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Neuromyelitis Optica Rehabilitation: Insights from a Single-Patient Case Report

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ABSTRACT:

Background and Purpose: This case examines the inpatient rehabilitation experience of an Indian male with Neuromyelitis Optica Spectrum Disorder (NMO).

Case Description: The patient was a 34-year-old Indian male with acute onset NMO with a unique presentation that affected his left visual field, left upper and lower extremity.

Intervention: The patient received rehabilitation according to his physical impairments. During his 16-day stay, the patient received a total of 2345 minutes of rehabilitation involving physical and occupational therapy. This patient did not require speech therapy services. Treatments included functional mobility training, gait training, activities of daily living training, strength and endurance training, orthotics, kinesiotaping, dry needling and neuromuscular reeducation.

Outcomes: Functional mobility and activities of daily living performance for physical and occupational therapy were assessed using the Inpatient Rehabilitation Patient Assessment Instrument per Centers for Medicare and Medicaid Services standards. Improvements in bed mobility, transfers, gait, upper and lower body dressing, selfcare, range of motion, strength, endurance, and overall functional mobility were observed. The participant was able to safely return home and begin outpatient physical therapy following discharge.

Discussion: This case analyzes the clinical presentation and course of inpatient rehabilitation in a young male with NMO. This case supports the use of individualized multidisciplinary therapeutic services in the rehabilitation of NMO. The outcomes in this case support further investigation for physical rehabilitation in NMO patients.

I. INTRODUCTION

Neuromyelitis Optica Spectrum Disorder (NMO) is a rare neurological disease commonly mis-diagnosed as Multiple Sclerosis (MS).¹ Patients typically present with central spinal cord lesions resulting in bilateral (B) vision loss and paralysis to the arms or legs and even the face. NMO is most prevalent in females as well as Asian, Black and Indian populations.^{2,3} In the United States, the average age of onset is 41.1 years.⁴ The 2015 international diagnostic criteria for NMO are summarized in Table 1.⁵ The six listed core clinical characteristics involve 1 of 6 central nervous system regions: optic nerve, spinal cord, area postrema of dorsal medulla, brainstem, diencephalon, or cerebrum respectively.⁵

Table 1 NMOSD diagnostic criteria for adult patients**Diagnostic criteria for NMOSD with AQP4-IgG**

1. At least 1 core clinical characteristic
2. Positive test for AQP4-IgG using best available detection method (cell-based assay strongly recommended)
3. Exclusion of alternative diagnoses^a

Diagnostic criteria for NMOSD without AQP4-IgG or NMOSD with unknown AQP4-IgG status

1. At least 2 core clinical characteristics occurring as a result of one or more clinical attacks and meeting all of the following requirements:
 - a. At least 1 core clinical characteristic must be optic neuritis, acute myelitis with LETM, or area postrema syndrome
 - b. Dissemination in space (2 or more different core clinical characteristics)
 - c. Fulfillment of additional MRI requirements, as applicable
2. Negative tests for AQP4-IgG using best available detection method, or testing unavailable
3. Exclusion of alternative diagnoses^a

Core clinical characteristics

1. Optic neuritis
2. Acute myelitis
3. Area postrema syndrome: episode of otherwise unexplained hiccups or nausea and vomiting
4. Acute brainstem syndrome
5. Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions (figure 3)
6. Symptomatic cerebral syndrome with NMOSD-typical brain lesions (figure 3)

Additional MRI requirements for NMOSD without AQP4-IgG and NMOSD with unknown AQP4-IgG status

1. Acute optic neuritis: requires brain MRI showing (a) normal findings or only nonspecific white matter lesions, OR (b) optic nerve MRI with T2-hyperintense lesion or T1-weighted gadolinium-enhancing lesion extending over >1/2 optic nerve length or involving optic chiasm (figure 1)
2. Acute myelitis: requires associated intramedullary MRI lesion extending over ≥ 3 contiguous segments (LETM) OR ≥ 3 contiguous segments of focal spinal cord atrophy in patients with history compatible with acute myelitis (figure 1)
3. Area postrema syndrome: requires associated dorsal medulla/area postrema lesions (figure 2)
4. Acute brainstem syndrome: requires associated periependymal brainstem lesions (figure 2)

Abbreviations: AQP4 = aquaporin-4; IgG = immunoglobulin G; LETM = longitudinally extensive transverse myelitis lesions; NMOSD = neuromyelitis optica spectrum disorders.

^a See table 2 and text discussion on serologic considerations for recommendations regarding interpretation of clinical and serologic testing.

Table 1. NMO Diagnostic Criteria

NMO can lead to severe disability. Disability is typically relapse related and treated aggressively to prevent permanent impairment.⁶ As with any neurological disease, physical rehabilitation is a key component in reducing disability and improving overall functional mobility. However, due to its low incidence, few studies address physical rehabilitation in patients with NMO. Rehabilitation services are generally based around MS protocols; however, efficacy is unknown. The limited research available suggests that a multidisciplinary rehabilitation program may lead to functional and neurological improvements in NMO patients.⁷⁻⁹

This case report outlines the inpatient rehabilitation (IRF) course of a 34-year-old Indian male with NMO. This report conforms to all CARE guidelines.¹⁰ This report did not require Institutional Review Board approval. Written consent was obtained from the patient.



II. CASE DESCRIPTION

History and Demographics Patient was a 34-year-old Indian male. He worked full time in the oil field prior to his diagnosis. Patient initially began experiencing thoracic spine and left (L) upper extremity (UE) pain, mild L lower extremity (LE) weakness, numbness, and tingling. He presented to a local rural hospital where he was given muscle relaxers, 6-day prednisone, and sent to local inpatient rehabilitation. The patient made some improvement initially, however over the course of a few weeks, his symptoms persisted and began to further affect his L UE and face. Patient reported to a distant level 1 trauma center for further workup. The patient underwent a lumbar puncture and cervical and brain magnetic resonance imaging (MRI). Cervical MRI revealed spinal cord thickening and edema as well as low-lying cerebellar tonsils into the foramen magnum. Brain MRI revealed asymmetric volume loss in the L optic nerve and low-lying cerebellar tonsils into the foramen magnum. There was no abnormal enhancement in the brain and cerebrospinal fluid was preserved. The patient was started on IV steroids and underwent plasmapheresis. Following bloodwork, the patient was diagnosed with seropositive NMO and discharged to a 40-bed inpatient rehabilitation facility. Differential diagnosis initially included cerebrovascular accident, transverse myelitis, and neoplasm. Unlike most NMO patients, there was minimal concern for MS secondary to his clinic presentation.

Initial Therapy Evaluation

Within 24 hours of admission, the patient was evaluated by physical therapy (PT) and occupational therapy (OT). Functional mobility and activities of daily living (ADL) were assessed using the Inpatient Rehabilitation Patient Assessment Instrument (IRF-PAI) per Centers for Medicare and Medicaid Services (CMS) standards. Mobility and self-care were scored as one of the following: Independent (IND), set-up or clean-up assistance (SU/CU), supervision or touching assistance (Sup, Touch A), partial/moderate assistance (Partial/Mod A), substantial/maximum assistance (Substantial/Max A), dependent (DEP), patient refused (Pat Ref), not applicable (Not App), not attempted due to environmental limitations (Not Att EL), or not attempted due to medical condition or safety concerns (Not Att MC). These scores are valued as 1-6 from dependent to independent. All not-attempted activities are valued as 1.

Weighted IRF-PAI quality indicators included eating, oral hygiene, toileting hygiene, shower/bathe self, upper-body dressing, lower-body dressing, putting on/taking off footwear, wash/dry hands, wash/dry face, comb/brush hair, tub/shower transfer, rolling left and right, sit to lying, lying to sitting edge of bed, sit to stand, chair to bed and bed to chair transfer, car transfer, picking up object, walk 10 feet, walk 50 feet with two turns, walk 150 feet, walking 10 feet on uneven surfaces, 1 step (curb), 4 steps, 12 steps, wheel 50 feet with two turns, and wheel 150 feet. Initial PT and OT scores can be found in Table 2 and Table 3, respectively. The patient's total mobility score on admission based on IRF-PAI coding was 31 for PT and 14 for OT.

Strength, range of motion (ROM), sensation, balance, vision, hearing, speech, bowel and bladder function, respiratory function, and cognition were assessed. All R sided assessments were WNL. Hearing, speech, and swallowing were WNL. Respiratory function and breath sounds were WNL. Bowel and bladder function were WNL; the patient was always continent. Cognition was WNL. The patient presented with isolated L visual, UE and LE impairments. Strength was assessed using the Medical Research Council Manual Muscle Testing (MMT) scale.¹¹

ROM was assessed visually. L UE and LE passive ROM was WNL however active ROM was impaired secondary to weakness. Cervical ROM was minimally limited in all planes and painful. Light touch, proprioception, deep pressure, and localization were assessed to gauge sensation; all were diminished in the L UE and LE. Sitting balance was assessed at edge of bed. The patient could sit independently and accept perturbations without UE support however he could not laterally shift to the left. The patient was unsafe for standing balance assessment on initial evaluation. Visual fields were assessed using the confrontation visual field test¹² Pt reported L visual field blurriness and demonstrated L peripheral vision deficits. The patient was categorized with a C2 American Spinal Injury Association (ASIA) level C L Brown Sequard spinal cord injury.

III. INTERVENTION

Treatment consisted of a multidisciplinary approach involving PT and OT. Based on his clinical presentation, treatment resembled that of a stroke patient. The patient did not require speech therapy services. Wheelchair Mobility Training¹³



Wheelchair propulsion was trained as a primary means of mobility. Due to L UE and LE weakness, wheelchair propulsion was performed with the R UE and LE. The L LE rested on a standard leg rest during mobility and the L UE rested on a standard arm tray. Initial cuing was required for path finding and sequencing. Wheelchair management including leg and arm rest management was performed with the R UE bilaterally.

Functional Mobility Training

Functional mobility began with training bilateral bed mobility and squat pivot transfers with handheld assistance. Maximum cuing was required for sequencing and safety initially. Out of bed mobility progressed as tolerance to upright positioning improved. Sit to stands and standing tolerance were initially trained in parallel bars. Sit to stands were progressed to a standard front wheeled rolling walker with L UE platform. The patient required cuing and facilitation for L scapular retraction and depression, L knee and hip extension for upright standing.

Activities of Daily Living Training ¹⁴

The patient presented to IRF with a basic understanding of hemiplegic upper and lower extremity dressing techniques from his previous rehabilitation. The patient initially utilized a reacher and dressing stick for UE and LE dressing. Dressing was progressed without the reacher and dressing stick as the patient improved. A standard sock aid was used for footwear initially but progressed and discontinued as well. Patient began shower training using a standard shower chair for most of the task. A long-handled sponge was used for general washing. As the patient increased standing tolerance, he began using grab bars to maintain standing balance for more of the task. As standing balance improved patient progressed toileting and toilet hygiene as well. He initially attempted to use a general bathroom buddy however eventually discontinued use.

Gait Training ¹⁵⁻¹⁷

Gait training began once the patient demonstrated safe upright standing and comfort with the rolling walker. Training initially began in parallel bars using an overhead body weight supporting harness with L UE and LE assistance and a step-to gait pattern. Gait was progressed to the standard front wheeled rolling walker with L UE platform used during standing. The patient initially presented with L knee buckling, L drop foot, R hip drop during L LE stance and decreased stride length bilaterally. The patient required max cuing and muscle facilitation initially. The patient eventually progressed to the addition of stair training using B rails, stepping up with the R LE and down with the L LE on each step.

Therapeutic Exercise ¹⁸⁻²⁰

General strength and cardiovascular endurance training were incorporated throughout the patient's stay. Strength training primarily focused on Triceps Surae, Anterior Tibialis, Quadriceps, Gluteus Medius, Gluteus Maximus, general abdominals, rotator cuff and scapular stabilizers, elbow, and forearm musculature. Strength training was progressed from active assisted range of motion to resisted range of motion using ankle weights, hand-held weights, and Thera Band. Cardiovascular endurance training was primarily trained using progressive reciprocal B UE and LE cycling. The patient was given a home training regimen and encouraged to continue addressing strength and endurance deficits outside of therapy.

Trigger Point Dry Needling (TPDN)

Throughout his stay, the patient presented with L upper back and UE musculoskeletal pain and palpable trigger points in his L upper, middle, and lower trapezius, latissimus dorsi, supraspinatus and infraspinatus. TPDN was performed on numerous occasions to the listed muscles. Limited evidence suggests TPDN may be effective in reducing pain in patients with myofascial trigger points.²¹ Visual twitch responses were observed, and the patient reported immediate pain relief. Despite evidence regarding TPDN efficacy lacking, this patient responded positively in the short-term.²²

Kinesiotaping

Kinesiotaping was used to facilitate L UE and LE musculature. Kinesiotaping to affected musculature has been shown to result in improved gait mechanics and balance in stroke patients.²³ Kinesiotape was consistently applied to the L anterior tibialis and L vastus medialis oblique using a facilitatory technique. The patient reported subjective improvement in muscle activation and motor control. Improvements in L knee extension and L ankle dorsiflexion were observed during gait. Taping was used in conjunction with orthotics for the above deficits. Additionally, tape was applied to the L shoulder. Kinesiotaping has been shown to decrease pain and improve glenohumeral joint subluxation in stroke patients.²⁴ This patient demonstrated increased joint space



between the humeral head and acromion as well as visual and palpable anterior humeral head translation. The patient reported an overall improvement in shoulder pain and function with consistent taping.

Orthoses

An over-the-counter generic ankle foot orthosis (AFO) was applied to the L ankle during all upright functional mobility to address dorsiflexion deficits and knee hyperextension.^{25,26} Notable improvements in L footdrop were observed during transitional mobility and gait. Once the patient demonstrated improved anterior tibialis muscle strength, a SaeboStep (Saebo, NC, USA) was utilized over the generic AFO. A DonJoy SE-4 (DJO Global, TX, USA) knee brace was applied to the L knee during all upright functional mobility to prevent knee buckling and hyperextension.²⁵ Improvements in L knee motor control and safety were observed. The patient reported increased confidence and perceived mechanics using orthotics with mobility. A SaeboStretch (Saebo, NC, USA) splint was applied to the L wrist and hand on a schedule to maintain adequate soft tissue mobility and prevent contractures. The L digits and wrist were splinted in neutral.

Neuromuscular Re-Education

Neuromuscular electrical stimulation (NMES) was utilized throughout the patients stay during therapeutic exercise and pre-gait training. Rectangular electrodes were applied to the L anterior tibialis and L quadriceps to address L foot drop and promote L knee extension and stability during L LE stance. Manual neuromuscular facilitation techniques and proprioceptive neuromuscular facilitation were used during functional mobility and gait to the necessary musculature. Techniques included stroking, tapping and deep pressure. These techniques have been shown to decrease muscle tone and increase overall function and motor control in stroke patients.^{27,28} L talocrural joint posterior mobilizations and manual stretching to the Gastrocnemius and Soleus and hamstrings were used in conjunction with therapeutic exercise. These techniques have been shown to improve range of motion and spatiotemporal gait parameters in stroke patients.²⁹

IV. OUTCOMES

The patient received 2345 minutes of therapy throughout his 16-day stay. His CMS risk-adjusted expected mobility score at discharge for PT and OT were 62 and 32 respectively. These expectations are based on his demographics, diagnosis, and associated comorbidities. The patient exceeded expectations with a total score of 93 and 36 for PT and OT on discharge, respectively. The patient was independent with bed mobility without the use of rails or mechanical bed function. The patient was independent with sit to stands, squat pivot transfers, and stand-step transfers to multiple surfaces using the same standard front wheeled rolling walker with L UE platform as noted above. The patient required setup for picking up an object secondary to the handing of the reacher. The patient was independent with ambulation totaling 200 feet using the standard front wheeled rolling walker with L UE platform, knee, and ankle braces. The patient required supervision for ambulation on uneven surfaces and 6-inch stairs due to safety concerns. The patient was independent with wheelchair mobility for 200 feet using his R UE and LE with his L LE on a standard leg rest during mobility and the L UE rested on a standard arm tray. The patient was independent with eating, oral hygiene, washing/drying his hands and face, combing/brushing his hair, upper body dressing and putting on and taking off footwear using the R UE. The patient required setup with toilet and bath/tub transfers using grab bars; the rolling walker utilized for standard transfers did not fit in the bathroom. The patient required supervision for toileting hygiene using grab bars and an elevated drop-arm commode. He required more than reasonable time and had mild instability causing safety concerns. The patient required supervision with lower body dressing. He would thread his legs B in sitting, using his R UE and a reacher. He would then stand-up using grab bars and pull up his pants using his R UE. The patient required supervision with showering using a long-handled sponge. He would sit in a shower seat with a back for most of the task and stand using grab bars for cleaning his perianal region. Discharge PT and OT scores can be found in Table 6 and 7, respectively.

V. DISCUSSION

Outcomes

The patient showed improvements in all aspects of functional mobility including strength, ROM endurance, and balance. The patient demonstrated an atypical NMO presentation, thus rehabilitation protocols mirrored stroke rehabilitation. The patients L sided impairments allowed him to compensate with his R UE and LE and use hemiplegic techniques as needed. The patient did not have any relapses during his stay. The patient was safely



discharged home and set to begin outpatient neuro rehabilitation the following week. There was no follow-up after discharge.

Limitations

This study was a retrospective design and therefore a few initial assessments were not reassessed at discharge and consequently left out of the outcomes. Consistent outcome measures and assessment may have provided further insight to the patient's functional improvements and recovery. Long term outcomes are unknown due to lack of follow up after discharge from IRF.

Summary

In most NMO cases, patients are initially mis-diagnosed with MS. Spinal cord lesions in NMO tend to be central, and patients typically present with B symptoms including vision loss and paralysis.¹ Additionally, symptoms can be relapsing and remitting.⁶ Therefore, rehabilitation is typically based on MS protocols. In this case, we discuss a patient with unilateral NMO affecting his L visual field, UE, and LE. To our knowledge, there are no studies describing NMO with this type of presentation. Additionally, studies regarding rehabilitation of any presentation of NMO are lacking. As such, this patient received rehabilitation primarily based on stroke protocols. The patient made notable improvements across all assessed domains and was safely discharged home and referred to outpatient neuro rehabilitation. This case supports the use of multidisciplinary therapeutic services in the rehabilitation of NMO.⁷⁻⁹ Therapeutic intervention should be based on clinical assessment of the patient's symptoms and functional impairments. Standard protocols may not always be viable in NMO patients secondary to varying clinical presentations and lack of efficacy. The outcomes in this case support further investigation for physical rehabilitation in NMO patients with varying clinical presentations.

Patient Perspective

The following perspective was recorded via a standard discharge survey. "All of the nursing staff, including Dr. X was very professional. They helped me out with everything that I needed from the therapy staff all the way to nursing and doctors." The patient rated his experience 10/10 and reported to be extremely likely to refer family or friends.

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