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Case Report

Chronic inflammatory demyelinating polyneuropathy: case report

Polineuropatia desmielinizante inflamatória crônica: relato de caso

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ABSTRACT: Chronic Inflammatory Demyelinating Polyneuropathy is a progressive, disabling, and rare disease, which requires greater understanding for a better propaedeutic and therapeutic approach, providing a better quality of life to the patient. This case describes a 48-year-old male patient seen in a Basic Health Unit with a complaint of loss of movement of the 4th and 5th fingers of both hands, associated with loss of upper limb strength for two months. The patient was diagnosed with Chronic Inflammatory Demyelinating Polyneuropathy by a specialized service and started treatment with immunoglobulin. He is currently clinically stable and on methylprednisolone. The current treatment is based on the reduction of the inflammatory process and consequent improvement of symptoms. However, there is no robust evidence indicating the better therapeutic regimen among the available options.

KEYWORDS: Polyneuropathies; Guillain-Barre syndrome; Mononeuropathies; Neurology; Medicine.

RESUMO: A Polineuropatia Desmielinizante Inflamatória Crônica é uma doença de caráter progressivo, incapacitante e rara, sendo necessário maior entendimento para melhor abordagem propedêutica e terapêutica, agregando maior qualidade de vida ao paciente. O objetivo deste estudo é relatar o caso de um paciente do sexo masculino, 48 anos com queixa de perda de movimentos de 4º e 5º quirodáctilo bilateral, associado a perda de força de membros superiores há dois meses atendido em Unidade Básica de Saúde. Realizado diagnóstico de Polineuropatia Desmielinizante Inflamatória Crônica em serviço especializado e iniciado tratamento com imunoglobulina. Atualmente encontra-se estável clinicamente com metilprednisolona. O tratamento atual baseia-se na diminuição do processo inflamatório e consequente melhora dos sintomas, entretanto não possui um esquema terapêutico com fortes evidências de superioridade entre as opções disponíveis.

PALAVRAS-CHAVE: Polineuropatias; Síndrome de Guillain-Barré; Mononeuropatias; Neurologia; Medicina.

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INTRODUCTION

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is an autoimmune disease associated with cellular and humoral immune responses that act against peripheral nerve antigens, leading to progressive sensorimotor impairment, affecting the patient's quality of life[1-4].

The disease was first reported in 1969 and was described by the British neurologist Peter Kynaston Thomas[5]. However, CIDP was only accepted as a nosological entity in 1975, in a case report presented by the American neurologist Peter J. Dick[6].

The prevalence of CIDP varies from 1 to 7.7 per 100,000 inhabitants[4,7]. Due to the diversity of its clinical manifestations and the absence of specific markers for the diagnosis, epidemiological studies present different incidence values[1,3,4].

CIDP can also affect children, usually between 5 and 18 years old. In adults, it occurs mainly between the fourth and sixth decade of life[4]. Moreover, males are more affected by the disease[4,7].

Regarding its pathophysiology, CIDP is an autoimmune disease mediated by cellular and humoral immune responses[4,8]. These manifestations attack peripheral nerve antigens, generating a demyelinating process and, consequently, axonal damage[4,8].

In its classic form, the clinical manifestations of CIDP are monophasic, progressive or relapsing, symmetrical, and evolve slowly over a period of two months[4,7,8]. The clinical findings include numbness, paresthesia, muscle weakness, sensory alterations, hyporeflexia or areflexia, fatigue, and balance alterations[7,8]. Cranial nerves may also be involved, with the seventh pair being the most affected[4,7,8].

It is also important to mention that there are other manifestations of the disease[4]. In addition to the classic (most common) form, it can also present as sensory, focal, motor, acute or as the Lewis-Sumner Syndrome[4,8].

The diagnosis of the disease is based on clinical findings, results of electrophysiological studies, analysis of the cerebrospinal fluid, and, in rare cases, biopsy of peripheral nerves[2].

The non-pharmacological treatment is based on physical activity and physical therapy[2,4]. As for the medical treatment, the most common types of treatment are intravenous immunoglobulin, corticosteroids, and plasmapheresis[4,9,10].

From 2016 to the present, only two cases of Chronic Inflammatory Demyelinating Polyneuropathy with no clear cause were found[11,12].

CASE REPORT

A 48-year-old married male patient, from Araçatuba-SP, sought care at a local Basic Health Unit (BHU) in January 2019, reporting loss of movement of the 4th and 5th fingers of both hands, associated with loss of upper limb strength for two months. He had a history of smoking, difficult-to-control insulin-dependent diabetes type 2 for seven years, treated with NPH insulin 40 IU/day, with low adherence to treatment, and hypothyroidism, treated with levothyroxine 50 mcg/day. Laboratory tests for antinuclear antibody, VDRL, rheumatoid factor, and HIV were requested. The patient was referred to the clinical neurology secondary service for specialist assessment.

The patient returned in February 2020 reporting worsened condition, loss of lower limbs strength for 6 months, and a noticeable hyperemia on the left hand for one month. He was referred again to the neurology service and had a chest X-ray, which showed mild posterior spondylosis, and a hand X-ray with no alterations.

The patient returned to the BHU in September, reporting that he had not yet been evaluated by a neurologist. An electroneuromyography (ENMG) of the upper limbs was requested due to paresthesia of the hands, in addition to cranial magnetic resonance. In November 2020, he returned with the result of the ENMG of the upper limbs, which indicated severe demyelinating sensory and motor neuropathy affecting the upper limbs.

In December 2020, the patient went to a neurologist at the medical specialty outpatient clinic, who requested a new ENMG of the upper limbs. The result came in January 2021 and showed severe demyelinating sensory and motor polyneuropathy with signs of denervation of the 1st dorsal interosseous muscles. In view of the test results, the neurologist referred the patient to the specialty outpatient clinic of a hospital in the capital of the state of São Paulo.

The patient was called for consultation with the neurology team of the hospital in the capital of São Paulo in April 2021, and reported further weakening, paresthesia, burning pain, muscle atrophy in the upper limbs, tactile and painful hypoesthesia in the sole of the foot and loss of strength in dorsiflexion and plantar flexion on both feet. At the time, the hypothesis of confluent multiple mononeuropathy, probably associated with vasculopathy secondary to diabetes type 2, was proposed.

A new ENMG was requested, and the result came in April 2021, with the following report: compound muscle action potential (CMAP) and sensory nerve action potential (SNAP) extremely reduced, with reduction in the conduction velocity of some nerves. The ENMG showed reinnervation patterns, mainly in distal muscles, with no signs of irritation, and demyelinating sensory and motor pattern with an associated axonal component. In view of the results, treatment with intravenous immunoglobulin for three months was proposed.

The patient returned in August 2021, without the expected result of the immunoglobulin therapy, and the treatment was augmented with pulse therapy with intravenous methylprednisolone 1g once a day for three

consecutive days. After one week, the same treatment was applied once a week for three weeks and, after that, once a month.

The neurology team of the neurology specialty outpatient clinic requested a cervical MRI in September 2021, to assess complications. The test showed straightening of the cervical spine, uncovertebral arthrosis at C5-C6, posterior disc bulge at C2-C3 and C3-C4 and disc-osteophyte complex at C5-C6 and C6-C7, with an indication of cervical stenosis.

The patient was referred to the neurosurgery team in the specialty outpatient clinic, which, after evaluation, chose not to pursue surgical treatment, as the risks outweighed the benefits.

Currently, the patient is clinically stable, maintaining treatment with methylprednisolone.

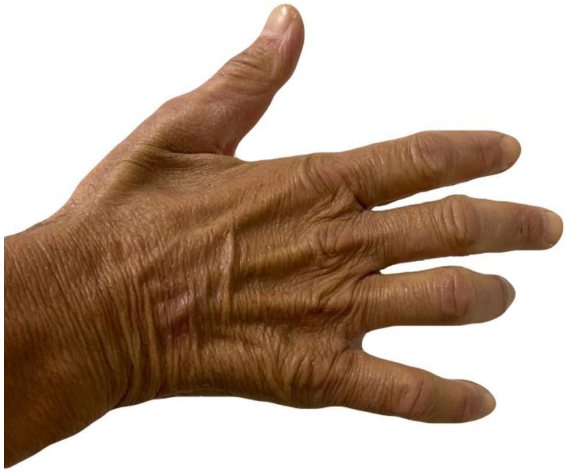

Figure 1 - Right hand showing atrophy of the dorsal interosseous muscles.

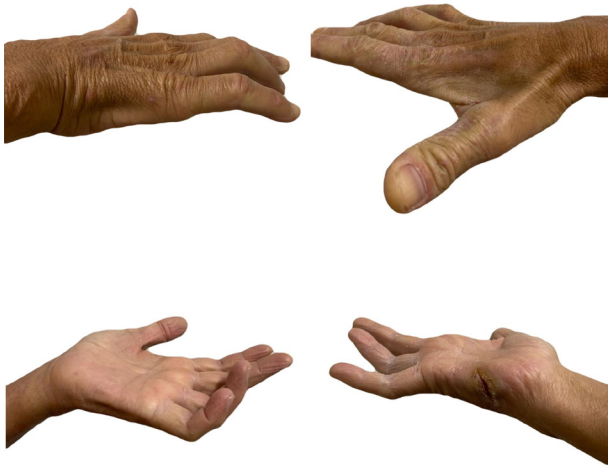

Figure 2 - Atrophy of the dorsal interosseous muscles of the hand. A: Pronation of the right hand. B: Pronation of the left hand. C: Supination of right hand. D: Supination of the left hand.

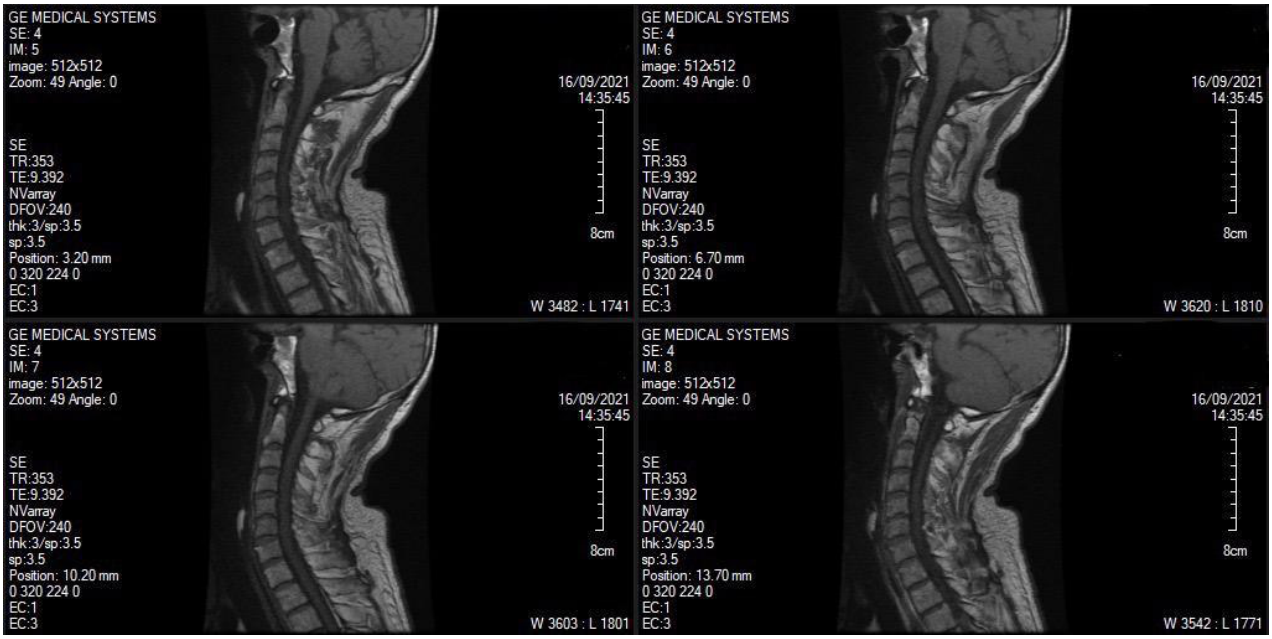

Figure 3 - Cervical MRI with cervical stenosis.

DISCUSSION

CIDP is an autoimmune disease described for the first time in 1969 and characterized by a progressive loss of sensorimotor function[1,2]. Epidemiologically, the incidence is higher in men between 40 and 60 years old, and the case presented corresponds to a 48-year-old patient[3,4].

CIDP is associated with systemic clinical factors in 10% to 20% of cases, of which the most common are infections associated with the human immunodeficiency virus, hepatitis B and hepatitis C[2]. In the case presented, the patient has diabetes mellitus without proper treatment and hypothyroidism.

Among the several types of CIDP, the most common is the typical presentation, which corresponds to 50% of the cases and is characterized by chronicity and sensory and motor symptoms[2,4]. However, it can also present as only motor, only sensory, only focal, acute, or multifocal demyelinating sensory and motor neuropathy, also known as Lewis-Summer Syndrome[2,4]. The patient in this case presents the typical form, as he had both sensory and motor symptoms.

The predominant motor symptom is symmetrical weakness of the upper and lower limbs, present in 90% of the cases[2,4]. Complete areflexia occurs in 70% of patients, mainly in the ankle jerk reflex[2,4]. On the other hand, pain is rare symptom in CIDP, and cranial nerves are rarely involved[4]. However, the patient in this case presented pain with burning sensation, with no alteration on cranial nerve examination.

CIDP is monophasic, progressive, relapsing, and symmetrical and evolves gradually over 2 months[2,3,4]. It has a high risk of disability, as up to 50% of patients may present severe temporary disabilities and up to 10% will have persistent and progressive disabilities[2,4].

One third of the cases are remitting and chronic, as the case presented, in which the symptoms appeared more frequently and more intensely[2,3,4]. On the other hand, two thirds of patients have a cyclic course, with episodes of progression and remission[2].

The main differential diagnosis of CIDP is Acute Inflammatory Demyelinating Polyneuropathy (AIDP), represented by Guillain-Barre Syndrome (GBS)[13-14].

Differentiating between them is challenging at the beginning of symptoms, and the longitudinal follow-up of the patient is important. It is known that the AIDP course usually has a clinical nadir within four weeks, while CIDP demonstrates gradual worsening or relapse of symptoms over more than an 8-week period[13-14].

The treatment of CIDP is based on the reduction of inflammation and consequent reduction of symptoms. The most common treatments are corticosteroid therapy, plasmapheresis, and intravenous immunoglobulin[2,4,13-15].

Corticosteroid therapy is an initial option for the treatment, both because of easier access and lower cost, but there are uncertainties regarding the method of use[15]. Evidence shows no difference between high-dose monthly oral dexamethasone for six months and low-dose daily prednisolone, but the latter had greater adverse effects[15].

Plasmapheresis and intravenous immunoglobulin show short-term improvement in symptoms and, consequently, in disability, but evidence does not show any difference between them[15].

CONCLUSION

CIDP is a progressive, debilitating, and rare autoimmune disease that affects mainly adult men. It has a complex pathophysiology and no specific diagnostic criteria. Its treatment is based on immunoglobulins, corticosteroids, and plasmapheresis, but further studies are still required to better elucidate its management. This study contributed to the scientific literature, as it presented a rare condition that is little reported worldwide and considering that it is essential that health professionals have knowledge of the disease, so that they can manage similar cases in the best possible way.

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