Refractory Form of Chronic Idiopathic Demyelinating Polyneuropathy with Rituximab Effect: A Case Report

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ABSTRACT

The basic characteristic of chronic idiopathic demyelinating polyneuropathy (CIDP) is demyelination of peripheral nerves of autoimmune origin. Antibodies against various myelin sheath proteins play an important role in the pathogenesis of the disease. Cell-mediated immunity, characterised by T-cell and macrophage infiltration in peripheral nerves and spinal roots, is also significantly involved (1). Treatment of CIDP is aimed at suppressing inflammation, but also at removing autoantibodies, cytokines and other pro-inflammatory molecules from the blood. Treatment options include corticosteroids, (intravenous or subcutaneous) immunoglobulins, and plasma exchange (plasmapheresis). If a patient with CIDP does not show an adequate clinical response to these three treatment modalities, it is then refractory CIDP. Based on various studies, this affects up to 10% of patients.

We present here a patient with CIDP whose condition gradually stopped improving after all three commonly used treatment modalities (corticosteroids, immunoglobulins, and plasmapheresis). His neurological findings were very severe. The case was ultimately diagnosed as a refractory form of CIDP, which is discussed here in the light of current knowledge of this issue.

KEYWORDS

Chronic Inflammatory Demyelinating Polyradiculoneuropathy; Rituximab; autoimmune diseases; peripheral nervous system diseases; immunotherapy; plasma exchange

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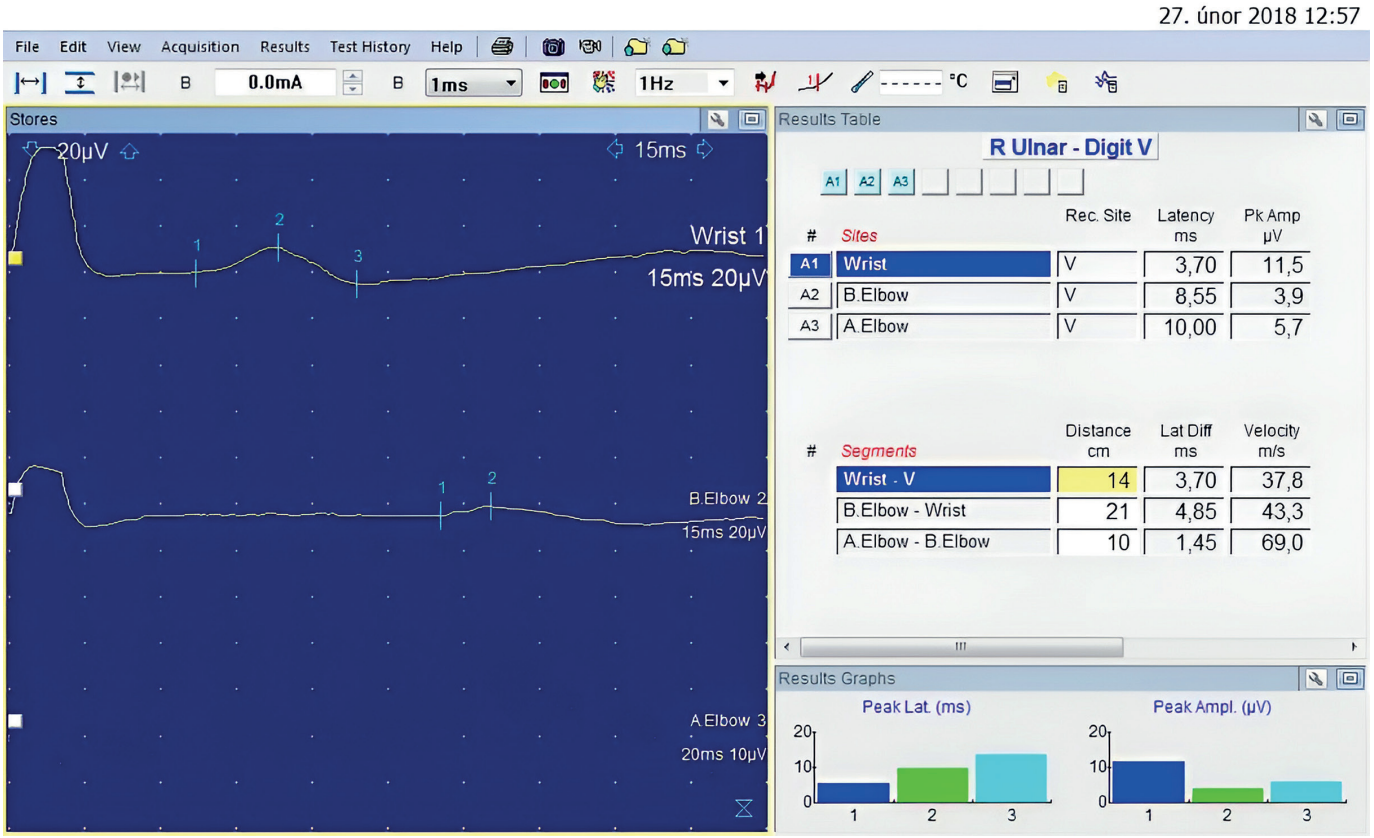
CASE REPORT

An active male, 184 cm tall, weighing 90 kg, born in 1979, was referred to the EMG laboratory in February 2018 with 5 months of progressive right hand motor impairment with loss of strength and sensory disturbance. The clinical findings included reduced finger duction, finger flexion (especially 4th–5th finger), weakening of lumbrical muscles in



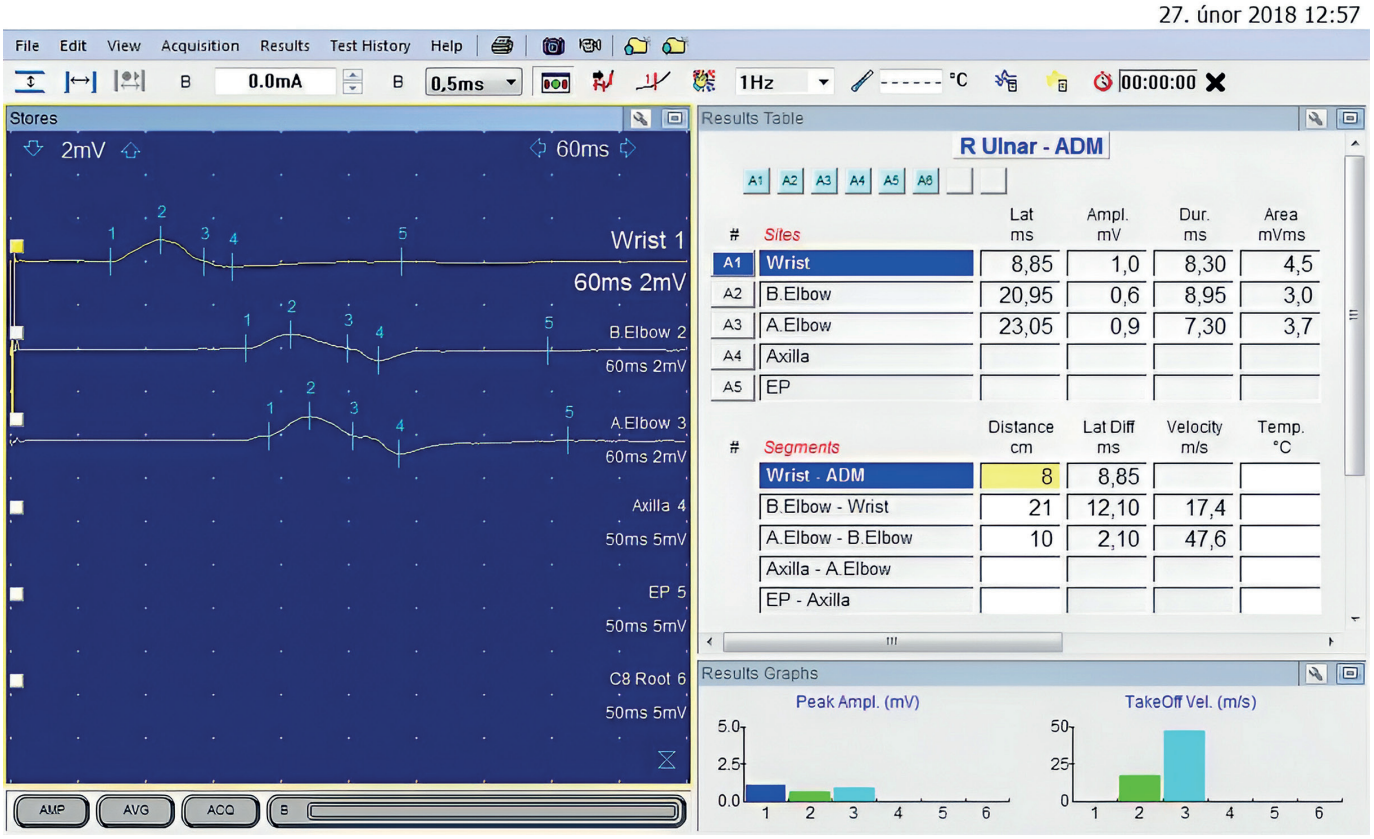


Fig. 1a, b R. ulnar nerve 27/02/2018.
Motor neurography – lesion of the distal part – DML 8.85 ms (3.8 upper limit of norm), duration of CMAP (8.00), A-CMAP 1.0 mV (6.0)m MCV in forearm 17.4 m/s (45.0).
Sensory neurography – longer distal sensory latency 3.70 ms (3.2), SCV 37.90 m/sw (45.0).

these fingers, and sensory disturbance in the hypothenar eminence, the 5th finger and half of the 4th finger. EMG revealed low CMAP following ulnar nerve stimulation (m. ADM, first dorsal interosseus) and slowed motor nerve conduction velocity in the proximal forearm Fig 1. Ultrasound examination showed no clear ulnar nerve

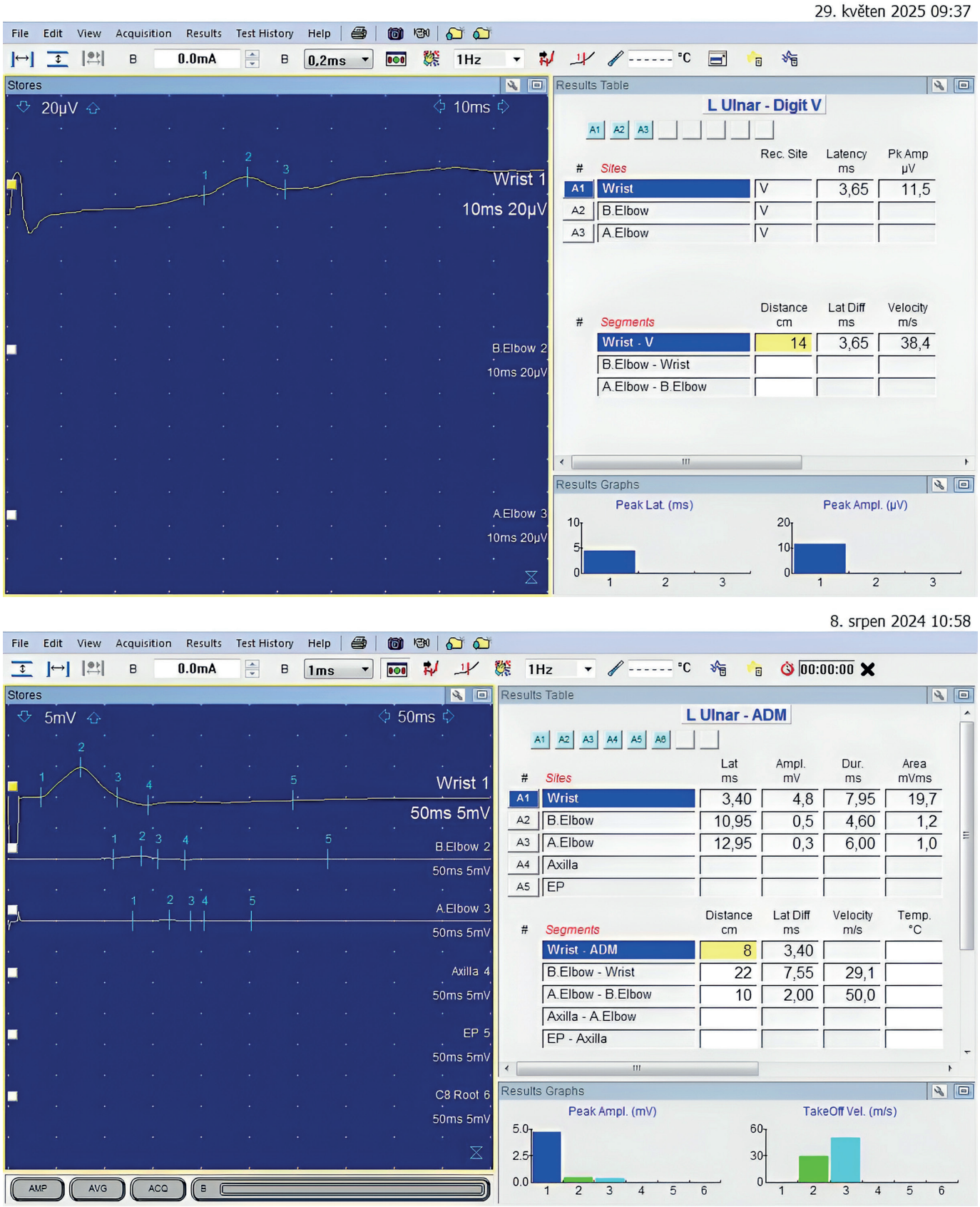


Fig. 2a, b L. ulnar nerve 08/08/2024.
Motor neurography – DML normal value, duration of CMAP 7.95 ms (borderline), A-CMAP 4.8 mV at wrist, 0.5 below the elbow, 0.3 above the elbow (6.0), MCV forearm 29.1 m/ and MCV ocress the elbow 50.00 (50).
Sensory neurography – SNAP latency 3.65 ms (3.80), SCV 38.4 m/s (45).

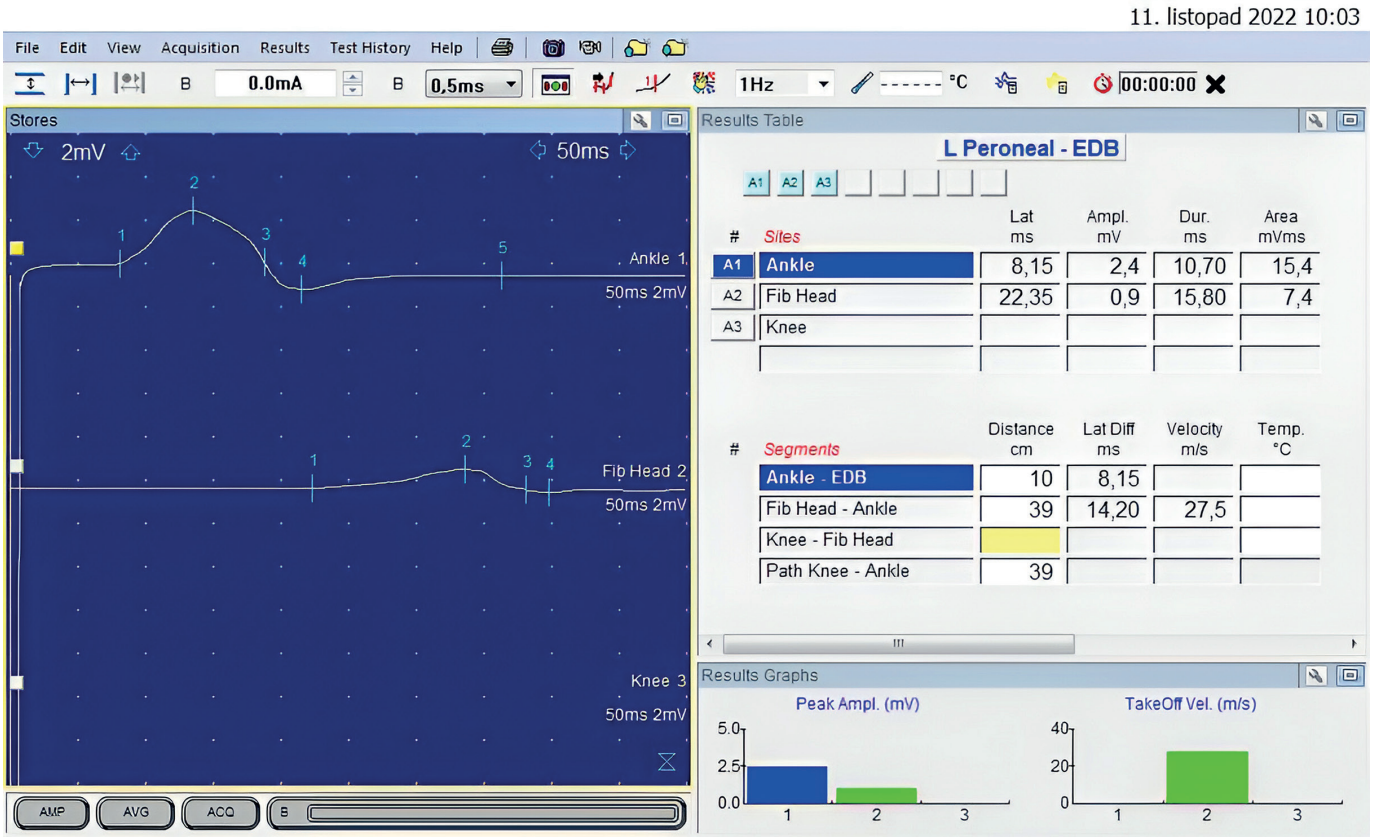


Fig. 3 L. peroneal nerve (extensor digitorum brevis muscle) 11/10/2022.
DML 8.15 ms (5.5), A-CMAP 2.4 and 0.9 mV (2.8), MCV 27.5 m/s (45).

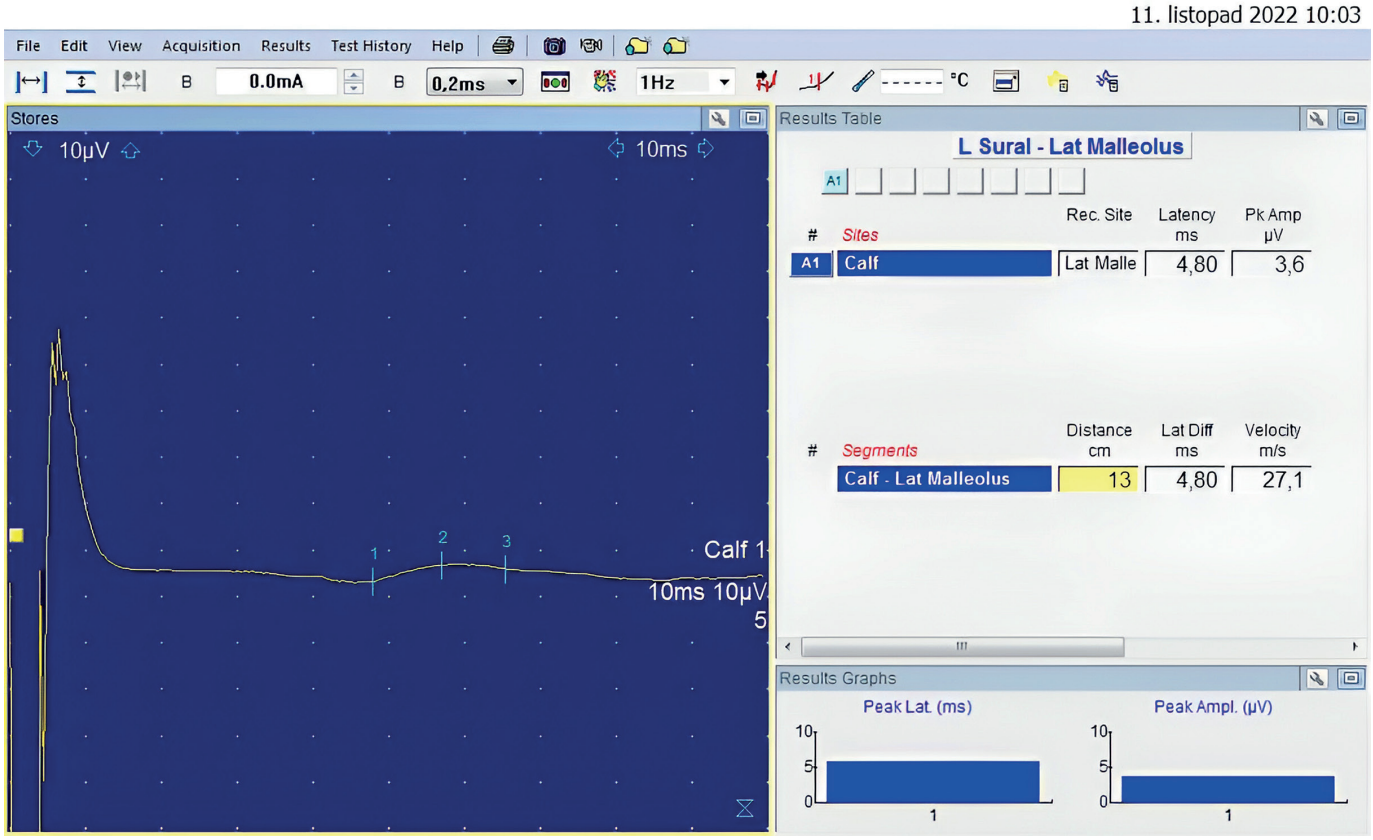


Fig. 4 L. sural nerve 11/02/2022.
SNAP latency 4.8 ms (4.0), A-SNAP 3.5 mV (8), SCV 27.1 m/s (45).

compression in the ulnar compartment. MRI of the ulnar nerve was recommended. On 25 April 2018, ulnar nerve surgery was performed at the elbow, after which there was a slight improvement lasting about 1 month, and then there was further progression, but the patient was not sent for EMG and no MRI was performed. In the spring of 2022, both motor function and sensation in the remaining fingers of the right hand deteriorated; in September 2022, gait and left-hand motor function were also affected. The control EMG from 31 October 2022 was suggestive of multifocal demyelinating disorder. CIDP, probably MADSAM (multifocal acquired demyelinating sensory and motor neuropathy, Lewis-Sumner syndrome) was suspected. Fig 2, Fig 3, Fig 4.

The samples were collected, including liquor (protein 0.8 g/l, glycorrhachia 3.44 mmol/l, 1 monocyte). IVIG therapy was started at a dose of 2 g/kg for 5 days. The patient's mobility improved and he was able to walk around his apartment with forearm crutches. Corticosteroid therapy was then initiated – prednisone 60 mg for 6 weeks with addition of cyclosporine 3×100 mg (Sandimmun). Nevertheless, the disease progressed and affected lower limb mobility so the patient had to use two forearm crutch. IVIG was re-administered at a dose of 70 g every 3 weeks, after which the findings stabilised. However, after the patient was switched to subcutaneous immunoglobulins, the pareses soon progressed, including gait and grip, with more marked deterioration on the left side. Since the autumn of 2023, there has been a steady progression of both paresis and atrophy. The patient then sat on the seat, could not stand up, had to be fed. The therapeutic effect of IVIG has waned gradually.

The laboratory findings identified only a fluctuating slight elevation of creatine kinase (CK) while antibodies against gangliosides were not detected. Cerebrospinal fluid was tested for IgG antibodies – against neurofascin-155, neurofascin-186, contactin, CASPR1 – which were not detected.

In August 2024, the patient was admitted to the neurological ICU, where a total of plasmapheresis (7 cycles) were performed. His condition did not improve, but no further attacks have occurred. He was then hospitalised for rehabilitation. Two weeks after plasmapheresis, IVIG was administered and after 4 days, the mobility of lower limbs and upper limb fingers slightly improved.

The patient has been treated with rituximab since 11 April 2025. At the follow-up in August 2025, no further attack was reported, and he was able to walk around his apartment with assistance and one forearm crutch. He was able to eat with a spoon in his right hand. Subjective sensory disturbances were less pronounced. He takes prednisone 20 mg daily, and cyclosporine 2×100 mg (Sandimmun). He continues with rituximab therapy at a dose of 1 g at first day, after 14 days another 1 g and then 1 g every 6 months. He performs rehabilitation exercises daily, both independently and with assistance from his family, and twice weekly at home under the supervision of a physiotherapist.

DISCUSSION

Recent diagnostic and therapeutic criteria (van den Bergh, 2021) define various forms of CIDP with the development of symmetric polyradiculoneuropathy during the course of the disease (2). There is also a treatment-refractory form (corticosteroids, immunoglobulins, and plasmapheresis), which affects 6–15% (3) of patients and is a serious and intractable issue. In a cohort of 45 patients (USA, Oregon), 11 people (24%) were refractory to first-line therapy. Seven out of 11 patients (64%) responded favourably to alternative treatment – rituximab or cyclophosphamide (4). Risk factors for refractory CIDP include multifocal form of CIDP, insidious onset, progressive course, and concomitant CNS involvement. According to various authors, initial corticosteroid therapy is successful in 68%. IVIG has an effect in 63–78% and plasmapheresis in 42–56% (5).

Our patient started with a lesion of the ulnar nerve on the right side when the neurosurgeon performed surgery in the elbow with little and short-term effect. The requirement for follow-up MRI examination of the ulnar nerve was not met. After 3 years, the patient experienced a significant deterioration in the right upper limb (n. ulnaris and n. medianus) but also had a significant nerve lesion in the left upper limb (unable to eat with his left hand) and in both lower limbs (only able to sit on a seat). This significantly delayed the diagnosis of CIDP, a multifocal variant of the MADSAM type.

Despite the initial good effect of immunomodulatory treatment, the therapeutic effect of immunoglobulins, corticosteroids and immunosuppressants gradually waned. At that time, plasmapheresis (7 cycles) was indicated, followed by 2 weeks of waiting for the immune system to stabilise. Only then did IVIG administration prove effective. Plasmapheresis has a substantially broader therapeutic effect than IVIG. Rituximab was administered in addition to the current therapy. His condition stabilised, sensitivity slightly improved with no further attacks.

It is important to accurately establish this diagnosis for the treatment strategy in the refractory form of CIDP. A large proportion of patients referred with a diagnosis of CIDP to a tertiary centre have another diagnosis (up to 40–60%) (5, 6). Such diseases do not improve with treatment. For example, the myelin sheath damage in nodopathies is not complement-dependent and patients do not respond to immunoglobulins (7).

Refractory forms of CIDP are not rare. Despite long-term administration of IVIG, the indication of plasmapheresis is necessary. Unlike IVIG, plasmapheresis removes autoantigens, cytokines and other pro-inflammatory substances (8). Then the therapeutic effect of IVIG and plasmapheresis is more likely.

In our patient, decompression of the ulnar nerve on the right side was performed hastily and without regard to the neurologist's request. The effect of the surgery was small and short-lived. Consequently, there was a significant delay in establishing a correct diagnosis and, in particular, in initiating effective therapy. After prolonged latency, multifocal polyneuropathy began to develop. Initially, treatment with intravenous immunoglobulins was moderately successful. Gradually, the effect of immunotherapy completely disappeared. The patient was bedridden, with near-plegic distal upper limbs. At that time, plasmapheresis (7 cycles) was indicated, followed by 2 weeks of waiting for the immune system to stabilise. Only then did the administration of IVIG have an effect. After slight improvement,

the patient is now stable on a regular dose of rituximab, prednisone 20 mg and cyclosporine 2×100 mg.

Treatment escalation and azathioprine, mycophenolate mofetil, and cyclosporine are used for the treatment of refractory form of CIDP while cyclophosphamide can be attempted for severe forms (Nascimento, 2025). Rituximab is a chimeric monoclonal antibody (IgG1) targeted against CD20, a surface antigen expressed on B lymphocytes responsible for pathological antibody production. The fact that CD20 is not expressed on stem cells or plasma cells makes rituximab a relatively safe medication, without a substantial risk of bone marrow suppression or impairment of long-term immunity. Various adverse events may occur after rituximab administration. The most common side effects include fever, bronchospasm, exanthema, angioedema, and hypotension. Rituximab is effective in approximately 50% of refractory CIDP cases, especially when associated with another autoimmune disorder (9).

Another group of modern medications consists of inhibitors targeting individual components of the complement system (eculizumab, ravulizumab, zilucoplan, riliprubart). By blocking complement activation, inflammatory changes in the myelin sheath do not develop, and demyelination is prevented (10).

Another group of modern therapies currently used in autoimmune neurological disorders includes neonatal Fc receptor (FcRn) blockers. Neonatal Fc receptors prolong the half-life of IgG and protect immunoglobulins from lysosomal degradation. Efgartigimod is an example of this therapeutic class (11).

Medicines to manage refractory CIDP should be taken for at least 3 months before switching to the next type of therapy.

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