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Utilization of Dantrolene in Stiff-Person Syndrome: A Case Report

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ABSTRACT

Setting: University hospital-based acute rehabilitation.

Patient: 75-year-old woman with Stiff-Person Syndrome (SPS) with a recent fall and Colles fracture.

Case Description: Four months prior to admission, the patient was diagnosed with SPS, negative for anti-GAD antibodies. Diagnosis was based on a 3-year history of progressive rigidity leading to frequent falls and fractures. Anxiety and fear of falling limited her mobility, and she sustained a sacral pressure ulcer during acute hospitalization. On admission, history was remarkable for undiary gait and muscle cramps exacerbated when startled or excited. Examination was remarkable for rigidity in her axial and limb muscles. She presented at the maximal assist level for transfers and toileting and moderate assist level for groming and ambulation using a platform walker (right arm in cast). She was unable to tolerate trialation of dantrolene due to rigidity in axial, then limb muscles, co-contraction of both agonist and antagonist muscles, and superimposed muscle spasms.

Results: Four months prior to admission, the patient was diagnosed with SPS, negative for anti-GAD antibodies. Diagnosis was based on a 3-year history of progressive rigidity leading to frequent falls and fractures. Anxiety and fear of falling limited her mobility, and she sustained a sacral pressure ulcer during acute hospitalization. On admission, history was remarkable for undiary gait and muscle cramps exacerbated when startled or excited. Examination was remarkable for rigidity in her axial and limb muscles. She presented at the maximal assist level for transfers and toileting and moderate assist level for groming and ambulation using a platform walker (right arm in cast). She was unable to tolerate trialation of dantrolene due to rigidity in axial, then limb muscles, co-contraction of both agonist and antagonist muscles, and superimposed muscle spasms.

Conclusion: Dantrolene is a useful additional treatment for SPS rigidity.

CASE REPORT

HISTORY
-75-year-old woman, history of anxiety and at risk Colles fracture after a fall, treated non-surgically.
-Progressed with present ambulatory dysfunction, rigidity, and frequent falls over 3 years.
-Always been stiff when startled or scared, had anxiety related to fear of falling.
-Diagnosed with anti-GAD-negative Stiff-Person Syndrome (SPS) 4 months before rehab admission.

PHYSICAL EXAMINATION
-Admitted to acute rehabilitation at the maximal assistance level for transfers and toileting, and moderate assistance level for grooming and ambulation with a platform walker.
-Exam at admission: intact strength without focal neurologic deficit with the exception of notable axial and limb rigidity, Modified Ashworth Score of 3 in all limbs.

TREATMENTS

PHARMACOLOGIC
-Physical Therapy
-Occupational Therapy

PHYSICAL THERAPY
-Physical therapy: strength training, balance, endurance, range of motion exercises.

PHARMACOLOGIC
-Neurotransmitters: GABA, serotonin, norepinephrine, dopamine.
-GABA agonists: diazepam, baclofen.
-Neurotransmitter deficits: GABA, serotonin, norepinephrine.

RESULTS
-Improvement of Modified Ashworth Score from 3 to 1-2 in all limbs with dantrolene.

Figure 1: Gordon Clinical Diagnostic Criteria for Stiff-Person Syndrome

1. Prodrome of episodic aching stiffness of axial muscles
2. Progression to include stiffness of proximal limbs
3. Painful spasms elicited by triggers
4. Increased lumbar lordosis
5. Normal sensation, motor function and intellect
6. Response to benzodiazepines

Figure 2: Pathogenic Mechanisms in Stiff-Person Syndrome

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DIAGNOSIS
-SPS, also known as Stiff-Man Syndrome, is a rare neuromuscular disorder characterized by rigidity, identified by Moersch and Woltman in 1956.
-Clinical diagnosis, still in use today, were first set by Gordon in 1967 (Figure 1).
-Rigidity and sudden spasms increase risk of falling.
-Associated anxiety and task-specific phobia, often misdiagnosed as psychological disorder.
-EMG demonstrates continuous motor unit activity at rest in both agonist and antagonist muscles; reduced EMG activity and rigidity when a patient is given dantrolene (GABA agonist) is often used by clinicians to confirm the diagnosis.
-Pathophysiology is one of excitation, with normal strength and without dystonia, extrapyramidal or pyramidal tract signs.

TREATMENT
-Physical and occupational therapies can improve mobilization and balance.
-Neuromuscular can reduce anxiety and avoidance behavior.

PHARMACOLOGIC
-Neurotransmitters: GABA, serotonin, norepinephrine.
-GABA agonists: diazepam, baclofen.
-Neurotransmitter deficits: GABA, serotonin, norepinephrine.

RESULTS
-Improvement of Modified Ashworth Score from 3 to 1-2 in all limbs with dantrolene.

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CONCLUSION
-SPS is a rare, progressive disorder characterized by rigidity, caused by deficient GABA utilization, that can severely limit quality of life.
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