

Locked-In Syndrome Due to Neuromyelitis Optica Spectrum Disorder in Acute Rehabilitation: A Case Report



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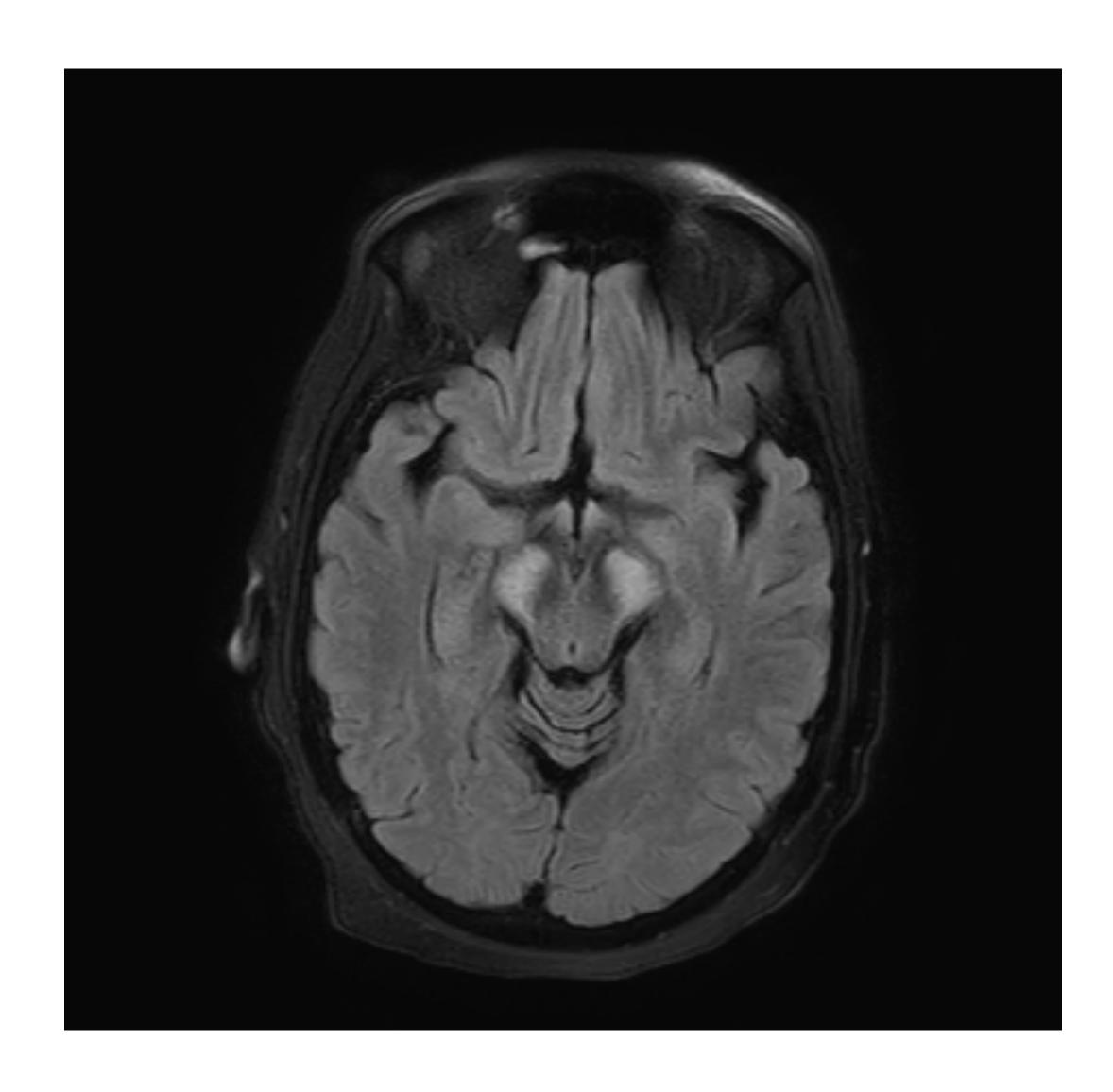
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Case Description

- 56-year-old female patient with prior questionable diagnoses of brainstem infarct, multiple sclerosis, and systemic lupus erythematosus who presented to the hospital with progressive dysarthria, lethargy, and weakness.
- MRI noted bilateral demyelinating lesions of the midbrain, pons, and middle cerebellar peduncles as well as nonspecific signal abnormality in the spinal cord at T6-T7.
- Her condition further deteriorated to flaccid complete tetraplegia.
 Communication was limited to extraocular movements and eye blinking.
- Serum aquaporin-4 antibody assay was positive.
- She was diagnosed with NMOSD resulting in transverse myelitis and locked-in syndrome.
- She was treated with IVIG and high-dose steroids and eventually plasma exchange.
- She completed 76 days of acute rehabilitation with a 3-day absence for rituximab infusions.

Progress at Acute Inpatient Rehab

- On admission, manual muscle testing revealed strength of 1/5 in bilateral intrinsic muscles of the hands; all other muscle groups were flaccid.
- At time of discharge, left upper extremity strength ranged from 2/5-3/5, right upper extremity strength ranged from 1/5-2/5, and strength in bilateral lower extremities remained 0/5.
- Initially, speech was intelligible 30-50% of the time compared to intelligible 90-100% of the time at discharge.
- She remained dependent for transfers, mobility, and most ADLs. Nutrition was provided through tube feeding at discharge.



Discussion

- Patient tolerated the acute rehabilitation well.
- Gained exposure to alternative methods of communication and demonstrated gains in voicing, volume, and intelligibility.
- Demonstrated improved seated stability and upper extremity use, allowing for increased participation in ADLs and transfers.
- Family was able to be trained, and the patient was able to independently direct care.

Conclusion

 Acute rehabilitation may be beneficial in the recovery of patients with NMOSD presenting with severe disability.

