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Multifocal Motor Neuropathy. A Clinical Case and Literature Review

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Summary. Multifocal motor neuropathy is a rare motor neuron disease characterised by a progressive, gradual loss of muscle strength, although sensory functions are preserved. The worldwide frequency of this disease is estimated at 1 person per 100,000 people. MMN is diagnosed when there are characteristic clinical signs, electroneuromyography test results support the clinical findings, and other possible diagnoses are excluded. To prove the progressive course of the disease, the patient must be under long-term medical supervision, since the deteriorating state of the patient may imitate other diseases affecting the motor neuron, such as amyotrophic lateral sclerosis, chronic inflammatory demyelinating polyneuropathy, spinal muscular atrophy, and other.

We present the clinical case of a 71-year-old patient who presented to the Neurology Department of the Hospital of Lithuanian University of Health Sciences Kauno Klinikos. She complained of pain in her right hip and progressing weakness of the right foot which had lasted for a year. After laboratory tests, long-term supervision, and exclusion of other diagnoses, the patient was diagnosed with an atypical type of multifocal motor neuropathy responsive to treatment with intravenous immunoglobulin.

Keywords: multifocal motor neuropathy, MMN, motoric, electroneuromyography, amyotrophic lateral sclerosis.

INTRODUCTION

Multifocal motor neuropathy (MMN) is a relatively rare disease affecting motor neurons and characterized by progressive muscle weakness with muscle atrophy. The worldwide incidence of the disease is about 1 case per 100,000 people [1]. If the disease is diagnosed in time, the prognosis is positive: most patients respond well to treatment with intravenous immunoglobulin, and some patients achieve long-term remission.

Amyotrophic lateral sclerosis (ALS) is a slightly more common (up to 3 cases per 100,000 people) [2] progressive neurodegenerative disease that causes muscle weakness, disability, and eventually death. In contrast to MMN, this disorder has a significantly worse prognosis, with expected survival of 3-5 years [3]. Disease-modifying drugs (e.g., riluzole, edaravone) can be prescribed to slow down, but not to cure the disease [4, 5].

Given the differences in treatment and outcomes, it is very important to differentiate and reliably diagnose these two diseases (as well as other motor neuropathies). Currently, the diagnosis of MMN, ALS, and other motor neuropathies is usually based on clinical presentation and neurophysiological (e.g., electroneuromyography) test results [6–8]. Unfortunately, even based on these criteria, up to 10% of ALS diagnoses are incorrect [9, 10]. If researched further, 1 in 5 of these cases is diagnosed as MMN, one of the most common ALS-like diseases [11].

Ultrasound examination of various peripheral nerves is considered to be a promising alternative method to clarify the diagnosis of ALS, MMN, and other motor neuropathies [12–14].

This article presents the case of a patient initially suspected of having ALS based on clinical and neurophysiological examination findings. Ultrasound examination of the vagus nerve increased suspicion for MMN. By administering appropriate treatment (intravenous immunoglobulin infusions), an improvement in motor function was

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achieved, and the positive response to treatment upheld the diagnosis of MMN.

CASE REPORT

A 71-year-old patient presented to the Outpatient Unit of the Neurology Department of the Hospital of Lithuanian University of Health Sciences Kauno Klinikos (LSMUL KK) in May 2018 due to a year-long pain in the loins, hips, left leg, and weakness of the right leg. The patient noticed that the ankle of the right leg began to swell, the muscles of the legs, arms, and torso (except for the face) were twitched, and the stomach began to tighten. According to the patient, the muscle twitching had recently become more frequent and widespread, due to which daily fatigue had increased.

Changes in muscle strength and tendon reflexes observed on neurological examination were similar to those observed in December 2017. Muscle strength in the right leg: 4 points in the proximal part, 4 points in the distal part, 4 points in dorsal flexion, 4 points in plantar flexion, and good strength in other limbs.

In November 2016, the patient suffered a myocardial infarction, a coronary artery angioplasty procedure was performed, and antiplatelet treatment was initiated. At the end of December 2017, she was treated at the Department of Neurology of the Republican Hospital of Kaunas. The results of the neurological assessment of muscle strength are shown in Table 1 and Fig. 1. Magnetic resonance imaging (MRI) of the lumbar spine showed moderate degenerative changes: stenosis of the spinal canal at the height of the L3-L4 and L4-L5 intervertebral spaces. After starting the treatment, the pain in the legs decreased, but muscle weakness in the right leg progressed. Due to progressive muscle weakness, atrophy and pain syndrome, it was decided to perform an electroneuromyography (ENMG), during which damage was detected at the level of the frontal horns or roots. Marked changes characteristic of acute and chronic neuropathic denervation-reinnervation were observed in L5 and S1 myotomes on both sides. Abundant fasciculations without clear signs of denervation in the proximal muscles of the legs and single fasciculations in the muscles of the arms were registered. No sensory nerve conduction disorders were detected.

As the patient's condition worsened and the diagnosis remained unclear, she was hospitalized at the Neurology Department of LSMUL KK in December 2018. An extensive examination for possible oncological, paraneoplastic processes and chronic infections was performed, but the cause was not found.

Firstly, the clinical condition was differentiated from ALS, however there was insufficient evidence to validate this illness. According to the El Escorial criteria (revised in 2015), to confirm this diagnosis using ENMG examination, it is necessary to determine the signs of damage to upper and lower motor neurons in at least 3 body regions. In this case, lower motor neuron lesions were found in the right and left leg areas. Although the patient's complaints lasted for about 1.5 years and had lately progressed, only the weakness of the right leg developed, and there were no signs of bulbar nerve damage characteristic of ALS disease (choking, swallowing disorder, voice change, shortness of breath). The diagnostic criteria were not sufficient to confirm this disease at that time.

Ambulatory follow-up under the supervision of a neurologist, ENMG, and transcranial magnetic stimulation (TMS) examination to evaluate central motoneuron function after 4 months were recommended. The ENMG in

Table 1. **Assessment of muscle strength according to the Lovett scale, change in symptoms over time, and additional symptoms**

Muscle group		December 2017	December 2018	April 2019	February 2020 (after treatment)
Right arm	Proximal	5	+ fasciculations in the shoulder girdle	5	5
	Distal	5		4	4
Left arm	Proximal	5	+ fasciculations in the shoulder girdle	5	5
	Distal	5		4	4
	Thenar eminence			Atrophy	Atrophy
Right leg	Proximal	4	+ significant atrophy	2 + patellar reflex ( )	3
	Distal	4	+ significant atrophy	2	3
	Dorsal flexion	4	3	0	2
	Plantar flexion	4	3 + Achilles reflex (–)	1	3
Left leg	Proximal	4	+ patellar reflex ( )	3	3
	Distal	4		2	3
	Dorsal flexion	4		3	4
	Plantar flexion	4	3 + Achilles reflex (–)	3	4

0 – no movement of the limb, 1 – weak muscle contraction without movement of the limb, 2 – full-amplitude movement of the limb is visible but it does not overcome the force of gravity, 3 – can overcome gravity but does not resist the opposing force, 4 – slightly resists the opposing force, 5 – fully resists the opposing force.
( ) – weakening, (–) – extinction

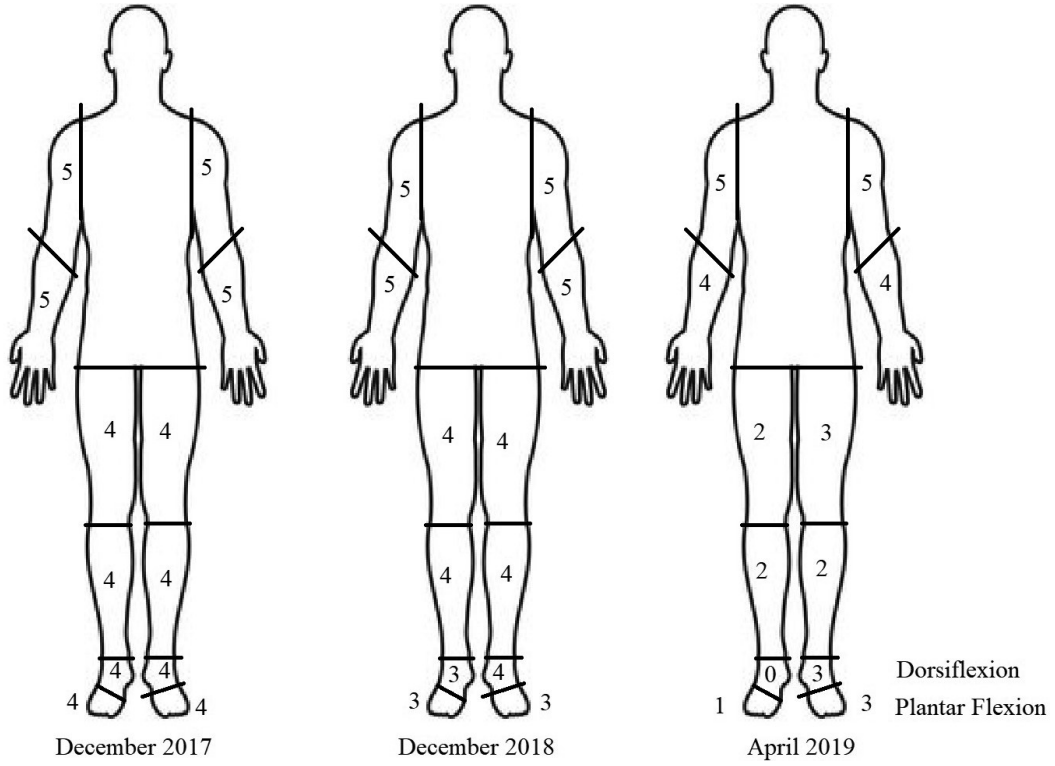

Fig. 1. **Dynamics of the patient's muscle strength assessment (according to the Lovett scale)**

April 2019 showed a worsening of partial axon degeneration of motor fibres compared to the results in November 2018.

Needle EMG showed signs of acute and chronic neuropathic denervation-reinnervation in distal muscles, especially in the right. Fasciculations were registered in the arms and legs proximally. Transcranial magnetic stimulation showed no abnormalities in central motoneuron conduction.

The patient was consulted several times at the Outpatient Department of Nervous System Diseases of LSMUL KK Despite rehabilitation and medical treatment, the patient's disease progressed. Especially since April 2019, her balance started to deteriorate, and there were repeated falls. The patient moved only with walking aids. Limping and urination disorders appeared. It was decided to re-hospitalize the patient at the Neurology Department.

In August 2019, the patient was treated at the LSMUL KK Neurology Department. Compared to previous neurological examinations, the strength of the right leg had further decreased. The patient underwent repeated blood and cerebrospinal fluid tests at the department, which showed no deviations from normal limits. No focal density, structural, or inflammatory changes were observed in the brain computed tomography (CT) scan.

To aid in the process of differential diagnosis, a combination assay was conducted to detect specific IgG antibodies against antigens that are commonly linked to paraneoplastic neurological syndromes. 12 specific antibodies were tested. Onconeuronal antibodies, antibodies related to neuronal cell surface antigens, and paraneoplastic diseases were studied: 1) Anti-amphiphysin (Anti-Amp), 2) Anti-CV2 (CRMP5), 3) Anti-PNMA2/Ta (Ma2/Ta), 4) Anti-Ri (ANNA-2), 5) Anti-Yo (PCA-1), 6) Anti-Hu (ANNA-1), 7) Anti-Recoverin (Rec), 8) Anti-SOX1, 9) Anti-Titin, 10) Anti-Zic4, 11) Anti-GAD65, 12) Anti-Tr (DNER). All antibody tests were negative. In addition, in search of genetic pathology, the patient was consulted by a geneticist and genetic material was taken for DNA testing for spinal muscular atrophy (type IV), but the patient refused a muscle biopsy. Spinal muscular atrophy was ruled out.

A repeat MRI of the C1-Th3 spine was ordered. The changes were considered to be of degenerative origin, although they did not explain the neurological symptoms. No pathological changes were found in the spinal cord.

After 4 months, the ENMG was repeated and negative dynamic was noted. In the legs, signs of severe axon degeneration of peripheral motor nerves were observed (right more than left). Fasciculations were registered in all leg muscles, especially in the proximal muscles. Both in the right leg and the calf of the left leg, there were abundant signs of acute denervation, as well as signs of chronic neuropathic denervation-reinnervation. The conduction velocities of sensory nerves were intact. Isolated fasciculations in the hands without signs of denervation. Signs of slight focal demyelination of the right sensory fibres of the median nerve in the area of the carpal tunnel. TMS again showed no evidence of impaired central motoneuron conduction.

In February 2020, the patient was re-hospitalized to the LSMUL KK Neurology Department to clarify the diagnosis. So far, most of the data were suggestive of ALS, although TMS still showed no signs of central motor neuron

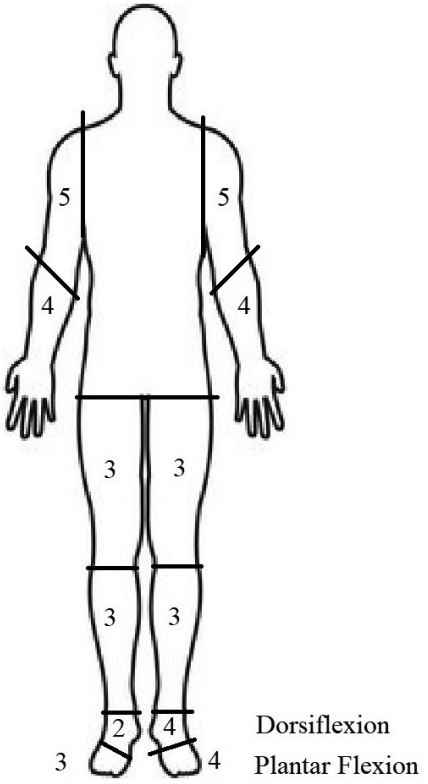

Fig. 2. **The patient's muscle strength after treatment (according to the Lovett scale)**

damage and bulbar symptoms had not appeared since 2017 (3 years since symptom onset).

From the patient's history it was known that the patient had been ill for 3 years, and that her condition worsened especially in April 2019. Until then, the woman walked with a cane, for the last year she walked with a low walker, but now she needed a wheelchair for movement. The woman claimed that it was becoming more difficult to serve herself at home, she could not lift things. The change in the results of the neurological examination is shown in Table 1. No deviations from normal limits in the performed blood, biochemical, and urine tests were observed. Repeated ENMG revealed signs of axon degeneration of the motor nerves in the legs (right more than left) and symptoms of acute and chronic neuropathic denervation and reinnervation in the proximal and distal leg muscles. If the conduction of the sensory nerves of the legs was intact, the mentioned changes could be due to the damage to the motor nerves in the proximal parts at the level of the frontal horns. Focal demyelination of the sensory and motor nerves of the right median nerve was also found in the wrist area; only fasciculations were registered in the hand muscles without other signs of denervation. Compared to the examination in December 2019, a worsening of the symptoms was observed and denervation in the proximal muscles of the legs also occurred.

To differentiate neurodegenerative disease from polyneuropathy, an ultrasound examination of the vagus nerve was performed. The changes found are characteristic of polyneuropathy: the area of the nerve in the transverse plane is increased and hyperintense inclusions are present. The condition was repeatedly discussed with the head of the Department of Peripheral Nerve Diseases. After assessing the course of the disease, deterioration of the condition, and diagnostic tests, it was decided that this could be motor multifocal polyneuropathy with an atypical course. A course of intravenous immunoglobulins was prescribed for treatment (2 g/kg). After the prescribed treatment, the patient's condition improved significantly (Table 1). The neurological examination showed (Fig. 2) that the patient's leg strength increased and she was able to walk without the help of a wheelchair.

Finally, when a good response to treatment with intravenous immunoglobulin was observed, knowing the patient's history (duration of symptoms (more than 3 years), progressive asymmetric leg and hand distal part weakness, atrophy of the right leg muscles) and taking into account ENMG data (persistent progression of tibial, peroneal and medial motor nerve fibre damage, unchanged sensory function), normal TMS results, as well as the exclusion of other diseases (blood, biochemical and urinalysis were normal, CT and MRI of the spine did not correlate with the progression of the disease), the patient was diagnosed with atypical multifocal motor neuropathy.

Ultrasonography of the vagus nerve

In the following paragraph, we present information collected from studies on ultrasound morphology of peripheral nerves in patients with motor neuron diseases. Several studies have described a reduction in the cross-sectional area of the ulnar nerve in patients with ALS [15], and the median nerve has also been investigated. It has been shown that the vagus nerve is also significantly smaller in patients with bulbar ALS than in healthy patients [16, 17]. Interestingly, an increase in the cross-sectional area of peripheral nerves has been observed in patients with MMN. Such changes have been described by Greek and South Korean researchers studying the median, ulnar, and tibial nerves [18, 19]. An increase in the cross-sectional area of the vagus nerve in patients with MMN has also been found [12], although it is theoretically known that signs of damage to cranial nerves are not characteristic of multifocal motor neuropathy [20].

An ultrasound examination of the vagus nerve was also performed on the patient treated at the LSMUL KK Neurology Department whose case we present: the vagus nerve with a cross-sectional area of 3.84 mm^2 on the right and 2.32 mm^2 on the left near the bifurcation of the carotid artery was found (Fig. 3). According to the literature, the right vagus nerve is statistically significantly larger since its average normal size is 2.7±0.6 mm^2. The average normal size of the left vagus nerve is 2.1±0.5 mm^2 [21, 22]. Based on this clinical measurement result of the vagus nerve and on literature data, it was observed that the patient's cross-sectional area of the nerve exceeded normal limits, which is consistent with the hypothesis of nerve enlargement in patients with MMN.

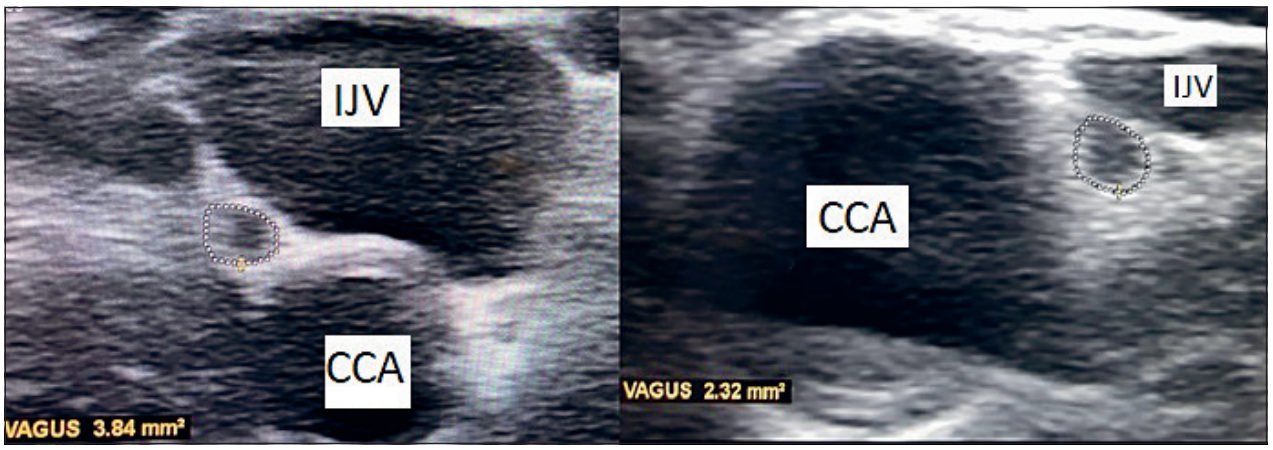

Fig. 3. **Vagus nerve measurements in the patient with multifocal motor neuropathy (MMN) in the presented clinical case**
Abbreviations: IJV – internal jugular vein, CCA – common carotid artery. Circled – vagus nerve. On the right – the left vagus nerve, on the left – the right vagus nerve.

LITERATURE REVIEW

Epidemiology

Multifocal motor neuropathy is a rare disease affecting motor neurons. According to data from different countries, the incidence of MMN is between 0.6-2 cases per 100,000 inhabitants [1, 23]. The disease usually first appears in patients younger than 50 years of age (80% of cases, average age 40 years), although the age of onset of the disease ranges from 20 to 70 years [1]. Cases have been published in which the disease occurred in a 6-year-old child [24] and an 8-year-old child [25] or older. MMN is 2.7 times more common in men than in women [26].

Aetiology

MMN is currently thought to be an autoimmune disease due to the presence of anti-GM, i.e., anti-ganglioside antibodies, in most patients. The immune origin of the disease is also supported by a positive clinical response to treatment with intravenous immunoglobulin and histological findings of nerve biopsy – perivascular infiltration of lymphocytes [27]. Antiganglioside GM1 is exceptionally abundant in motor nerve myelin compared to sensory nerve fibres [26]. A positive anti-GM1 antibody test is likely to explain the clinical course of MMN, namely, motor nerve damage and preserved sensory function. Although MMN was previously thought to be caused by neural demyelination, studies have shown that anti-GM1 antibodies disrupt the function of sodium and potassium channels in the isthmus of Ranvier, thereby disrupting conduction in motor neurons [26]. Articles have been published describing the iatrogenic manifestation of MMN: the disease occurred after immunotherapeutic treatment of Crohn's disease [28, 29], rheumatoid arthritis [30], psoriasis [31, 32] with tumour necrosis factor-alpha (TNF) inhibitor infliximab. After discontinuation of the drugs, the motor functions of the limbs partially or completely recover within 1 year.

Clinical features

The disease usually manifests as subacute distal asymmetric muscle weakness of the limbs (mostly hands, but possibly also legs) without sensory disturbances. Patients may complain of unilateral weakness of the muscles of the wrist, fingers, forearm, and foot [26]. Frequent muscle spasms (cramps), fasciculations, tingling sensations or even pain can also be bothersome. These symptoms can be exacerbated by cold in up to 83% of cases [28], although they are less pronounced compared to motor dysfunction. It has been observed that symptoms more often progress in the non-dominant hand, and cramps and fasciculations occurring in up to 40% of cases may cause hand muscle hypertrophy [28]. Impairment of motor function usually progresses and affects other limbs. Cranial nerves or respiratory muscles are usually not involved [33]. During the neurological examination, MMN-specific signs of damage to the lower motor neurons can be observed: in addition to decreased strength and preserved sensory function, weakened deep tendon reflexes may also be detected, but not always [34]. The absence of signs of upper motor neuron damage – increased tendon reflexes, spasticity – helps to differentiate it from ALS [26].

Diagnostics

Diagnostic criteria, according to the latest (2010) guidelines of the European Federation of Neurological Societies and the Peripheral Nerve Society [20], are based on the two following main criteria: slowly progressive focal and asymmetric limb weakness, in other words, impairment of the function of more than two motor nerves for more than a month. If symptoms are found in only one nerve, the diagnosis is only suspected. In addition, no sensory disorders should be found on objective examination, except for a possible impaired sensation of slight vibration in the legs. The diagnosis is ruled out if there are upper motor neuron and prominent bulbar symptoms, if sensory symptoms are not limited to a mild loss of vibration sensation in the legs, or if there is widespread symmetrical weakness in the first weeks.

Other, auxiliary clinical criteria can also help in establishing the diagnosis: the hands are most affected, weakened or absent tendon reflexes in the affected limbs, intact cranial nerves, spasms and fasciculations in the affected limb. In addition, an achievable response (observed relief of disability or increase in muscle strength) during the administration of immunomodulatory treatment with intravenous immunoglobulin in conjunction with anti-GM1 antibodies, a positive MRI, or cerebrospinal fluid test also help in confirming the diagnosis.

Diagnostic criteria

1. Clinical: must meet both primary criteria and none of the exclusion criteria.
2. Neurophysiological: reliable or probable conduction block (CB) in at least one nerve (Table 2).
3. Supportive: anti-GM1 antibodies found, positive MRI, cerebrospinal fluid test results, positive response to treatment.
4. Possible categories: confirmed diagnosis of MMN or probable diagnosis of MMN.

Guidelines for diagnostic tests

1. All patients suspected of having this condition should undergo an objective examination and neurophysiological electrodiagnostic tests.
2. In selected patients, laboratory tests for antibodies to ganglioside GM1, MRI imaging of the brachial plexus, and cerebrospinal fluid should be considered.
3. In individual cases, possible additional tests to detect or rule out other diseases may be considered.

Additional criteria supporting the diagnosis

1. Increased levels of IgM anti-ganglioside GM1 antibodies.
2. Laboratory findings: increased protein level in cerebrospinal fluid (<1g/L).
3. Increased signal intensity on T2 MRI sequence, which is characteristic of a widespread inflammatory reaction of the nerves of the brachial plexus.
4. Objectively observed relief of clinical symptoms after administration of intravenous immunoglobulin.

DIFFERENTIAL DIAGNOSES

Amyotrophic Lateral Sclerosis

Although both diseases, ALS and MMN, initially manifest with similar motor symptoms, disease progression is slower in the latter. The most significant difference is that in the case of ALS, in contrast to the MMN, the central (upper) motoneuron as well as the respiratory and cranial nerves are affected [26]. Muscle weakness in MMN occurs in areas innervated by specific motor nerves, rather than in the entire myotome at once, which is more common in ALS. Muscle atrophy and fasciculations are also more common in ALS. ENMG shows typical conduction blocks along with demyelination, while in the case of ALS, conduction in motor and sensory fibres is normal, but fibrillations can be detected in arm and leg muscles along with large action potentials of motor units (increased amplitude and duration) [35]. Transcranial magnetic stimulation can also help objectively investigate central motor neuron damage.

Table 2. **Electrophysiological criteria for conduction block** [20]

1. Definite motor CB*
Negative peak CMAP area reduction on proximal vs. distal stimulation of at least 50% whatever the nerve segment length (median, ulnar, and peroneal). Negative peak CMAP amplitude on stimulation of the distal part of the segment with motor CB must be >20% of the lower limit of normal and >1 mV and increase of proximal to distal negative peak CMAP duration must be 30%
2. Probable motor CB*
Negative peak CMAP area reduction of at least 30% over a long segment (e.g., wrist to elbow or elbow to axilla) of an upper limb nerve with increase of proximal to distal negative peak CMAP duration 30%
OR
Negative peak CMAP area reduction of at least 50% (same as definite) with an increase of proximal to distal negative peak CMAP duration >30%
3. Normal sensory nerve conduction in upper limb segments with CB (see exclusion criteria)

CB – conduction block; CMAP – compound muscle action potential.
*Evidence for CB must be found at sites distinct from common entrapment or compression syndromes.

Cases have also been described where ultrasound examination of peripheral nerves has helped distinguish ALS from chronic autoimmune neuropathies [11]. The inhomogeneous, asymmetric nerve cross-sections visible in our case study allowed us to suspect an autoimmune inflammatory process and thus distinguish motor neuropathic disease from ALS.

It is important to distinguish MMN from ALS because the former has a relatively good response to treatment (intravenous immunoglobulin) and a better prognosis.

Progressive Muscular Atrophy

One form of ALS, progressive muscle atrophy (PMA), affects only the peripheral (lower) motoneuron [36, 37]. This form of ALS is more difficult to distinguish from MMN because it does not have the central motor neuron lesions characteristic of the classical ALS form, although postmortem data show ALS-like (but not clinically evident) lesions [38]. As the mechanism of the disease is similar to ALS, typical changes (muscle denervation in 88% of patients, as well as motoneuron conduction disorders) are observed on ENMG examination [39], which helps to distinguish PMA from MMN.

Chronic Inflammatory Demyelinating Polyradiculoneuropathy

Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) usually presents with chronic symmetrical muscle weakness, more commonly affecting the legs than the arms, as well as hyporeflexia, and with sensory disturbances (unlike MMN). Differential diagnosis can be complicated by the fact that sometimes CIDP can manifest only with motor symptoms. In addition, conduction block can be detected in ENMG in both diseases. As for the cerebrospinal fluid examination, both CIDP and MMN show an increase in protein without an increase in cell count. In contrast to MMN, anti-GM1 antibodies are usually absent in CIDP. Patients with CIDP are effectively treated with corticosteroids and plasmapheresis [34]. One of the rare variants of CIDP, multifocal acquired demyelinating sensory and motor neuropathy affecting individual nerves, may resemble MMN, but the presence of prominent sensory disturbances and neurophysiological studies help distinguish this disease from MMN [26, 34].

Hereditary Neuropathy with Liability to Pressure Palsies

Hereditary Neuropathy with Liability to Pressure Palsies (HNPP) is manifested by asymmetric weakness of muscles innervated by various nerves and conduction blocks detected by ENMG examination. In contrast to MMN, these blocks appear at typical sites of nerve compression (i.e., the median nerve in the carpal tunnel or the ulnar nerve at the elbow). In addition, unlike MMN, sensory symptoms also occur. The disease is inherited in an autosomal dominant pattern – similar cases often occur in the family. If the disease is suspected, appropriate genetic tests are available (searching for *PMP-22* gene deletion) [26, 34].

Spinal Muscular Atrophy, type IV

Spinal muscular atrophies (SMA) are a group of hereditary diseases characterized by degeneration of peripheral (lower) motor neurons, leading to progressive muscle weakness and atrophy. Cell death is caused by the SMN protein deficiency developed due to the *SMN1* gene mutation, which can only be partially compensated by the *SMN2* gene product. The more copies of this gene variant, the later and lesser the symptoms of the disease. In the case of type IV, slowly progressive muscle weakness, atrophy, and deformities occur around the age of 35. EMG examination shows signs characteristic of muscle denervation (abnormal spontaneous activity: fibrillations, fasciculations, positive sharp waves). Serum creatine kinase may be normal or elevated. The diagnosis is based on the *SMN1* gene deletion/mutation detected by molecular genetic testing [35].

Treatment

Treatment options for patients with multifocal motor neuropathy are currently limited. Unlike some similar polyneuropathies, corticosteroids or plasmapheresis have not been proven effective in this disease, but there is evidence of their potential harm [33]. Currently, the most effective known drug is intravenous immunoglobulin (IVIg) [40]. The standard dose is 0.4 g/kg/d for 5 days, the total dose is 2 g/kg. More than 3 out of 4 patients respond positively to IVIg administration, but only 1 out of 5 achieve long-term remission, and the rest require periodic infusions. Administration of IVIg has been proven to increase muscle strength, which is the most important cause of disability in MMN [26]. No association was found between treatment, anti-ganglioside GM1 antibodies, and ENMG test results [41]. Treatment with alternative drugs (i.e., cyclophosphamide, rituximab) may also be appropriate for IVIg-resistant cases, but their benefits are still controversial [42].

Prognosis

The prognosis for multifocal motor neuropathy is generally positive. About 4 out of 5 patients have a positive response to treatment with intravenous immunoglobulin; 1 out of 5 achieve long-term remission, other patients require repeated IVIg infusions. Treated patients maintain their ability to work; in one study lasting about 5 years, 94% of patients did not lose their job [43]. Without treatment, slow deterioration of motor function with muscle atrophy in the last stages is predicted, and spontaneous remission is not expected.

CONCLUSION

In the article, we discussed the clinical case of a 71-year-old female patient who presented to the LSMUL KK Neurology department in May 2018 due to a year-long pain in the loins, hips, and right leg weakness. The patient noticed that the muscles of the legs, arms, and torso (except for the face) were twitching and that stomach muscles began to tighten. Body muscle twitches, according to the patient, had become more frequent and widespread, due to which daily fatigue increased. These complaints had been disturbing the patient for about a year. In the last 2 years, the patient was treated 3 times in the LSMUL KK Neurology Department where her condition was actively monitored. Since the first visit, the patient's muscle strength had decreased significantly. In the proximal part of the right leg, muscle strength decreased from 4 to 2 points, dorsiflexion from 4 to 0 points, and plantar flexion from 4 to 1 point. Muscle weakness in the left leg also progressed: proximal 2 points, distal 2 points. Fasciculations appeared in the muscles of the shoulder girdle, the strength of the hands decreased to 4 points, and atrophy of the thumb muscles of both hands developed.

According to the guidelines of the European Federation of Neurological Societies and the Society of Peripheral Nerves reviewed in the literature [20], the clinical course of this patient's disease meets 2 main clinical criteria: firstly, she developed a slowly progressive focal and asymmetric limb weakness, which was manifested by more than two

motor nerve dysfunctions lasting for more than a month. Also, neither the data of the objective examination, nor the results of the ENMG and TMS tests showed signs of damage to the sensory and upper motor neurons. During the 3 years of the disease, the patient did not develop bulbar symptoms, which helped to rule out the diagnosis of ALS.

An ultrasound examination of the vagus nerve was also performed on this patient in the LSMUL KK Neurology Department. As some studies show, in patients with MMN there is an increase in the cross-sectional area of peripheral nerves [18, 19], as well as an increase in the cross-sectional area of the vagus nerve [12]. Since the patient's ultrasound results showed that the cross-sectional area of the vagus nerve was significantly larger than it should be, this helped to suspect the diagnosis of MMN and conduct further research in this direction.

Other, auxiliary clinical criteria also helped to establish the diagnosis of MMN: undetectable tendon reflexes in the affected limbs, intact cranial nerves, spasms and fasciculations in the affected limb. Also, an achievable response (observed relief of disability or increase in muscle strength) when administering immunomodulatory treatment with intravenous immunoglobulin helped prove the diagnosis of MMN. After the patient was given a 5-day intravenous course of human immunoglobulin, her condition improved significantly. Until then, the patient hardly walked and used a wheelchair. After the prescribed treatment, the patient was able to stand up independently and walk with a walker.

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DAUGIAŽIDININĖ MOTORINĖ NEUROPATIJA. KLINIKINIS ATVEJIS IR LITERATŪROS APŽVALGA

Santrauka

Daugiažidininė motorinė neuropatija diagnozuojama esant charakteringiems klinikiniams požymiams, kurie paremti elektroneuromiografijos tyrimo rezultatais ir kitų panašių ligų atmetimu. Tam tikrais atvejais, ligai progresuojant, reikalingas paciento stebėjimas, kadangi simptomai dažnai būdingi ir šoninei amiotrofinei sklerozei, lėtinei uždegiminei demielinizuojančiai polineuropatijai, spinalinei raumenų atrofijai ar kitoms ligoms.

Mes pristatome klinikinį atvejį: apie 71 metų pacientė atvyko į Lietuvos sveikatos mokslų universiteto ligoninės Kauno klinikos Neurologijos ambulatorijos skyrių dėl metus trunkančio skausmo strėnose, klubuose ir atsiradusio dešinės kojos silpnumo. Po atliktų tyrimų, ilgai trukusio stebėjimo, atmetus kitas ligas, pacientei nustatyta netipinės eigos veiksmingai intraveniniu imunoglobulinu gydyta daugiažidininės motorinės neuropatijos forma.

Raktažodžiai: daugiažidininė motorinė neuropatija, DMN, motorika, elektroneuromiografija, šoninė amiotrofinė sklerozė.

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