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Polineuropatía crónica inflamatoria desmielinizante: caso clínico

Chronic inflammatory demyelinating polyneuropathy: clinical case

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Palabras claves:
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Resumen

Introducción: la Polineuropatía crónica inflamatoria desmielinizante es un raro trastorno autoinmune que afecta al sistema nervioso periférico, el diagnóstico, variabilidad en la evolución y respuesta al tratamiento son desafíos clave. Además, el alto costo de tratamientos y el impacto en la calidad de vida subrayan la importancia de investigar esta condición. **Objetivo:** analizar el manejo de la patología, incluyendo su descripción, plan de atención y tratamiento, mediante estudio de un caso clínico. **Metodología:** Caso clínico con revisión bibliográfica tipo descriptivo, retrospectivo, se abordan aspectos como definición, fisiopatología, factores de riesgo, diagnóstico, pronóstico, signos y síntomas. La sistematización del caso incluye motivo de consulta, enfermedad actual, antecedentes, medicamentos, examen físico, laboratorios y desenlace. La discusión destaca influencias holísticas en el desenlace, comparando datos con otras investigaciones para enriquecer el artículo. **Resultados:** Se considera el caso de un hombre de 41 años con múltiples condiciones médicas preexistentes, incluyendo hipotiroidismo, hipertensión arterial, trombosis venosa profunda y tromboembolismo pulmonar. Ingresa a emergencia con síntomas compatibles con síndrome coronario agudo, pero se descartan signos cardíacos significativos en los estudios. Dada su historia de trastornos previos y una polineuropatía inflamatoria, se sospecha un síndrome multiinflamatorio sistémico relacionado con un posible síndrome de hipercoagulabilidad. Se inicia tratamiento y se realiza una exhaustiva evaluación clínica, destacando la presencia de estenosis de canal medular y evidencia de polineuropatía desmielinizante crónica inflamatoria (CIDP) en estudios complementarios. **Conclusiones:** La Polineuropatía Desmielinizante Crónica Inflamatoria (CIDP) es un desafío en los trastornos autoinmunes del sistema nervioso periférico, evidenciando complejidades en diagnóstico, evolución y tratamiento. El estudio de un caso clínico subraya la importancia de un enfoque integral que incluye medicamentos, control vital, fisioterapia y seguimiento neurológico. La investigación destaca la necesidad de más estudios y atención especializada

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para abordar eficazmente la CIDP. **Área de estudio general:** medicina. **Área de estudio específica:** cardiología. **Tipo de estudio:** Casos clínicos.

Keywords:
Chronic Inflammatory Demyelinating Polyneuropathy (CIDP), Peripheral Nervous System, Autoimmune Disorders, Specialized Clinical Care

Abstract

Introduction. Chronic inflammatory demyelinating polyneuropathy is a rare autoimmune disorder affecting the peripheral nervous system, diagnosis, variability in evolution and response to treatment are key challenges. In addition, the high cost of treatment, together with the impact on quality of life, underlines the importance of investigating this condition.**Objective:**to analyze the management of the pathology, including its description, care plan and treatment, by means of a clinical case study.**Methodology:**clinical case with bibliographic review, descriptive, retrospective type, aspects such as definition, physiopathology, risk factors, diagnosis, prognosis, signs and symptoms are approached. The systematization of the case includes reason for consultation, current illness, history, medications, physical examination, laboratories and outcome. The discussion highlights holistic influences on the outcome, comparing data with other investigations to enrich the article.**Results:**We present the case of a 41-year-old man with multiple pre-existing medical conditions, including hypothyroidism, arterial hypertension, deep vein thrombosis and pulmonary thromboembolism. He is admitted to the emergency department with symptoms compatible with acute coronary syndrome, but significant cardiac signs are ruled out on workup. Given her history of previous disorders and an inflammatory polyneuropathy, a systemic multi-inflammatory syndrome related to a possible hypercoagulability syndrome is suspected. Treatment is initiated and a thorough clinical evaluation is performed, highlighting the presence of spinal canal stenosis and evidence of chronic inflammatory demyelinating polyneuropathy (CIDP) in complementary studies.**Conclusions:**Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is a challenge in autoimmune disorders of the peripheral nervous system, evidencing complexities in diagnosis, evolution and treatment. A clinical case study highlights the importance of a comprehensive

approach that includes medications, vital management, physical therapy, and neurological follow-up. The research highlights the need for further studies and specialized care to effectively address CIDP.

Introduction

Chronic inflammatory demyelinating polyneuropathy is a rare autoimmune disorder affecting the peripheral nervous system, causing inflammation and damage to the myelin sheath. It causes muscle weakness, numbness, tingling, and fatigue.(1). One of the main challenges lies in the diagnosis, variability in evolution, response to treatment(2),High cost of treatment, since both intravenous immunoglobulin (IVIG) and corticosteroids, which are frequently used, are expensive and require periodic administration(3); additionally it affects the quality of life and causes severe disability(4).

Exploration in the field of autoimmune disorders, such as chronic inflammatory demyelinating polyneuropathy (CIDP), is of utmost importance due to the similarities it shares with other diseases such as multiple sclerosis and lupus. By exploring the pathology of CIDP, one seeks to gain detailed insights into the underlying mechanisms that trigger these diseases, allowing for a more complete understanding of their pathophysiology. Identifying the specific immunological processes involved in CIDP and comparing them to other autoimmune disorders may shed light on the common and distinctive factors that contribute to the development of these diseases. This, in turn, could open new perspectives for the design of more targeted and effective treatments that address the immunological mechanisms responsible for CIDP and other related autoimmune conditions. (5).

During the 1980s, significant progress was made in the interpretation of the pathophysiology of chronic inflammatory demyelinating polyneuropathy. Detailed studies were carried out to identify the underlying immunological mechanisms that contribute to chronic demyelination and inflammation in the peripheral nervous system, allowing CIDP to be recognized as a unique entity with distinctive features, gradually separating it from Guillain-Barré syndrome. In the following decade, during the 1990s, substantial progress was made in the diagnosis of CIDP, establishing more precise criteria that allowed health professionals to more reliably identify this disease, even in mild or atypical presentations. Improvements in diagnostic techniques, such as

neurophysiology and imaging tests, contributed to a more accurate identification of CIDP.(6).

The incidence and prevalence are difficult to determine due to the variability of presentation and lack of a definitive diagnostic test. It is estimated that it affects between 1 and 8 of every 100,000 people worldwide, and can affect people of any age, although it is common between 50 and 60 years of age. It affects both sexes equally and there is no known racial or ethnic predisposition to the disease.(7).

The exact source is unknown, but it is believed to be caused by immune dysfunction towards the myelin sheath, with the presence of inflammatory cells and antibodies, which contribute to axonal destruction and damage.(8), and genetic actors that are associated with increased risk, including HLA-DRB1, PTPN22 and CTLA-4(9)Risk factors include autoimmune disorders, HIV, hepatitis C, Lyme disease, exposure to heavy metals(10); pathologies such as diabetes, which weakens the immune system, another involved to a lesser extent is monoclonal gammopathy of uncertain significance(11).

It is characterized by a variation in the intensity and duration of its symptoms. Affected patients predominantly experience muscle weakness and decreased sensitivity in the extremities, either symmetrically or asymmetrically. In addition to motor and sensory impairment, CIDP can have an impact on balance, which can result in frequent falls.(12). Other common symptoms include persistent fatigue, muscle cramps, and difficulties in performing fine tasks. This variability in clinical presentation makes CIDP a heterogeneous disorder, and the diversity of symptoms can affect patients' quality of life, affecting their mobility and ability to carry out daily activities. Early recognition of these signs and symptoms is crucial for an accurate diagnosis and effective management of the disease.(13).

Variants may occur, the most common of which are: distal or distal acquired symmetrical demyelinating neuropathy, “multifocal demyelinating neuropathy” or “Lewis-Sumner syndrome”(14); less frequent: focal, affects the brachial or lumbosacral plexus(15), motor with symmetrical proximal and distal weakness, and sensory with gait ataxia,(16)It can affect the autonomic nervous system, with different changes in blood pressure, heart rate, bowel and bladder function. It can be associated with autoimmune disorders, such as systemic lupus erythematosus, rheumatoid arthritis or Sjogren's syndrome.(17).

Diagnosing chronic inflammatory demyelinating polyneuropathy (CIDP) can be a lengthy process because it has similarities to other conditions and lacks a definitive test. During the clinical evaluation, signs of muscle weakness, reduced reflexes, and sensory abnormalities are sought, while medical history, recent infections, vaccination history,

and exposure to possible toxins are inquired about.(18)Diagnostic tests include nerve conduction studies, electromyography, antibody detection, and inflammatory markers. In some cases, a nerve biopsy may be performed, and supervision by a specialist in autoimmune disorders of the nervous system is essential. The combination of these approaches contributes to a more accurate diagnosis, although the complexity and variability of CIDP underscore the importance of a multidisciplinary approach to the diagnostic process.(19, 20).

Treatment of chronic inflammatory demyelinating polyneuropathy (CIDP) is usually started with corticosteroids, such as prednisone, being the first line of therapy.(21)In cases of poor response, alternatives such as the use of methylprednisolone for a period of four weeks are used, and oral steroids such as dexamethasone or prednisone are considered.(22)For more severe cases, plasma exchange is used to remove harmful antibodies, and intravenous immunoglobulin is presented as another effective option.(23)In situations where the response to initial treatment is unsatisfactory or in refractory cases, immunosuppressants such as methotrexate or cyclophosphamide are used.(24)Furthermore, the use of Rituximab stands out as another important option in the CIDP therapeutic armamentarium. The variety of therapeutic options highlights the importance of an individualized approach and the need to consider the patient's response to each modality to optimize treatment outcomes.(25).

The prognosis of chronic inflammatory demyelinating polyneuropathy (CIDP) is closely linked to early diagnosis, disease severity, and response to treatment. Approximately 60% to 80% of patients respond positively to immunosuppressive drugs or intravenous immunoglobulin, which can slow or stop disease progression and even reverse nerve damage in some cases.(26)However, in cases where there is no response, a continued worsening of symptoms is observed, increasing the risk of disability and decreasing quality of life. In these situations, ongoing treatment and periodic assessments are required to monitor symptoms and adjust the therapeutic approach as needed.(27).

Based on the above, the main purpose will be to observe the management of the pathology in order to identify novel aspects. The specific objectives include a detailed description of the disease, as well as planning nursing care and medical treatment. The aim is to systematize the information from the clinical case. Finally, the findings from the clinical case will be compared with previous research in a discussion.

Methodology

This article presents a descriptive and retrospective clinical case report on sinoatrial nodal block disease. It also includes a compilation of literature. The medical history was reviewed and analyzed to obtain information about the case. Vancouver style was used for writing, and the following structure was used to describe the pathology: definition of

the pathology, pathophysiology, risk factors, diagnosis, prognosis, signs and symptoms, consequences, nursing care plan, and medical treatment. Scientific databases such as Scopus, Porquest, Pubmed, Web of Science, and Lilacs were used to compile literature on the subject.

Clinical case data were systematically collected from the patient's medical history (secondary database), which included the reason for consultation, the patient's current illness upon admission, the diagnostic impression (IDX), personal and family history, medications the patient regularly uses, physical examination, the first laboratory tests taken from the patient, the therapeutic management plan, complementary tests, and the outcome.

In the last section, knowledge about the characteristics that comprehensively affect patient outcome was discussed and synthesized. Finally, data were compared with those from other research to organize the clinical case article.

Results

Case presentation

A 41-year-old male patient with a history of hypothyroidism, arterial hypertension, deep vein thrombosis and pulmonary thromboembolism, goes to the emergency room presenting dyspnea, diaphoresis, pain in the cervical area, with radiation to the shoulders and retrosternal region, is diagnosed with acute coronary syndrome, for which treatment is started and is admitted to the internal medicine service, laboratory tests are performed where negative troponins and an electrocardiogram without corresponding signs are evident, due to a history of pulmonary thromboembolism, deep vein thrombosis and inflammatory polyneuropathy, systemic multiinflammatory syndrome associated with hypercoagulability syndrome is suspected.

Discussion

The case presented is a 41-year-old male patient with a significant medical history who presents to the emergency room with symptoms suggestive of acute coronary syndrome, ruled out by negative troponins and electrocardiogram. Based on his medical history, a systemic multiinflammatory syndrome associated with hypercoagulability syndrome is suspected.

The patient was placed on a treatment regimen that included several medications commonly used to address his medical conditions. These included rivaroxaban, an oral anticoagulant used to prevent blood clots; furosemide, a diuretic used to control fluid retention; irbesartan, an angiotensin II receptor antagonist used to treat hypertension;

and levothyroxine, a synthetic thyroid hormone that helps regulate thyroid function in hypothyroidism.

Physical examination revealed the presence of tenderness on acupressure in the anterior chest area, suggesting tenderness in that region. In addition, decreased strength is noted in all four limbs, indicating possible neuromuscular or motor function-related impairments. Pain in the lower limbs adds another layer of complexity to the clinical picture, highlighting the need for further evaluation to determine the underlying cause of these symptoms.

Laboratory data showed abnormal values in leukocytes, erythrocytes, hemoglobin, hematocrit and platelets, as well as elevated levels of erythropoietin, regarding this Marques et al, highlight the elevation of erythropoietin as a compensatory response to counteract the negative effects of anemia induced by inflammation, when the production of proinflammatory cytokines stimulates the expression of the liver hormone hepcidin that promotes iron retention, in response to this, the body increases the production of erythropoietin to stimulate the production of red blood cells and compensate for anemia.(28).

Complementary examinations include magnetic resonance imaging of the lumbar spinal canal, bone scintigraphy, and sensory-motor neuroconduction studies and electromyography, which reveal spinal canal stenosis, joint inflammatory process, and findings suggestive of motor and sensory polyneuropathy of a secondary axonal demyelinating type. Fisse et al mention the limitation of nerve conduction studies to reproduce clinical dynamics, and the use of electromyography for monitoring CIDP, however, they highlight various clinical scores, such as the Medical Research Council (MRC) Summary Score and the INCAT Global Disability Summary Score (ODSS) as standard tools to assess muscle strength and disability in patients with CIDP.(29).

In therapeutic management, the administration of methylprednisolone, enoxaparin for coagulation, clopidogrel for antiplatelet aggregation, and other medications to address the various clinical aspects present in the patient stand out, the growing shortage of IVIg in many countries, which is why the administration of methylprednisolone is highlighted as part of therapeutic management, highlighting the importance of optimizing treatment algorithms and seeking alternatives when necessary.

In a comparable case, in 2023, Segura et al. investigated a case of a 38-year-old patient who experienced paresthesias, progressive weakness in all four limbs, and diplopia and dysphagia for eight months. The patient presented with symmetrical flaccid quadriparesis with distal predominance, atrophy in the dorsum and palm of both hands, generalized areflexia, postural tremor in the upper limbs with left predominance, appendicular dysmetria, dysdiadochokinesia, ophthalmoparesis on dextroversion of the

right eye, absent gag reflex, ataxic gait with increased base of support, and positive Romberg sign. Electromyography results revealed a predominantly demyelinating sensorimotor polyneuropathy, while cerebrospinal fluid results showed dissociated albuminocytology. Anti-neurofascin antibody testing was performed upon clinical suspicion.(30).

For his part, in the study conducted by Al, 2023. He states that the cumulative mortality rate was 3.3% (95% confidence interval [CI], 1.9% to 5.7%). The cumulative fraction of non-ambulatory patients was 8.2% (95% CI, 5.7% to 11.6%) and overall, 47.1% (95% CI, 39.5% to 54.9%) of CIDP patients had a good outcome without disability. The cumulative remission rate was 40.8% (95% CI, 30.6% to 51.8%). Noting that future research is required on how to prevent long-term deterioration in CIDP. Care should be taken in developing clinical strategies to avoid immunomodulatory therapy in many patients in remission.(31).

In a similar case, in 2023, Segura et al. examined a case of a 38-year-old patient who experienced paresthesias and progressive weakness in all four extremities for eight months, as well as diplopia and dysphagia. The patient presented with symmetrical flaccid quadriparesis with distal predominance, atrophy in the dorsum and palm of both hands, generalized areflexia, postural tremor in the upper extremities with left predominance, appendicular dysmetria, dysdiadochokinesia, ophthalmoparesis on dextroversion of the right eye, absent gag reflex, ataxic gait with increased base of support, and positive Romberg sign. Electromyography demonstrated a predominantly demyelinating sensorimotor polyneuropathy, and cerebrospinal fluid results showed albuminocytological dissociation. Anti-neurofascin antibody testing was performed upon clinical suspicion. Despite an improvement in diplopia and dysphagia, no effect was observed on limb strength, and functional deterioration was even recorded. For this reason, treatment with rituximab was started at a dose of two grams, achieving a substantial improvement in distal muscle strength, tremor, gait stability, coordination and functionality, measured by the modified Rankin scale.(32).

In a similar case, in 2023, Segura et al. investigated a case of a 38-year-old patient who experienced paresthesias, progressive weakness in all four limbs, and diplopia and dysphagia for eight months. The patient presented with symmetrical flaccid quadriparesis with distal predominance, atrophy in the dorsum and palm of both hands, generalized areflexia, postural tremor in the upper limbs with left predominance, appendicular dysmetria, dysdiadochokinesia, ophthalmoparesis on dextroversion of the right eye, absent gag reflex, ataxic gait with increased base of support, and positive Romberg sign. Electromyography findings indicated a predominantly demyelinating sensorimotor polyneuropathy, and cerebrospinal fluid findings indicated albuminocytology dissociation. Based on clinical suspicion, anti-neurofascin antibody

testing was performed. Laboratory tests were normal. Electrodiagnostic studies showed diffuse motor and sensory dysfunction in all extremities; consequently, a diagnosis of CIDP was made. Imaging studies revealed a large hepatic mass in the left lobe of the liver. Subsequent biopsy showed histopathological findings characteristic of hepatocellular carcinoma. After failure of medical treatment with intravenous immunoglobulin and corticosteroids, laparoscopic resection of the tumor was planned and carried out, resulting in complete resolution of symptoms. At 18 months postoperatively, the patient was asymptomatic.(33).

Rostovtseva et al. (34), chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare disease characterized by heterogeneous clinical findings, absence of specific laboratory markers, and good response to treatment. Typically, CIDP manifests as weakness of varying severity, from minimal paresis to paralysis, as well as symmetrical loss of sensitivity in all limbs. A distinctive clinical feature of CIDP is the involvement of both proximal and distal parts of the limbs. At the same time, there are a large number of atypical variants of CIDP, clinically similar to other chronic immunologic neuropathies. Neuroimaging findings typically include involvement of the cauda equina, brachial plexus, and lumbar plexus.(34).

In the same context, Ferraz et al. (35) describes a case of a 42-year-old woman with poorly controlled diabetes who presented with a nine-month evolution of ataxic gait, decreased motor and sensory function in the upper and lower extremities, and postural anesthesia in the fingers. Deep tendon reflexes were abolished in the lower extremities and markedly decreased in the upper extremities. The study of cerebrospinal fluid (CSF) showed a high level of proteins, and both imaging and serological studies were normal. Although she had a previous electrophysiological study showing a distal symmetrical polyneuropathy (DSPN) with axonal injury, nerve conduction studies were repeated and a chronic inflammatory demyelinating polyneuroradiculopathy (CIDP) was diagnosed. According to the latest scientific evidence, the administration of intravenous immunoglobulin (IVIg) was started. The Inflammatory Neuropathy Cause and Treatment (INCAT) score and the Medical Research Council (MRC) summed score improved after two cycles. Unfortunately, symptoms recurred rapidly and corticosteroids were introduced to try to delay recurrence of symptoms, although they worsened diabetes control. This case highlights the diversity and complexity of inflammatory polyneuropathies, as well as the variability in therapeutic responses.(35)Both cases present challenges in the diagnosis and management of clinical conditions involving multiple neurological and medical systems.

In addition to the above, it is noted that according to Rajabally (36), chronic inflammatory demyelinating polyneuropathy is a well-described clinical entity with typical and variant forms, among which the distal form stands out, being very rare.

Clinically, distal CIDP may resemble M-glycoprotein (MAG)-associated neuropathy, which is not considered part of the CIDP spectrum. Anti-MAG neuropathy is characterized by a slowly progressive clinical course, predominantly sensory features, ataxia, and tremor. Although typical CIDP is clinically distinguished from distal CIDP or anti-MAG neuropathy, some cases with the presence of anti-MAG antibodies have been recently described.(36).

Also Suponeva et al. (37) agree that chronic inflammatory demyelinating polyneuropathy (CIDP) is a treatable dissimilar neuropathy. The variety of clinical forms and the evolution of the disease can make adequate diagnosis and early treatment difficult. In a quarter of cases, CIDP begins acutely, mimicking Guillain-Barré syndrome. Early diagnosis is especially important due to the differences in treatment and prognosis of these conditions. In this article, we present a clinical case of acute onset of CIDP with a detailed analysis of the differential diagnosis between acute and chronic immune-mediated neuropathies.(37).

In the same line, Gogia et al. (38) points out that chronic inflammatory demyelinating polyradiculoneuropathies (CIDP) are a type of acquired immune-mediated disorder that affects the peripheral nervous system. Although it presents diverse clinical manifestations, the classic presentation includes symmetrical involvement of both sensory and motor in the proximal and distal regions. CIDP can have a monophasic, recurrent or progressive course, developing over a period of more than eight weeks. The 8-week course, as well as the duration to reach the nadir, help distinguish CIDP from Guillain-Barré syndrome (GBS) or other acute inflammatory demyelinating polyneuropathies (AIDP). The first case was described by Eichhorst Burns in 1890. Approximately 16% of patients present an acute picture of GBS.(38).

In this regard, Allen & Lewis (39) describe that, since the introduction of CIDP in the 1970s, more than 15 sets of diagnostic criteria have been proposed, with those published in 2021 by the European Academy of Neurology/Peripheral Nerves Society (EAN/PNS) being especially relevant to clinical practice. These criteria offer physicians a valuable tool to interpret the data collected during the evaluation of patients with possible CIDP symptoms. The correlation between diagnosis and treatment highlights the importance of objectifying the response to treatment, and although no approach is perfect, the use of weakness and disability results in clinical practice can help clarify the difficulties in interpreting the response to treatment. The absence of objective benefit should be considered when reconsidering the diagnosis of CIDP.(391).

In contrast, Malik et al. (40) describe the case of a young woman diagnosed with chronic inflammatory demyelinating polyneuropathy (CIDP). Treatment with intravenous immunoglobulins and prednisone did not improve her condition, and her neurological symptoms worsened. She was subsequently readmitted with dyspnea on

exertion, dizziness, malaise, and black stools. Colonoscopy revealed a necrotic mass in the ascending colon directly invading the second part of the duodenum. Pathological results confirmed the diagnosis of a locally advanced CRC. Following surgical resection of the cancer, her CIDP showed dramatic resolution without the need for additional therapy. Patients with CRC may develop CIDP as a type of paraneoplastic syndrome. Health care workers should be aware of this possible association, as it is of utmost importance for the necessary comprehensive clinical management.(40).

This research presents a case of a 41-year-old man with various pre-existing medical conditions, admitted to the emergency room with symptoms suggestive of acute coronary syndrome. Despite ruling out significant cardiac signs in the studies, his medical history and the presence of inflammatory polyneuropathy suggest a systemic multi-inflammatory syndrome related to a possible hypercoagulability syndrome. Treatment includes medications, monitoring of vital signs, physical therapy, and neurological follow-up to address these complex comorbidities, highlighting the presence of spinal canal stenosis and evidence of CIDP in complementary studies. In contrast, Chauvet, et al. 2022, emphasize that a subset of adult patients with treatment-resistant CIDP have been observed to present antibodies against paranodal proteins, however in the case of a pediatric patient, a 14-year-old adolescent with a severe form of CIDP in whom positive antibodies against neurofascin 155 were found in his serum. Resistant to conventional therapies, he experienced dramatic improvement with Rituximab treatment, showing mild to moderate functional motor disability at 24-month follow-up. In pediatric patients with CIDP who remain refractory to conventional treatments(41).

Based on the above, it is concluded that, Chronic inflammatory demyelinating polyneuropathy (CIDP) represents a chronic form of immune-influenced peripheral polyneuropathy. The lack of reliable diagnostic biomarkers makes the identification of CIDP difficult, making its diagnosis challenging. The lack of early recognition in many patients underlines the importance of establishing an accurate and timely diagnosis, as well as avoiding diagnostic errors and unnecessary treatments. Identifying the distinctive clinical, electrophysiological and laboratory manifestations of the disease is essential to facilitate a rapid diagnosis, while understanding potential diagnostic pitfalls helps prevent errors.

This pathology remains a challenging area in the field of autoimmune disorders affecting the peripheral nervous system. The variety in clinical presentation, the difficulty in diagnosis and the variability in response to treatment have been highlighted in numerous studies and, in particular, in the review of the clinical case presented. The lack of specific biomarkers for the diagnosis of CIDP contributes to the challenges in its early identification, often leading to delays in the initiation of treatment.

The comparison between the clinical case and the existing literature highlights the importance of a multidisciplinary approach in the management of CIDP. The integration of medications, vital control, physical therapy and neurological monitoring has proven to be essential to address the complexities associated with this disease. The clinical case with multiple pre-existing medical conditions underlines the need for a thorough evaluation that considers the patient's complete medical history.

Conclusions

- Extensive research on Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) has significantly deepened our understanding of this autoimmune disease impacting the peripheral nervous system. CIDP, manifesting substantial challenges in terms of diagnosis, variability in course, and response to treatment, underscores the intrinsic complexity of this pathology. A detailed analysis of a specific clinical case, in particular the management of a 41-year-old man with multiple pre-existing medical conditions, has highlighted the imperative need for a comprehensive approach to effectively address CIDP.
- The systematization of the information derived from the clinical case has allowed us to identify novel aspects in the treatment of CIDP, highlighting the crucial importance of multidisciplinary approaches that encompass medications, vital monitoring, physiotherapy and neurological follow-up. The comparison of the findings with previous research has enriched the contextualization of the clinical case, revealing similarities and differences that can guide future therapeutic approaches.
- The high cost of treatments and their impact on lifestyle highlight the urgency of further research to develop more effective and accessible strategies. Ultimately, this research underscores the need for further studies and specialized clinical care to effectively address CIDP, thereby improving the quality of life of users affected by this rare autoimmune disease.

Conflict of interest

The authors declare that there is no conflict of interest in relation to the submitted article.

Authors' contribution statement

Author 1: I actively participated in the planning and design of the literature review. I also carried out a critical evaluation of the selected studies, analyzing both the methodological quality and the validity of the results.

Author 2: Significantly contributed to the interpretation and discussion of the findings obtained in the clinical case. He/she also played an important role in the writing and revision of the content of the manuscript.

Author 3: Provided valuable input by providing comments that improved the clarity and coherence of the work. Actively participated in the development of the results and conclusions of the study.

Bibliographic References

1. Darbà J, Marsà A. Chronic inflammatory demyelinating polyneuropathy in Spain: a retrospective analysis of hospital incidence and medical costs. https://doi.org/101080/1473716720222000862 [Internet]. November 1, 2021 [cited April 25, 2023];22(4):665–70.Available in:https://www.informahealthcare.com/doi/abs/10.1080/14737167.2022.2000862

2. Kanda T, Iijima M, Sobue G. [A Review of the History of Treatment: From Symptom Improvement to Maintenance Therapy- with a Focus on SCIg].Brain Nerve [Internet]. July 2021 [cited 25 April 2023];73(7):819–28. Available at:https://pubmed.ncbi.nlm.nih.gov/34234040/

3. Divino V, Mallick R, DeKoven M, et al.The economic burden in the United States: A case-control study. PLoS One [Internet]. October 1, 2018 [cited April 25, 2023];13(10). Available at:https://pubmed.ncbi.nlm.nih.gov/30352101/

4. Querol L, Hartung H, Lewis R, et al.The Role of the Complement System in Chronic Inflammatory Demyelinating Polyneuropathy: Implications for Complement-Targeted Therapies. Neurotherapeutics [Internet]. 2022 Apr 1 [cited 2023 Apr 25];19(3):864–73. Available in:https://pubmed.ncbi.nlm.nih.gov/35378684/

5. Vallat J, Mathis S, Vegezzi E, et al.Antibody- and macrophage-mediated segmental demyelination in chronic inflammatory demyelinating polyneuropathy: clinical, electrophysiological, immunological and pathological correlates. Eur J Neurol [Internet]. 2020 Apr 1 [cited 2023 Apr 25];27(4):692–701. Available in:https://pubmed.ncbi.nlm.nih.gov/31769579/

6. Dyck P, Tracy J. History, Diagnosis, and Management of Chronic Inflammatory Demyelinating Polyradiculoneuropathy.Mayo Clin Proc [Internet]. 2018 Jun 1 [cited 2023 Apr 25];93(6):777–93. Available at:https://pubmed.ncbi.nlm.nih.gov/29866282/

7. Broers MC, Bunschoten C, Nieboer D, et al.Incidence and Prevalence of Chronic Inflammatory Demyelinating Polyradiculoneuropathy: A Systematic Review and Meta-Analysis. Neuroepidemiology [Internet]. 2019-01-22 [cited 2023-04-25];52(3–4):161. Available at:https://pubmed.ncbi.nlm.nih.gov/30669140/

8. Stino A, Naddaf E, Dyck P, et al.Chronic inflammatory demyelinating polyradiculoneuropathy-Diagnostic pitfalls and treatment approach. Muscle Nerve [Internet]. February 1, 2021 [cited April 25, 2023];63(2):157–69. Available at:https://pubmed.ncbi.nlm.nih.gov/32914902/

9. Lehmann H, Burke D, Kuwabara S. Chronic inflammatory demyelinating polyneuropathy: update on diagnosis, immunopathogenesis and treatment.J Neurol Neurosurg Psychiatry [Internet]. 2019 Sep 1 [cited 2023 Apr 25];90(9):981–7. Available at:https://pubmed.ncbi.nlm.nih.gov/30992333/

10. Doneddu PE, Bianchi E, Cocito D, et al.Risk factors for chronic inflammatory demyelinating polyradiculoneuropathy: antecedent events, lifestyle and dietary habits. Data from the Italian CIDP Database. Eur J Neurol [Internet]. 2020 Jan 1 [cited 2023 Apr 25];27(1):136–43. Available in:https://pubmed.ncbi.nlm.nih.gov/31325350/

11. Doneddu P, Cocito D, Manganelli F, et al.Frequency of diabetes and other comorbidities in chronic inflammatory demyelinating polyradiculoneuropathy and their impact on clinical presentation and response to therapy. J Neurol Neurosurg Psychiatry [Internet]. 2020-10-1 [cited 2023-04-25];91(10):1092–9. Available at: https://pubmed.ncbi.nlm.nih.gov/32868387/

12. Rodríguez Y, Vatti N, Ramíres C. Chronic inflammatory demyelinating polyneuropathy as an autoimmune disease.J Autoimmun [Internet]. August 1, 2019 [cited April 25, 2023]; 102:8–37. Available at:https://pubmed.ncbi.nlm.nih.gov/31072742/

13. Franques J. [Chronic inflammatory demyelinating polyneuropathy: Diagnosis and therapeutic update].Rev Med Interne [Internet]. 2019 Dec 1 [cited 2023 Apr 25];40(12):808–15. Available at:https://pubmed.ncbi.nlm.nih.gov/31677862/

14. Van den Bergh P, van Doorn P, Hadden R, et al.European Academy of Neurology/Peripheral Nerve Society guideline on diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy: Report of a joint Task Force—Second review. Journal of the Peripheral Nervous System [Internet]. 2021 Sep 1 [cited 2023 Apr 25];26(3):242–68. Available in:https://onlinelibrary.wiley.com/doi/full/10.1111/jns.12455

15. Benoit C, Svahn J, Debs R, et al.Focal chronic inflammatory demyelinating polyradiculoneuropathy: Onset, course, and distinct features. J Peripher Nerv Syst [Internet]. June 1, 2021 [cited April 25, 2023];26(2):193–201. Available at:https://pubmed.ncbi.nlm.nih.gov/33660880/

16. Ikeda S, Koike H, Nishi R, et al. Clinicopathological characteristics of subtypes of chronic inflammatory demyelinating polyradiculoneuropathy.J Neurol Neurosurg Psychiatry [Internet]. 2019 Sep 1 [cited 2023 Apr 25];90(9):988–96. Available at:https://pubmed.ncbi.nlm.nih.gov/31227562/

17. Querol L, Crabtree M, Herepath M, et al.Systematic literature review of burden of illness in chronic inflammatory demyelinating polyneuropathy. J Neurol [Internet]. October 1, 2021 [cited April 25, 2023];268(10):3706–16. Available at:https://pubmed.ncbi.nlm.nih.gov/32583051/

18. Bunschoten C, Jacobs B, Van den Bergh P, et al.Progress in diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy. Lancet Neurol [Internet]. August 1, 2019 [cited April 25, 2023];18(8):784–94. Available at:https://pubmed.ncbi.nlm.nih.gov/31076244/

19. Doneddu P, Cocito D, Manganelli F, et al.Atypical: diagnostic criteria, progression and treatment response. Data from the Italian Database. J Neurol Neurosurg Psychiatry [Internet]. 2019-02-01 [cited 2023-04-25];90(2):125–32. Available at:https://pubmed.ncbi.nlm.nih.gov/30297520/

20. Kuwabara S, Misawa S, Mori M. Atypical chronic inflammatory demyelinating polyneuropathies.J Neurol Neurosurg Psychiatry [Internet]. 2019 Feb 1 [cited 2023 Apr 25];90(2):121. Available at:https://pubmed.ncbi.nlm.nih.gov/30297522/

21. Svačina M, Lehmann H. Chronic Inflammatory Demyelinating Polyneuropathy: Current Therapies and Future Approaches.Curr Pharm Des [Internet]. 26 Mar 2022 [cited 25 Apr 2023];28(11):854–62. Available at:https://pubmed.ncbi.nlm.nih.gov/35339172/

22. Lewis R, Muley S. UpToDate[Internet]. 2022 [cited 25 April 2023]. Chronic inflammatory demyelinating polyneuropathy: Treatment and prognosis - UpToDate. Available at:https://www.uptodate.com/contents/chronic-inflammatory-demyelinating-polyneuropathy-treatment-and-prognosis?search=The%20treatment%20of%20CIDP&source=search_result&selectedTitle=1~63&usage_type=default&display_rank=1#H483050551

23. Jacob S, Mazibrada G, Irani S, et al. The Role of Plasma Exchange in the Treatment of Refractory Autoimmune Neurological Diseases: a Narrative Review.J Neuroimmune Pharmacol [Internet]. 2021-12-01 [cited 2023-04-25];16(4):806–17. Available at:https://pubmed.ncbi.nlm.nih.gov/34599742/

24. Briani C, Visentin A. Therapeutic Monoclonal Antibody Therapies in Chronic Autoimmune Demyelinating Neuropathies.Neurotherapeutics [Internet]. April 1, 2022 [cited April 25, 2023];19(3):874–84. Available at:https://pubmed.ncbi.nlm.nih.gov/35349079/

25. Muley S, Jacobsen B, Parry G, et al.Rituximab in refractory chronic inflammatory demyelinating polyneuropathy. Muscle Nerve [Internet]. May 1, 2020 [cited April 25, 2023];61(5):575–9. Available at:https://pubmed.ncbi.nlm.nih.gov/31922613/

26. Doneddu P, Nobile E. Management of chronic inflammatory demyelinating polyradiculopathy.Curr Opin Neurol [Internet]. October 1, 2018 [cited April 25, 2023];31(5):511–6. Available at:https://pubmed.ncbi.nlm.nih.gov/30045075/

27. Chen Y, Tang X. Chronic Inflammatory Demyelinating Polyradiculoneuropathy in Associationwith Concomitant Diseases: Identification and Management. Front Immunol [Internet]. July 4, 2022 [cited April 25, 2023];13. Available at:https://pubmed.ncbi.nlm.nih.gov/35860284/

28. Marques O, Weiss G, Muckenthaler M. The role of iron in chronic inflammatory diseases: from mechanisms to treatment options in anemia of inflammation.Blood [Internet]. November 10, 2022 [cited January 22, 2024];140(19):2011–23. Available at:https://dx.doi.org/10.1182/blood.2021013472

29. Fisse AL, Motte J, Grüter T, et al.Comprehensive approaches for diagnosis, monitoring and treatment of chronic inflammatory demyelinating polyneuropathy. Neurol Res Pract [Internet]. 1 December 2020 [cited 22 January 2024];2(1):1–14. Available at:https://neurolrespract.biomedcentral.com/articles/10.1186/s42466-020-00088-8

30. Desai U. How I Treat Chronic Inflammatory Demyelinating Polyneuropathy Podcast.Neurol Ther [Internet]. October 1, 2023 [cited January 23, 2024];12(5):1409–17. Available at:https://pubmed.ncbi.nlm.nih.gov/37358694/

31. Al A, Jakobsen J. Outcome in chronic inflammatory demyelinating polyneuropathy: A systematic review and meta-analysis.Muscle Nerve

[Internet]. October 1, 2023 [cited January 23, 2024];68(4):388–96. Available at:https://onlinelibrary.wiley.com/doi/full/10.1002/mus.27820

32. Segura D, Sanchez A, Alvarez K, et al.Chronic inflammatory demyelinating polyradiculoneuropathy with antineurofascin-155 antibodies: A first case report in Peru. Medwave [Internet]. 1 January 2023 [cited 23 January 2024];23(1). Available at:https://pubmed.ncbi.nlm.nih.gov/36652591/

33. Sawma T, Kanso M, Khalife J, et al.Chronic inflammatory demyelinating polyneuropathy caused by hepatocellular carcinoma. Case Rep [Internet]. February 10, 2023 [cited January 23, 2024];16(2). Available at:https://pubmed.ncbi.nlm.nih.gov/36764741/

34. Rostovtseva T, Dolgushin M, Lopatina A, et al.Involvement of the central nervous system in a patient with chronic inflammatory demyelinating polyneuropathy: a case report. Lučevaâ diagnostika i terapiâ [Internet]. 19 October 2022 [cited 23 January 2024];13(3):77–87. Available at:https://typeset.io/papers/involvement-of-the-central-nervous-system-in-patient-with-3o9t1ht3

35. Ferraz I, Correia I, Cruz S, et al. Chronic Inflammatory Demyelinating Polyradiculoneuropathy and Diabetes: A Case Report.Cureus [Internet]. September 21, 2022 [cited January 23, 2024];14(9). Available at:https://www.ncbi.nlm.nih.gov/pmc/articles/PMC9586193/

36. Rajabally Y. Chronic inflammatory demyelinating polyneuropathy with positive anti-myelin associated glycoprotein antibodies: Back to clinical basics.Eur J Neurol [Internet]. February 1, 2023 [cited January 23, 2024];30(2):301–2. Available at:https://pubmed.ncbi.nlm.nih.gov/36409135/

37. Suponeva N, Grishina D, Ryabinkina Y, et al.[Chronic inflammatory demyelinating polyneuropathy with an acute onset. Clinical case]. Ter Arkh [Internet]. 2022 [cited 2024-01-23];94(4):544–51. Available at:https://pubmed.ncbi.nlm.nih.gov/36286806/

38. Gogia B, Rocha F, Khan M, et al.Chronic Inflammatory Demyelinating Polyradiculoneuropathy - PubMed. StatPearls [Internet]. 2023 [cited 2024-01-23]; Available at:https://pubmed.ncbi.nlm.nih.gov/33085396/

39. Allen J, Lewis R. Diagnosis of chronic inflammatory demyelinating polyneuropathy.Muscle Nerve [Internet]. November 1, 2022 [cited January 23, 2024];66(5):545–51. Available at:https://pubmed.ncbi.nlm.nih.gov/36259623/

40. Malik A, Inayat F, Naeem M, et al.Colorectal carcinoma and chronic inflammatory demyelinating polyneuropathy: is there a possible paraneoplastic association? Clin Endosc [Internet]. 2023-03-01 [cited 2024-01-23];56(2):245–51. Available at:https://pubmed.ncbi.nlm.nih.gov/34315209/

41. Chauvet E, Rohner G, Manel V, et al.Autoantibodies to a Nodal Isoform of Neurofascin in Pediatric Chronic Inflammatory Demyelinating Polyneuropathy. Child Neurol Open [Internet]. January 2023 [cited 2024-01-23]; 10:2329048X2211496. Available in:https://pubmed.ncbi.nlm.nih.gov/36816544/

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