Movement Disorder: A Case Study in myoclonus vs. ALS vs. MSA

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Introduction

- Movement disorders are an important presentation to recognize in the hospital and can have long term implications in patients' overall prognosis and quality of life. This case will discuss a patient who was initially admitted for altered mental status and tremors. Differentials during patient's hospitalization include myoclonus, ALS (Amyotrophic lateral sclerosis) vs MSA (Multiple system atrophy). We will discuss the pathophysiology's and patient presentations of each of these respective diseases, as well as what ultimately could have been done differently in this case for more prompt diagnosis and further management as an outpatient.

Objectives

- **Spread awareness, and educate healthcare professionals about movement disorders**
- **Elaborate warning signs to establish in common presentations of the movement disorders presented in this case**
- **Identify obstacles in this patient case and establish steps the primary team could do differently in a similar case in the future**

Case Presentation

This is a 49-year-old male with a history of GERD initially presenting to Atlanticare Regional Medical Center (ARMC) with altered mental status, weakness, tremors, burning abdominal pain, and poor oral intake. Patient attempted to relieve the burning abdominal sensation with Mylanta but was unsuccessful. Initial workup included creatinine of 19, wbc of 15k, and UDS positive for cannabis. All imaging including chest x-ray, Ct head, and CT abdomen/pelvis was negative. He was initially admitted to the ICU, started on vancomycin, cefepime, with blood cultures later noted to contain MSSA. Under the recommendation of infectious disease, he was started on nafcillin. An echocardiogram was completed and ruled out endocarditis as a source of infection. The patient was then started on CVVHD, followed by hemodialysis until renal function improved. An MRI of the brain was completed which did not show acute abnormalities. Per neurology's recommendations, the patient was transferred to a different facility for expedited lumbar puncture with a movement disorder specialist consultation.

He later was readmitted to ARMC for inpatient care while awaiting rehabilitation placement. Workup from the secondary facility was largely negative for any neurological/genetic abnormalities that may be causing his myoclonus/tremors. Patient had severe tremors and pain during his second admission. He was started on baclofen, oxycodone, and gabapentin for pain. In addition, valium was given for anxiety.

His primary concerns were spasms in the lower extremities that caused shooting pain to the rest of his body. On day 3 of admission, he had a sustained heart rate of 200 bpm for several minutes, and a MET (Medical Emergency Team) call was made. EKG revealed sinus tachycardia during this incident, and he was subsequently given several interventions including boluses of normal saline, 2 doses of IV Lopressor, and an early dose of his valium to further calm him down. After his heart rate had dropped to a baseline of 180 bpm following the MET, he was started on Metoprolol 25 mg twice daily. The next day he underwent an echocardiogram which revealed an ejection fraction of 35-40%, which was unchanged from his most recent echocardiogram completed in the outside facility. Cardiology was consulted and his GDMT regimen was optimized prior to his discharge.

Neurology was also consulted on this case due to an unclear etiology of his spasms; a 5 day course of IVIG was completed while in the hospital with a significant improvement of his spasms, pain, and overall anxiety. Discontinuing the patient's home Gabapentin, and initiating Lyrica while in the hospital also played a factor in the patient's overall improvement. Prior to discharge, the patient still lacked a diagnosis to explain symptomatology. However, the two most likely differentials discussed with the Neurology team were amyotrophic lateral sclerosis ALS and MSA. Myoclonus was deemed less likely to be the source of his spasms given the fact that he was able to communicate with the providers and nurses while he was having spasms, a feature that is not commonly seen in true myoclonus. MSA was also discussed to be less likely given the acute nature of his symptoms. He was ultimately deemed safe for discharge once his spasms were acutely improving and his heart rate was within significantly safer parameters; he was discharged with Valium, Lyrica, and his optimized GDMT regimen to continue in the outpatient setting. The final recommendation on discharge was for him to complete physical therapy at a rehabilitation center, complete an outpatient EMG for further investigation, and follow up with Neurology further in the outpatient setting.

Discussion

This case was selected due to its complexity, length of hospitalization, and amount of complications that presented throughout its course. In this discussion we will attempt to provide education on myoclonus, ALS , and MSA due to their relevance to the case. However, the differential diagnosis for movement disorders in the acute setting is very broad.

Myoclonus presents as a brief and sudden movement of muscles more commonly in the upper extremities, lower extremities, and the face. Myoclonus can be due to physiologic reasons including sleep, hiccups, or reactions such as being surprised or startled; these typically do not require any treatment. There are also myoclonic movements that occur secondary to seizures, particularly in patients with status epilepticus; these require patients to be on maintenance medications for their seizures to prevent further myoclonic movements. Secondary causes of myoclonus include autoimmune disorders, vitamin deficiencies, brain lesions, degenerative brain disorders, infections (such as herpes or lyme disease), spinal injuries, nerve injuries, medications, and toxins; treating these typically include treating the underlying cause or removing the offending agent respectively. Myoclonus was the primary diagnosis for the majority of the case; however, causes of myoclonus were ruled out based on the patient's physical examination and lab work. The important investigations during this hospitalization included blood count, metabolic panel, vitamin panel, CT imaging of the head, and lumbar puncture that did not reveal any significant findings. Physical examination also revealed that he was able to maintain active conversation with nurses and providers even while his spasms were occurring, which is not a typical feature of myoclonus.

Amyotrophic Lateral Sclerosis or "Lou Gehrig's Disease" is a movement disorder noted for generalized weakness, muscle wasting, fasciculations, and increased reflexes. The pathophysiology centers around loss of function in both upper and lower motor neurons, the affected anterior and lateral columns of the spinal cord become hard hence the description "lateral sclerosis". The most common causes of death due to this disease are respiratory failure and cachexia, and mortality is typically within 5 years. Diagnostic criteria for ALS involves lower motor and upper motor neuron signs in 3-4 regions with signs of progression for a definite diagnosis, at least 2 regions and progression for probable ALS, and distribution of upper motor and/or lower motor neuron signs in 1 or more regions with or without signs of progression for possible ALS. The only approved pharmacotherapy to treat ALS at this time is Riluzole, which can be an expensive medication to obtain in the outpatient setting. The other agents involved are for symptomatic relief, which include Baclofen, Valium, or Dantrolene to relieve spasticity in ALS patients. However, these may ultimately increase progression of the weakness and cause sedation and dizziness. Supportive therapy includes early initiation of physical therapy treatments in order for the patient to physically adjust to the fasciculations and pain. ALS was considered as a potential differential diagnosis. The patient had shown signs of upper moton weakness mainly in the lower extremities but there was milder fasciculations in the upper extremities. The patient was started on several of these medications including Baclofen and Valium, with Lyrica being an alternative therapy. The patient also completed physical therapy while in the hospital and was referred for therapy outpatient.

MSA is a movement disorder defined by dysregulation of the autonomic nervous system which can be classified into two main variations. Parkinsonian MSA presents with stiff muscles, bradykinesia, tremors at rest or while moving, dysarthria, and slurred or soft speech. Cerebellar MSA involves trouble with movement/coordination, dysarthria, dysphagia, and changes in vision. Some of the more general symptoms in MSA include constipation, bladder or bowel incontinence, changes in sweat production, agitated sleep, stridor, erectile dysfunction, loss of libido, and trouble with controlling emotions. There are currently no known genetic or environmental causes of MSA. Of note, there is a correlation between people who have REM sleep disorders, as people diagnosed with MSA often have these sleep disorders. The pathophysiology of MSA is largely believed to be atrophy of regions of the brain including cerebellum, basal ganglia, and brainstem. MSA typically has a mortality of 7-10 years after symptoms first appear with the most concerning complications being difficulty with breathing, and being more susceptible to infections or clots in the lungs due to autonomic dysregulation.

Conclusion

This case highlights the diagnostic challenges and complexities associated with movement disorders, emphasizing the importance of a thorough evaluation and broad differential diagnosis. Despite the initial diagnosis of myoclonus, further investigations ruled out typical causes revealing atypical features inconsistent with myoclonus. The suspicion of ALS was supported by clinical findings such as lower extremity weakness and fasciculations leading to targeted pharmacological and physical therapy interventions. Additionally, features of MSA were considered due to autonomic dysregulation. This case underscores the necessity of a systematic approach to movement disorders, integrating detailed clinical observations, diagnostic criteria, and multidisciplinary management to optimize patient outcomes. In the future, this knowledge of movement disorders will aid the primary team in making more prompt diagnosis given warning signs of each of the pathologies presented in this report.