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Prognosis and outcome in patients with chronic inflammatory demyelinating polyradiculoneuropathy in a tertiary center in Oman

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Chronic Inflammatory Demyelinating Polyradiculoneuropathy; Prognosis; Nerve Conduction Studies; Treatment Outcome; Oman

Abstract

Background: Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an immune-mediated condition with variable clinical characteristics and different treatment modalities. We aim to present different clinical and demographic features of all patients with CIDP presented to the neuromuscular clinic within four years and their follow-up results.

Methods: A retrospective study from a hospital database of 23 patients met the European Federation of Neurological Societies/Peripheral Nerve Society (EFNS/PNS) diagnostic criteria for CIPD. Complete demographic and clinical data were collected. Progress and outcome were assessed using two clinical score systems at regular intervals at 6, 12, and 18 months.

Results: Mean age of patients was 43.4 ± 20.9 years (male-to-female ratio was 1.6:1). Age of onset was 39.7 ± 18.0 years. At the presentation, the Medical Research Council sum score (MRCss) was 50 (39.7-51.3), and the modified Rankin Scale (mRS) was 3 (2.2-3.4). There was a significant improvement in MRCss during four periods ($P < 0.001$). Multiple comparisons revealed a significant difference in MRCss between the baseline and 12 and 18 months but no significant change between the baseline and 6 months. Likewise, mRS showed a significant improvement between the baseline and 18 months (no significant change between the baseline and 6 months or 12 months).

Conclusion: The clinical characteristics of CIDP in our cohort were similar to other reported studies, and most of the studied patients had good outcomes. Our results could be utilized as baseline data for a better understanding of the characteristics of CIDP in Oman and, consequently, for better management of the disease.

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Introduction

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an acquired, immune-mediated condition characterized prominently by progressive muscle weakness and sensory dysfunction, which affects 1.0 to 8.9 persons per 100000.[1,2] It is a heterogeneous disease with different clinical presentations as well as variability in the clinical spectrum, ranging from almost asymptomatic patients to wheelchair-bound ones, reflecting heterogeneity in peripheral nervous system (PNS) involvement and response to therapies.[3]

CIDP diagnoses depend on pattern recognition, clinical symptoms and signs, electrodiagnostic studies, and other laboratory tests.[4] Typical forms of CIDP present as symmetrical proximal and distal muscle weakness in all four limbs, large-fiber sensory loss, and reduced or absent reflexes.[5] As CIDP became better recognized, researchers proposed various diagnostic criteria based on clinical features, specific electrodiagnostic criteria, and ancillary studies, including nerve biopsy or lumbar puncture.[6]

Despite available effective immune therapies, treatment response varies, and outcomes range from complete remission to severe disability. The two common first-line treatments for CIDP are corticosteroids [either tablet or intravenous (IV) infusion] and immunoglobulin (Ig).[7]

Neurological impairment could be assessed by routine neurological examination and by using various impairment scales, such as Medical Research Council sum score (MRCss), Inflammatory Neuropathy Cause and Treatment (INCAT) disability score, and Rasch-built Overall Disability Scale (RODS).[8]

The main goal of the current study was to investigate all patients who presented to our hospital within four years, collecting different clinical and demographic data, and correlating them with the various clinical responses.

Materials and Methods

This study was retrospective and descriptive from a hospital database system. Data analysis of 23 patients who presented to the neuromuscular clinic in Khoula Hospital, Muscat, Oman, from January 2016 to May 2020 was completed after obtaining ethical approval from the Institutional Ethics Committee (code: RO052022096).

All patients who met the European Federation of Neurological Societies/Peripheral Nerve Society (EFNS/PNS) diagnostic criteria for CIPD were included in the study.[6] The patients were excluded if they did not meet the diagnostic criteria or had significant comorbidities or CIDP mimics such as hereditary neuropathy and multifocal motor neuropathy with conduction block. Demographic data were collected, including age, gender, and area of residency. The full detailed medical background was documented. Different variabilities were assessed regarding the age of onset, disease duration, weakness distribution, symmetry, sensory involvement, and clinical features at the time of diagnosis using the MRCss and modified Rankin Scale (mRS). MRCss and mRS were assessed at baseline, 6, 12, and 18 months to determine the functional recovery. For MRCss, we evaluated bilateral muscle powers in the upper limbs (arm abduction, elbow flexion, and wrist extension) and in the lower limbs (hip flexion, knee extension, and foot dorsiflexion) with a maximum MRCss of 60. A nerve conduction study (NCS) was done on all patients. Cerebrospinal fluid (CSF) analysis was performed on almost all patients.

Statistical analyses were performed using RStudio (R version: R-3.6.1). The compliance of variables with normal distribution was tested with the Shapiro-Wilk test and probability graphics.

The descriptive data following a normal distribution were reported as mean ± standard deviation (SD), and those not following a normal distribution were reported as median (25^{th}-75^{th} percentiles). We used repeated measure methods to assess the clinical improvement of the patients in four time periods of baseline (T0), 6 months (T1), 12 months (T2), and 18 months (T3). Since the data did not follow a normal distribution, for repeated measure analysis, the Friedman test was used for within-group comparisons of MRCss and mRS. A P-value below 0.05 was considered significant.

Results

A total of 23 patients who presented to the neuromuscular clinic at Khoula Hospital, from January 2016 to May 2020 with CIDP diagnosis were included. The mean age was 43.4 ± 20.9 years, and the male-to-female ratio was 1.6:1. Duration of symptoms before diagnosis ranged from one month to 5 years, with a mean duration of 14.3 ± 18.6 months**.** Age of onset ranged from 10 to 83 years old with a mean of 39.7 ± 18.0 years.

The demographic and clinical data are summarized in table 1.

Table 1. Demographic and clinical characteristics of the patients with chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)

Characteristics	Total	Men (n = 14)	Women (n = 9)
Age (year)	46.0 ± 19.3	51.0 ± 20.0	37.8 ± 15.6
Delay to diagnosis (month)	14.3 ± 18.6	15.0 ± 20.0	13.0 ± 16.8
Age of onset (year)	39.7 ± 18.0	44.7 ± 18.5	32.0 ± 14.6
Other disorders			
Diabetes	5 (21.7)	4	1
Hypertension	3 (17.4)	2	1
Thyroid disease	4 (17.4)	2	2
Dyslipidemia	2 (8.7)	2	0
Heart disease	1 (4.3)	0	1
Connective tissue disease	1 (4.3)	0	1
Presentation			
Distal weakness	13 (56.5)	8	5
Sensorimotor distribution	15 (65.2)	10	5
Symmetrical distribution	19 (82.6)	12	7
MRCss at presentation	47.0 ± 10.0	48.0 ± 8.9	46.0 ± 12.6
mRS at presentation	2.7 ± 1.0	2.8 ± 1.0	2.8 ± 1.0
NCS			
Typical demyelinating features	19 (82.6)	10	9
CSF			
Elevated CSF protein (n = 19)	16 (84.2)	9	7

Data are presented as mean ± standard deviation (SD) or number and percent
MRCss: Medical Research Council sum score; mRS: Modified Rankin Scale; NCS: Nerve conduction study; CSF: Cerebrospinal fluid

At the presentation, the MRCss was 50 (39.7-51.3), and the mRS was 3 (2.2-3.4).

Nine patients (39.1%) lived in the Muscat area, and 14 (60.9%) came from regional governorates. Five patients (21.7%) had diabetes mellitus (DM), four (17.4%) with thyroid disease, four patients (17.4%) had hypertension (HTN), two (8.7%) with dyslipidemia, one (4.3%) had cardiac disease, and another one (4.3%) with connective tissue disease (Table 1).

Seven patients (30.4%) presented with pure motor symptoms, one (4.3%) with pure sensory, and 15 (65.2%) with mixed sensorimotor manifestations. Thirteen of them (56.5%) had distal symptoms, and ten patients (43.5%) had both proximal and distal symptoms. Nineteen patients (82.6%) had symmetrical manifestations at the diagnosis.

All 23 patients were investigated with a NCS. Nineteen (82.6%) had typical features of CIDP, and four (17.4%) were categorized as atypical CIDP. Nineteen patients (82.6%) went through a CSF study, sixteen (84.2%) patients had elevated CSF protein levels (> 45 mg/dl), and three patients (15.8%) had a normal study.

During the management of the acute stage, 21 patients (91.3%) received intravenous immunoglobulin (IVIg), of whom, 17 patients (81%) received a single course, and four patients (19%) required repeated courses due to the severity of symptoms and slow recovery. Three patients (13%) received one course of methylprednisolone.

During follow-up, 14 patients (60.9%) required polytherapy as maintenance therapy in addition to the monthly IVIg or IV methylprednisolone, with immunosuppressive drugs such as azathioprine or mycophenolate mofetil. Ten (43.5%) patients received oral prednisolone as long-term therapy, and one refractory case was treated with rituximab as maintenance therapy.

Outcome measures at follow-up: Friedman test revealed a significant improvement of MRCss during four time periods of baseline, 6 months, 12 months, and 18 months ($P < 0.001$) (Figure 1). Multiple comparisons revealed a significant difference in MRCss between the baseline and 12 and 18 months but no significant change between the baseline and 6 months.

Likewise, a significant improvement in mRS was found during follow-up ($P < 0.001$) (Figure 1). However, mRS showed a significant improvement between the baseline and 18 months (no significant change between the baseline and 6 months or 12 months).

Discussion

In our study, there was a male predominance

among the patients with CIDP in Oman with a male-to-female ratio of 1.6:1, which is similar to some other studies,[9-14] while other studies reported different ratios: Rajabally et al.: 2.3:1,[15] Chio et al.: 2.1:1,[16] Mahdi-Rogers and Hughes: 1.9:1,[17] and Abraham et al.: 4:1.[18]

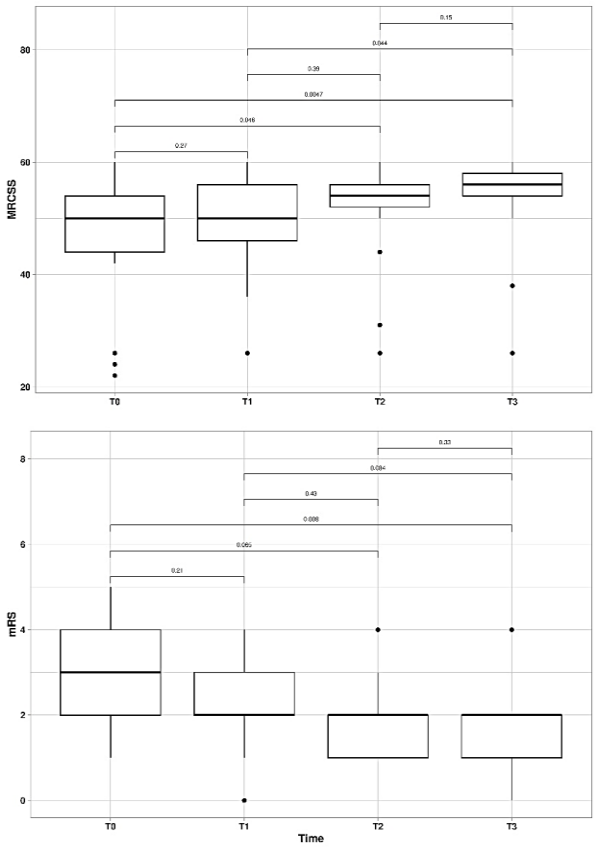

Figure 1. Medical Research Council sum score (MRCss) and modified Rankin Scale (mRS) change in the periods of baseline, 6 months, 12 months, and 18 months

The average age of onset was 43 years, similar to the Okhovat et al.[19] study at 40 years, while Mygland and Monstad[14] reported a mean age of 48 years. However, some studies stated older ages of onset ranging from 53 to 61 years.[18,20-23]

Our study's most commonly reported medical disorder associated with CIDP was diabetes (21.7%). Abraham et al.[18] and Alabdali et al.[23] reported a higher predominance of hyperlipidemia in patients with CIDP.

The average time from onset until the first medical assessment was 14 months, while other studies reported a shorter diagnosis delay ranging from three to seven months.[9,20] One patient presented within one month with Guillain-Barré syndrome (GBS)-like symptoms and was ultimately diagnosed as CIDP as his disease progressed in a waxing and waning pattern. Such a presentation has been reported in several studies.[24,25]

Mixed sensory motor manifestations were predominant in our study, with a percentage of 65%, while Rentzos et al.[9] reported 92%, and Mygland and Monstad[14] reported 100% of patients with mixed sensory and motor manifestations. The distal manifestation was predominant in 57% of patients, as reported by Panaite et al.,[26] and 82.6% showed symmetrical presentations, which is similar to Rentzos et al.[9]

CSF protein was higher than normal in 84% of patients compared to Rentzos et al.[9] and Panaite et al.[26] who reported a predominance of 65%. Meanwhile, Rajabally and Narasimhan[22] reported that 69% of their patients had high CSF protein.

Considering outcome measurements of MRCss and mRS, a significant improvement of both scores was achieved at 18 months. However, in shorter intervals, a substantial improvement of MRCss between the baseline and 12 and 18 months was seen, but this significant improvement was not observed after six months. Hence, it is required to continue treatment for more than a minimum of six months to attain considerable improvement.

The limitation of our study is an observational retrospective analysis with a low sample size. A well-designed prospective analysis of a larger sample of patients with CIDP from different healthcare centers and hospitals, including more specific outcome measures, needs to be carried out to achieve more reliable and representative data.

Conclusion

The clinical presentation of CIDP in most patients in Oman is similar to what was reported by previous studies, and most of the included patients had favorable prognoses. Our results could be utilized as baseline data for a better understanding of the characteristics of CIDP in Oman and, accordingly, for better management of the disease.

Conflict of Interests

The authors declare no conflict of interest in this study.

Acknowledgments

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Ethical approval of this study was obtained from the Institutional Ethics Committee of Khoula Hospital (ethical code: RO052022096).

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