 4.2, 4.6, 9.4) showed a checkerboard pattern distribution and atrophy of two types of muscle fibers.

few muscle fibers. NSE (neuronspecific enolase) staining strongly dyed the angular atrophic muscle fibers. ORO (oil red O) staining and PAS (para-aminosalicylic acid) staining did not show any obvious abnormalities. ATPase (adenosine triphosphatase) staining (pH 4.2, 4.6, 9.4) showed a checkerboard pattern and atrophy of two types of muscle fibers.

IHC (immunohistochemistry) results of muscle biopsies (**Figure 3**) included: CD8 (cytotoxic T lymphocyte) (-), CD4 (T-Cell surface specific molecules) (+), CD68 (surface molecules of macrophages) (++), CD45RO (later B cells) (+), CD20 (differentiation antigen of B cells) (-), and MHC-I (major histocompatibility complex I) were commonly detected partially in muscle bundles

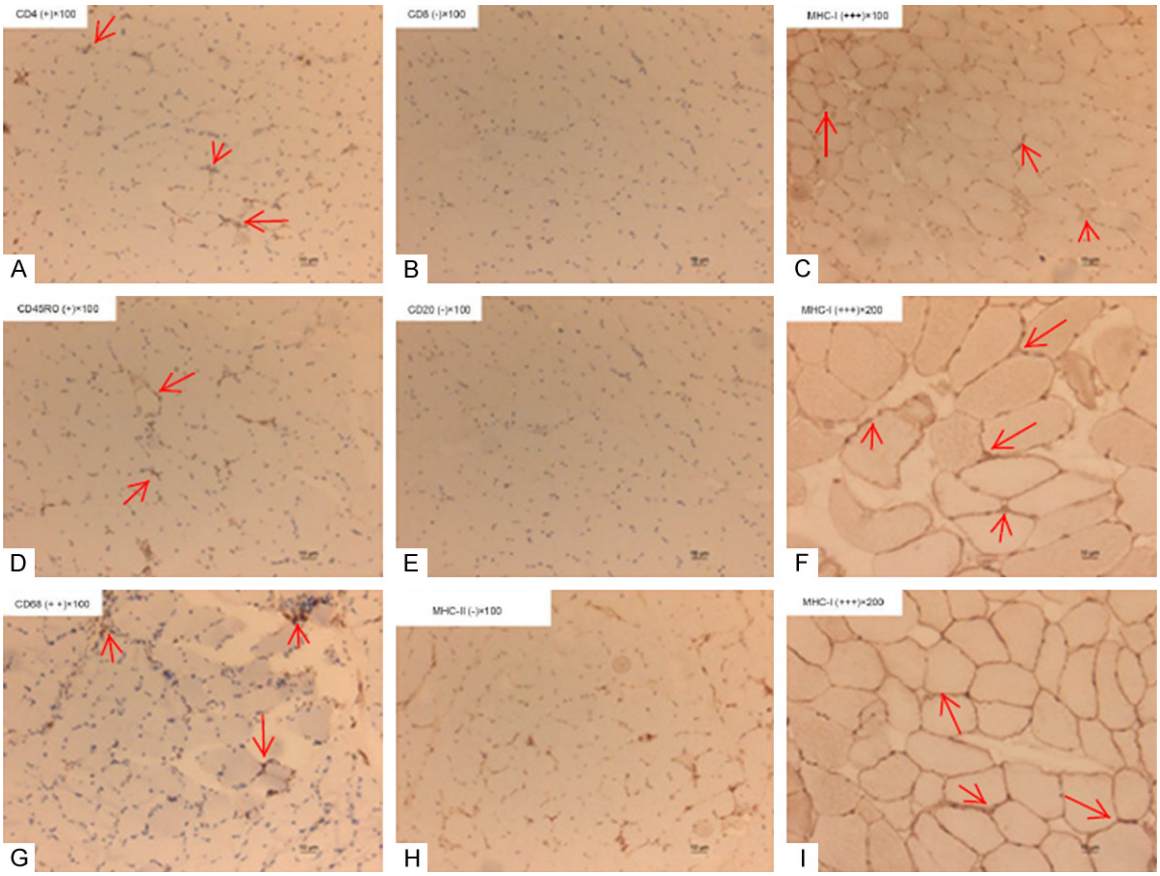

Figure 3. Immunohistochemistry results of muscle biopsies on frozen section. A small amount of inflammatory cell infiltration is seen in the endometrium, including the macrophages of CD68 positive (++), T lymphocytes of CD4 positive (+) (A), CD45RO positive (+) (B), and CD68 positive (++) (C), but lacking the T lymphocytes of CD8 negative (-) (D) and CD20 negative (-) (E). On myofibrous membrane and in partial myofibrous cytoplasm, MHC-II antigen negative (-) (F) and MHC-I antigen strong positive (+++) (G-I) widely.

in sarcolemma, while MHC-II (major histocompatibility complex II) did not demonstrate any distinct abnormalities.

Working diagnosis and management

Middle-aged women often display chronic and progressive limb weakness, normal tendon reflexes, and negative pathologies. CK (creatine kinase) was progressively elevated and the anti-SRP antibody was positive, while the electromyogram showed myogenic damage. Imaging findings showed a slightly longer T_2 signal for the muscle of the double thigh. The muscle biopsy showed active necrotic myopathy.

Investigation and hospital course

On July 11, 2017, the injection dosage was 1 g × 3 days, 0.5 g × 6 days, 40 mg × 3 days, and 120 mg × 3 days. This was halved for 3 days after oral administration. Immunoglobulin was administered 0.4 g/kg/d × 5 days from July 24, 2017. Moreover, 10 mg methotrexate was administered 1 time/week on August 05, 2017. A total of 75 days later, both upper limbs in the patient's distal muscle showed a strength grade of 4 and proximal 3. They exhibited dyspnea and dysphagia, while choking disappeared. The patient was discharged on September 8, 2017.

Further management

On Sep 23, 2017, the disease relapsed (**Table 1**). Symptoms continued to worsen. Dyspnea required noninvasive ventilator support. The patients was discharged on October 24, 2017.

Follow-up

In another hospital, the patient underwent three plasma exchanges and received methyl-

prednisolone (250 mg halved every 3 days). Presently, prednisolone (10 mg daily) and methotrexate (10 mg, weekly) are used as maintenance therapy. The dyspnea does not currently need ventilator support, while the dysphagia needs a liquid diet through a gastric tube. The upper limbs cannot be utilized for combing her hair but she can grab objects. The lower limbs cannot be raised from the bed.

Discussion

There are five main risk factors for IMNM: ① Malignant tumors [6]; ② Virus infections, such as Hepatitis C [7] and AIDS (Acquired Immune Deficiency Syndrome) [8]; ③ Connective tissue diseases, such as scleroderma [9]; ④ Myositis-specific antibodies (anti-SRP antibody and anti-HMGCR(Antibody of trihydroxytrimethyl coenzyme A reductase) antibody) [10]; and ⑤ Drugs (taking statins) [11] and poisoning (ethanol). In the current case, no positive findings were detected in the detailed screening of tumors. Moreover, all relevant indicators of virus infections were negative. There were no signs of connective tissue disease or history of administering statin anti-SRP antibodies. Therefore, this case was characterized as anti-SRP antibody-positive IMNM.

IMNM is similar to clinical muscular dystrophy regarding serum muscle enzymes, electromyography, and pathology of muscle biopsies [10, 12, 13]. Differential diagnosis heavily depends on the detection of specific antibodies, considered diagnostic markers of IMNM [14, 15]. Two kinds of specific antibodies in IMNM are anti-SRP and anti-HMGCR antibodies, with positive rates of 39% and 26%, respectively. The two antibodies are mutually exclusive. Hence, double-positive patients have been rare [10] Approximately 30-40% of IMNM have shown no known antibodies. One study reported that the positive rate of anti-SRP antibody in PM and DM patients was 5-10% [16], while the positive rate was 18% in non-sIBM IIM [17]. Thus, accurate diagnosis was based on the combination of antibody detection and muscle biopsy pathology. In 1986, Reeves et al. first found anti-SRP antibodies in the serum of patients with PM. The titer was correlated with serum CK levels, which indirectly reflected the link between the structure and pathology [18]. In the current case, the detection of anti-SRP antibody was not quantitative but qualitative. Therefore, it could not be used to monitor disease activity. However, anti-SRP IMNM was diagnosed satisfactorily.

A previous study reported [19] the clinical characteristics of anti-SRP antibody-positive IMNM. Of the enrolled 68 patients, 52 showed that legs were dominant, 1 patient showed distal dominance, 43 patients showed severe involvement, and 12 patients showed laterality. Moreover, 48 patients showed neck weakness, 46 patients showed dysphagia, 8 patients showed respiratory insufficiency, 3 patients showed facial involvement, 1 patient showed cardiac involvement, and 46 patients showed muscle atrophy. Extra-muscular symptoms, cancer, and rheumatic disease were rare. In the present case, the distal dominant onset of two ulnar fingers on the right hand had not been reported. The weakness progressed and lateralized into the right forearm, right upper arm, distal of right lower limbs, proximal of right lower limbs, left forearm, left upper arm, distal of left lower limbs, and proximal of left lower limbs. Neck-dysphagia-respiratory insufficiency was also noted. Proximal leg muscle atrophy was obvious. Involvement of the organs occurred sequentially, which formed the specific characteristics of this case. However, during recovery, the distal limbs recovered first.

Muscle enzymes were widely found in the skeletal muscle, myocardium, and brain tissue. Skeletal muscle enzymes included CK, LDH (lactate dehydrogenase), ALT (alanine aminotransferase), and AST (glutamate transaminase). Of these, CK was extremely sensitive to skeletal muscle damage. ALT and AST were elevated. The liver lesion was not considered clinically, indicating skeletal muscle damage. Levels of CK-MB (CK isozyme) and TnI (troponin I) were elevated and the myocardial lesion was considered clinically. Elevated serum CK suggested existing muscle damage before the change in muscle strength. Before and after treatment, the change in muscle strength and muscle enzymes was often not parallel. Thus, improving muscle strength was clinically essential. Some IIMs subtypes, such as IBM and DM, although active in inflammatory infiltration in muscle tissue, showed that CK can be normal or mildly elevated. However, after extensive muscle atrophy, CK release was reduced [20].

The anti-SRP antibody-positive IMNM demonstrated that CK was significantly higher than in other myositis or myopathy, which was useful for diagnosis. Reportedly, CK is 6589 ± 4233 IU/L [21]. In this case, muscle enzymes markedly increased for muscle cell necrosis. After glucocorticoid treatment, inflammation and muscle enzymes decreased. Levels of CK/LDH/ALT/AST axis were similar. CK-MB and TnI tests ruled out myocardial damage.

In 2004, the European Neuromuscular Disease Center used MRIs as the diagnostic criteria for IIMs, especially IMNM. However, the diagnostic criteria of the IMACS (International Myositis Assessment and Clinical Studies) group did not include thigh MRIs from the study population. T_2WI-FS (T2-weighted image-fat suppression) or STIR (short-tau inversion recovery) showed a high signal in skeletal or fascial muscle, indicating active inflammatory edema. Sensitivity was 89-100% higher than the muscle biopsy (66%) [22]. T_1WI (T1-weighted image) showed a high signal in skeletal muscle, indicating adipose infiltration or adipose replacement. The shape and size of the low signal reflected the muscle's compensatory hypertrophy or atrophy. Simultaneously, distribution characteristics of muscle inflammatory edema, fat infiltration/substitution, and compensatory hypertrophy/atrophy in different muscles were observed. These characteristics may assist in guiding the muscle biopsy site and shortening the range of differential diagnosis. DM muscle fascial edema was prominent. The anterior chamber and distal legs muscles of sIBM were predominant. Muscular dystrophy was accompanied by selective muscle atrophy/aliphatic edema [17]. Lateral muscular edema of the anti-SRP-positive IMNM was obvious. Vastus intermediates were relatively conservative and thigh muscle edema, atrophy, and fat infiltration were noted [23]. Edema displayed a low signal on T_1WI, while a high signal was observed on T_2WI and PDWI-FS (proton density weighted imaging-fat suppression). Fat infiltration showed a high signal on T_1WI, T_2WI, and PDWI, but low on PDWI-FS. In the current case, vastus lateralis muscle mild atrophy and fat infiltration/substitution were observed on T_1WI. Combining T_2WI with PDWI-FS, irregular patchy high signals were displayed, suggesting active inflammatory edema. In addition, compared to muscular dystrophy, active inflammatory edema was remarkable in IMNM.

Intriguingly, no spontaneous potential (fibrillation potential and positive sharp wave) was detected in myogenic damage electromyography. Classical electromyography of IMNM is characterized by spontaneous fibrillation potential, positive sharp wave, and insertion of excitatory activity in most IMNM patients [20, 24]. Fibrillation potential occurred occasionally in normal muscles. However, more than two locations in a muscle was considered a pathological scenario, usually representing the neurological deficit lesion observed in inflammatory myopathy and some muscular dystrophy. Ratings of fibrillation potential and positive sharp: (+) 2-3 second spontaneous potential release was detected at least two locations in at least one muscle; (++) spontaneous potential was detected at three or more locations in at least one muscle. In the current case, myograms in the extremities revealed spontaneous fibrillation potential and positive sharp wave + ~ ++ reflected severe myocytes necrosis and secondary neurogenic lesions in IMNM.

Enzyme histochemistry and immunohistochemistry are the major methods for diagnosis and differential diagnosis of myositis. Enzyme histochemistry in IMNM was the predominant method that detected myocytes necrosis, with degeneration and regeneration and lack of inflammatory cells infiltration. In PM, the inflammatory cells were distributed in a patchy pattern. In DM, the inflammatory cells were focally distributed among small veins. Myocytes atrophy surrounded the muscle fiber beam. In IBM, characteristic inclusion bodies were seen in the cytoplasm and/or nucleus. Immunohistochemistry staining is vital for differential diagnosis [25]. In IMNM, the necrotic fiber membrane or cytoplasmic antigen, MHC-1, expressed positive granular brown substance, C5b-9 membrane complex on the capillaries, and myolemma [26]. Macrophages eventually perform muscle cell necrosis. In PM, "CD_8/MHC-I complex" was expressed. In DM, MHC-I/B cells/CD_4^+ T-cells antigens were fascicularly distributed among vessels. In the current case with little inflammatory cell infiltration and CD8 (-), the PM was removed. No perivascular inflammatory cell infiltration or scattered distribution was observed in the atrophic muscle fibers. B-cells related to CD20 (-), helper T-cells related to CD_4 (+), and MHC-I (++) led to the removal of DM. In addition, the characteristic inclusion body was not found and IBM was removed.

Macrophage-related antigen CD68 (++) and sarcolemma MHC-I (++) were factors that led to the diagnosis of IMNM.

IMNM is a rare disease. Randomized controlled clinical studies have been lacking. Thus, no feasible treatment strategy is available. Moreover, 77% of SRP-positive patients require combined immunotherapy. In Japan, combining IVIG with tacrolimus is common. In the USA and Europe, the combination of mycophenol, methotrexate, and azathioprine is common. More than half of SRP-positive patients show poor effects on all kinds of immunotherapy [26]. Hormone therapy for anti-SRP antibody IMNM is usually 1 mg/kg. Hormone and immunosuppressive agents rarely stop drug administration due to high recurrence rates (40-70%) [27]. Treatment recovery can be reflected by improvement of muscle strength, decreased serum CK, and decreased doses of immunosuppressive agents. In this case, the initial use of intravenous methylprednisolone caused a marked reduction in muscle enzymes, but muscle strength progressively declined. Sequential intravenous immunoglobulin and oral methotrexate administration resulted in low levels of muscle enzymes. Recovery of muscle strength was slow. What caused hormones to take effect too slowly? It inhibits immune inflammation to avoid further immune damage and stabilize the muscle cell membrane. Levels of muscle enzyme decrease significantly. However, recovery of muscle strength is obviously delayed. After 2 months of symptom recurrence, progression in ventilation, immunotherapy of IVIG, and tacrolimus-improved muscle strength are not distinctly observed. Underlying factors may be early hormone withdrawal, leading to relapse. After three plasma exchanges and methotrexate (15 mg, 1/week) and small doses of hormone maintenance therapy, the patient was confined to the bed, but escaped the ventilator. Furthermore, the prognosis of anti-SRP-positive IMNM was not good.

Disclosure of conflict of interest

None.

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