Case Report

Chronic Inflammatory Demyelinating Polyneuropathy In A 39-Year-Old Nigerian Lady and Challenges of Care in A Resource Constrained Environment

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ABSTRACT

A 39-year-old lady presented with progressive limb weakness of twelve (12) months duration with associated tingling, shock-like burning sensations on both hands and feet. Her physical examination revealed flaccid quadriparesis with moderate bilateral hand muscle atrophy as well as impairment in light, crude touch and joint position sense. Laboratory and electrodiagnostic work up strongly suggested acquired immune mediated peripheral neuropathy. She received intravenous immunoglobulin and muscle power gradually improved. Apart from the high cost of intravenous immunoglobulin, other challenges were encountered in the course of management of this patient. The aim of this case report is to highlight the challenges of care of this condition in a resource poor setting

Keywords: Acquired neuropathy,chronic inflammatory demyelinating polyneuropathy, intravenous immunoglobulin, inflammatory neuropathy.

INTRODUCTION

Chronic inflammatory demyelinating polyneuropathy is an uncommon cause of acquired disabling peripheral nerve disorder characterized by symmetrical, proximal and distal sensory, and motor impairment developing for more than 8 weeks.[1,2]It is a treatable autoimmune disorder and the most common chronic acquired immune-mediated inflammatory neuropathy. Clinical features are heterogeneousand analogous to those of acute inflammatory demyelinating polyradiculoneuropathy, which runs acute courses.[1,3]Epidemiologic studies of CIDP are limited probably due to its rare nature.[4] However, its prevalence and incidence vary in different countries and regions.[1,4,5] Globally, prevalence of CIDP was observed to range from 0.67 to 10.3 cases per 100,000 with its incidence varying between 0.15 and 1.6 cases per 100,000 person-years.[4] While its incidence rises with advancing age, its peak occurrence is between 40 and 60 years with a male preponderance.[4,6] In Sub Saharan Africa, only very few studies on CIDP are documented and most of them are largely case reports.[7,8]

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Diagnosis of CIDP is based on clinical, electrodiagnostic (mandatory) and supportive criteria, according to the European Federation of Neurological Societies/Peripheral Nerve Society (EFNS/PNS) Criteria guidelines.[9,10] Diagnosis in our environment is often bedeviled by unavailability of facilities for electrodiagnostic work up, making it difficult for clinicians to recognize potential cases. Even when recognized, limited options for immunomodulatory treatment pose additional challenge. Intravenous immunoglobulin is not readily available in Nigeria, and if seen at all, comes with a huge cost. Thus, the overall burden of care for CIDP is high with a huge economic burden on the patients. The aim of this article is to report this rare disorder as well as highlight the challenges encountered in the course of management of this young Nigerian patient.

CASE REPORT

A 39-year-old Nigerian lady presented to our Neurology Out Patient Clinic with progressive limb weakness of about twelve (12) months duration. Weakness was initially noted in both upper limbs and later progressed to both lower limbs. Few weeks later, she developed gradual onset tingling, burning and shocking sensations on both distal upper and lower limbs. Over the next few months, her illness had progressed to the extent she could no longer ambulate without assistance and had to use wheelchair more regularly. However, there was no dysphagia or other bulbar symptoms. Her sphincteric function was also intact. Her past medical history was not significant for any other conditions such as hypertension, diabetes or connective tissue disorders. Prior to her referral to our facility, she had been managed in a peripheral hospital where she was placed on oral steroids, which did not help her symptoms and consequently she was subsequently referred to our clinic for expert care.

Physical examination findings revealed flaccid quadriparesis (MRC power grade of 2/5 on both upper limbs; and 1/5 on the lower limbs). Significant atrophy was noted on the small muscles of her hands with poor grip strength bilaterally.There was impairment in both crude and light on both distal upper and lower limbs. Joint position and vibration sense was also impaired.

Her INCAT disability score was 10/10 suggesting a severe presentation.

A diagnosis of CIDP was suspected, at the first evaluation, on clinical ground and she was then requested to do a number of investigations to confirm her diagnosis and possibly establish presence of comorbidities. Most of her laboratory and imaging results returned normal except for cerebrospinal fluid chemistry which showed elevated protein with normal cells (albuminocytologic dissociation). See Table 1

Electrodiagnostic studies showed impairment in both sensory and motor nerve conduction parameters.

Sensory nerve conduction study showed prolonged distal latencies, slow conduction velocities, conduction block with temporal dispersions and low amplitudes. See Table II

The motor nerve conduction study result showed significantly impaired parameters in both distal lower limb nerves. See Table III.

The needle exam showed some positive sharp waves and fibrillation potentials in some lower limb muscles with some large polyphasic motor unitswith reduced recruitment.

Patient was subsequently commenced on intravenous human immunoglobulin (Globucel) @ 2g/kg after obtaining consent from her. She also received medication with intravenous hydrocortisone prior to IVIG infusion. In addition, she also received SC Clexane (Enoxaparin) for venous thromboembolism prophylaxis and normal saline for rehydration. Pregabalin was prescribed for her neuropathic symptoms. While on IVIG, she developed moderate generalized headache which responded to antipyretic and she became stable. Patient was discharged home after one week on admission and was commenced pulse dexamethasone therapy (40mg daily, four days every month for 6 months) according to PREDICT trial.[11]Neurorehabilitation was also advised.

At one month follow up, she admitted to significant improvement in her overall functional status and muscle power, especially hand grip, with overall INCAT disability scale of 6/10. As at this time, neurorehabilitation was still ongoing and she was

assured regular telephonic communication with the managing team to complement physical follow up.

Table 1: Investigation Result of the Patient

INVESTIGATIONS	RESULTS	NORMAL RANGE
Cerebrospinal fluid analysis		
Protein	0.76g/l	0.15 – 0.45
WBC cell:	0 – 2 cells/ml	
Yeast and epithelial cells	Nil	
Creatinine kinase (CK)	279 u/l	15 – 170
AST (SGOT)	33u/l	10 – 42
Fasting blood glucose	5.2mmol/l	4.2 – 6.4
Triglycerides	0.69mmol/l	<1.71
LDL cholesterol	3.49 mmol/l	<3.30
HDL cholesterol	0.93 mmol/l	1.04 -1.55
Serum Urea	2.1 mmol/l	1.5 -6.6
Serum Creatinine	55 µ mol/l	60 -120
Calcium	2.55mmol/l	2.02 – 2.6
HIV I & II	Sero -negative	
HBsAg	Sero -negative	
HCV antibody	Sero -negative	
ANA	Normal	
dsDNA Antibody	Normal	
ESR	74mm/hr	2-11mm/hr
Packed Cell volume (PCV)	36%	36 -52%
TOTAL	9.9 X 10^9/L	4.8 – 10.8 %
Differentials:		
Neutrophil	80%	40 – 75%
Lymphocyte	12%	0 – 45%
MRI		
Brain	Normal	
Cervical	Normal	
Lumbosacral spines	Normal	

Table 2: Sensory nerve conduction parameters of the patient with CIDP

Nerve/Site	Rec. site	Onset Lat (ms)	Peak latency (ms)	NP Amp (µV)	PP Amp	Segment	Dist cm	Velocity (m/s)	Comment
R. Median-Dig 2 (Antidromic)									
Wrist	Index	2.92	3.33	21.4	21.2	Wrist-Index	14	48	
L. Median-Dig 2 (Antidromic)									Temporal
Wrist	Index					Wrist-Index	14		Dispersion
R.Ulnar-Digit 5(Antidromic)									
Wrist	Digit 5	2.76	3.07	2.4	5.3	Wrist-Digit 5	14	51	
L. Sural-(Antidromic)									
Calf	Ankle	2.19	2.60	5.8	10.1	Calf-Ankles	14	64	
R. Sural-(Antidromic)									
Calf	Ankle	2.66	3.39	4.3	6.6	Calf-Ankle	14	53	

Table 3:Motornerve conduction parameters of the patient with CIDP

Nerve sites	Muscle	Latency ms	Amplitude mV	Segments	Dist cm	Lt Diff ms	Velocity m/s	Comment
R. Median-APB								
Wrist	APB	11.08	0.7	Wrist-APB	8			
Elbow	APB	24.17	0.2	Elbow-Wrist	18.5	13.08	14.1	
L. Median APB								
Wrist	APB	12.96	0.6	Wrist-APB	8			
Elbow	APB	21.44	0.4	Elbow-Wrist	18.5	8.48	21.8	
R Ulnar-ADM								
Wrist	ADM	12.17	0.3	Wrist-ADM	8			
B. Elbow	ADM	23.52	0.3	B-Elbow-Wrist	18	11.35	15.9	
A. Elbow	ADM	28.87	0.2	A.Elbow-B.Elbow	12	5.35	22.4	
L. Ulnar-ADM								
Wrist	ADM	16.04	0.2	Wrist-ADM	8			
B. Elbow	ADM	19.15	0.2	B. Elbow-wrist	18	3.10	58.0	
A.Elbow	ADM	22.40	0.1	A. Elbow-B. Elbow	12	3.25	36.9	
R. Peroneal-EDB								
Ankles	EDB			Ankle-EDB	8			***No Response**
L. Peroneal-EDB								
Ankle	EDB				8			***No Response**
R. Tibial-AH								
Ankle	AH			Ankle-AH	8			***No response**
L. Tibial-AH								
Ankle	AH			Ankle AH	8			***No response**

This case report highlights not only the rarity of CIDP in our environment but also the challenges of care of this condition in resource constrained settings. Peripheral neuropathies are heterogeneous in their presentations and would often confuse clinicians whose practice focus is not on peripheral nerve disorders. CIDP is quite rare accounting for about 53% of all acquired neuropathies.[5,12] Only few case reports of this condition have emanated from Nigeria further buttressing the rare nature of this disorder. She appeared to have had the typical variant of CIDP which accounts for 50-60% of the disease.[13,14] Typical CIDP is a symmetric sensorimotor polyneuropathy with proximal and distal motor involvement that exceeds sensory involvement as seen in our patient (motor predominant chronic demyelinating polyneuropathy). Most patients with typical CIDP exhibit a slowly progressive trajectory as seen in our patient but about one-third of patients may have a relapsing-remitting course.[6] However, about 30% of all CIDP patients have been noted to achieve a cure following appropriate interventions.[6]

Atypical variants are less common and include distal acquired symmetric neuropathy (DADS), [15] multifocal acquired demyelinating sensory and motor neuropathy (Lewis-Sumner syndrome),[16] focal CIDP forms, sensory and motor CIDP.[4,17]

Iyagba et al[8] had also reported a similar case in an adult female Nigerian in Port Harcourt who presented with prominent autonomic features. Our patient did report any autonomic symptoms at presentation, or even during hospitalization, but autonomic dysreflexia is a recognized complication of CIDP in some cases.[18] Electrodiagnostic evaluation of peripheral neuropathies is important in classifying the pathological process as well as providing clues on overall outcome.[19] Our patient was able to carry out nerve conduction studies which further increased the credibility of our diagnosis.

Our patient also had idiopathic form of CIDP in which other systematic, metabolic, inflammatory, neoplastic and infectious diseases are absent.[4] Some of these conditions include diabetes mellitus, monoclonal gammopathy of undetermined significance(MGUS), POEMS.[4,20] Others are Human Immunodeficiency Virus(HIV) infection, systemic lupus erythematosus (SLE) Hepatitis C, Hepatitis B infection, Sjogren syndrome, thyroid disease, Hodgkin lymphoma and melanoma.[4]

Diagnosis of CIDP is challenging due to its overlapping features with several other neuromuscular disorders which often confuse most clinicians, especially non neurologist physicians, making it difficult to recognize this condition early for timely intervention. Late detection leads to severe and irreversible peripheral nerve damage and this leaves patient with permanent disability. For instance, the index patient was earlier diagnosed in a peripheral hospital as having muscular dystrophy and was managed as such. Apparently, her earlier clinical evaluations failed to tease out the salient features that made CIDP most likely. So, there was significant delay before she was eventually referred to a bigger facility.

Comprehensive investigation of peripheral nerve disorders is not only tedious but also challenging usually due to the diverse nature of their causes. For most suspected cases of CIDP in our environment, arriving at a definite diagnosis is often not easy due to lack of diagnostic facilities. Diagnosis requires fulfillment of both clinical and electrodiagnostic criteria and the later requires EMG/NCS machine which is not available in most hospitals in Nigeria, and where such exists, cost of investigation is usually an issue. Apart from electrodiagnostic study, neuromuscular ultrasound is another important, novel and cheaper tool for investigating peripheral nerve disorders[10,21] Unfortunately, this modality is still new and yet to gain ground in our routine clinical practice. Another method for diagnosing peripheral nerve disorders is nerve biopsy.[22] Histological examination of nerve tissue is also invaluable in clarifying pathological mechanisms of various peripheral disorders. The consequence of these diagnostic limitations is huge, either resulting in delayed or missed diagnosis

Lastly, treatment of CIDP is generally expensive especially with intravenous immunoglobulin or plasmapharesis. Intravenous steroids are cheaper compared to most other immunodulatory medications. Our patient procured intravenous

immunoglobulin at the cost of 5000USD and quite frankly, it would not have been affordable to an average Nigerian patient with a similar disorder

CONCLUSION

CIDP is quite rare in our environment but sure exists. Factors that contribute to delay in diagnosis are numerous, but more importantly, its heterogeneous clinical presentations often confuse most clinicians.The challenge of care of this condition mostly stems from lack of diagnostic facilities as well as high cost of treatment, especially in resource poor clinical setting. Thus, there is need for government and other relevant stake holders to make available needed infrastructures for diagnosis and treatment of various peripheral nerve disorders.

Abbreviations

CIDP Chronic inflammatory demyelinating polyneuropathy

IVIG Intravenous immunoglobulin

INCAT Inflammatory Neuropathy Cause and Treatment Disability Scale

EMG/NCS Electromyogram and nerve conduction studies

HIV Human Immunodeficiency Virus

MRC Medical Research Council Sum Score

POEMS Polyneuropathy, Organomegaly, Endocrinology, Monoclonal gammopathy and skin changes

CMAP Compound Muscle Action Potential

EFNS/PNS European Federation of Neurological Societies/Peripheral Nerve Society

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