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Case Report

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Stiff Person Syndrome

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Introduction

A rare, progressive condition of the central nervous system is known as stiff person syndrome (SPS). The symptoms can include severe immobility, rigidity, and painful muscle spasms in the trunk and limbs. Spasms can produce enough force to break a bone. Muscle spasms can occur in SPS patients because they are more sensitive to noise, rapid movements, and mental disturbance. The condition affects women twice as frequently as it does males. Increased muscular activity brought on by a decline in brain and spinal cord inhibition is the cause of SPS. Once known as stiff-man syndrome, this disorder is now also referred to as Moersch-Woltman Syndrome. High antibody titers against acid decarboxylase are prevalent, and SPS is closely associated with autoimmune disorders (GAD65) [1].

The current clinical classification of SPS includes:
1. Classic SPS
2. Partial SPS variants
3. Progressive encephalomyelitis with rigidity and myoclonus (PERM) [2].

We present the case of a 57-year-old woman with a history of migranous headaches, anxiety, and depression who developed diffuse painful muscle spasms in her shoulders, upper and lower back, and inability to move her neck. On examination, dense diffuse muscle stiffness was found throughout the body, and SPS was diagnosed after ordering a test for Glutamic acid decarboxylase antibodies. It is positive in roughly two-thirds of the patients.

Even though it is uncommon, SPS should be considered in patients who present with progressive diffuse muscle spasms. Dysautonomia, diffuse muscle stiffness, and recurrent headaches were amongst her symptoms. The patient described great problems bending and turning and walking like a "tin-man." Over time, the rigidity spread to the proximal upper and lower extremities. Furthermore, the patient reported painful generalized muscle spasms and exaggerated startle responses caused by unexpected tactile, visual, or acoustic stimuli in addition to strong emotions. She also encountered anxiety and depression, coupled with pathological startling. Because it is frequently misdiagnosed, practitioners should perform a thorough physical examination on any patient who presents with such unusual symptoms.

Case Presentation

A 57-year-old woman presented to the office due to diffuse muscle stiffness, pain, and spasm of the back of the head, neck and back since 15-20 years.

The patient is allergic to fish product derivatives, contrast media containing iodine, porcine derivatives, shellfish, and erythromycin. Her mother had colon cancer and osteoarthritis. She is married with two children. She is not a smoker or an alcoholic. The patient is currently employed, but she claims she is going to retire early due to the limitations imposed by her condition. Her supervisors are oblivious about her situation and have been rude to her in the past, especially when she had to be absent or leave early. She had to leave early from work at times to take her medications and "pass out" in bed. She has suffered from a lot of pain her entire life, and it's only getting worse as she gets older.

The patient is currently on the following medications: Albuterol, Budesonide-formetrol, cetirizine, epinephrine, famotidine, fluticasone propionate, fovatriptan, Gabapentin, Ibuprofen, Lisinopril, Magnesium oxide, Naproxen, Ondansetron, Pantoprazole, Polyethylene glycol, Polyvinyl alcohol, Ropivacaine, triamcinolone, zolmitriptan

Patient has been in the armed forces (air force) from 1980 to 1999 and had onset of some symptoms during her tenure, most prominently in the late 1990's, but they were not as pronounced compared to today. The patient describes a head injury in the 1990, but denied any weakness, or exposure to any rusty metal Symptoms in the 90's began with neck pain, numbness and tingling. A part of her job requirements at that time involved lifting of heavy equipment. The stiffness is associated with bilateral numbness, tingling in her arms and burning in her fingertips. When sitting for more than 15 minutes, she feels numbness in her gluteal region. At times, when she's not completely locked in position, her muscles twitch. She complains of stiffness throughout her spine. She experiences loss of balance, and describes it as if she's stepping off a boat. As per the patient the muscle spasms were diffuse, starting from the nape of the neck, shoulders and back. She stated the pain as pricking which is diffusely distributed throughout the body. She also has difficulty sleeping and she has difficulties in bending and turning. She also has autonomic symptoms like urinary urgency and constipation. She stated that pain is exacerbated by constant posture and its not relieved by naproxen and oxycodone. It is

difficult to maintain her relationship with her husband as she couldn't be involved in an intimate relationship.

While she describes her symptoms, the patient is stiff and moves like a block, with no trunk axial twisting. She initially refuses to sit due to her stiffness and prefers to explain her symptoms while standing.

She also reported arm and leg weakness, difficulty lifting her leg while walking, and dragging her feet at times. The weakness is intermittent and has a variable duration ranging from a few hours to a few days. Even doing the dishes is difficult for her due to her poor hand grip. She's kept a journal for years, and her weakness appears to be consistent. She describes her weakness as more peripheral than proximal. She had physical therapy for her neck and lower back, and she still does the exercises. She enjoys exercising and continues to do so, but it is more difficult.

The patient was not in acute distress. The vitals are as follows: BP 155/91, Pulse 76/min regular, Weight 147lbs, RR 16/min and afebrile. On neurological examination, there were painful spasms of the shoulder and back, as well as stiffness of the limb muscles, with no other neurological signs. Her deep tendon reflexes in both limbs were normal, no pathological reflexes were elicited, distal muscle strength was decreased more than proximal muscle and her muscle tone was 3/5 in both upper and lower extremities. Aside from her presbyopia glasses, the patient's ocular examination was normal. The patient had normal coordination, no discernible dysdiadochokinesia, dysmetria, or gait abnormalities.

A complete blood count, metabolic panel, thyroid studies, Vitamin B12, Vitamin D, folate, anti-nuclear antibodies, and cretinitine kinase levels were all ordered. Furthermore, confirmation of antibodies against GAD is ordered. An MRI of the cervical spine six months ago revealed no abnormalities. An EMG and nerve conduction studies are also planned.
Clonazepam and Baclofen have been prescribed to the patient.

Discussion

The differential diagnosis for SPS is broad, and it includes disorders of the brain, spinal cord, and muscles:

- Myelopathy: compressive, inflammatory, infectious, ischemic
- Myopathies and muscular dystrophies
- Idiopathic Parkinson's disease and Parkinson-plus syndromes
- Autoimmune encephalitis
- Primary lateral sclerosis
- Progressive multiple sclerosis
- Generalized or focal dystonia
- Neuromyotonia
- Isaac syndrome
- Ankylosing spondylitis
- Hereditary spastic paraplegia
- Hereditary hyperekplexia
- Leukodystrophies
- Neuroleptic malignant syndrome, serotonin syndrome, or malignant hypothermia
- Tetanus
- Functional neurological disorder [1].

The known incidence of SPS is 1 case per million per year, with an estimated prevalence of 1 to 2 cases per million, even though it is most likely underreported [3]. Cases with onset in childhood are unusual, with less than 15 GAD-seropositive cases reported [4]. If SPS is not promptly treated, it frequently goes undiagnosed or is incorrectly diagnosed, which can result in physical and mental health that is permanently compromised. As a result, healthcare professionals should try to recognize and treat the disease as early as feasible, under the guidance of a neurology specialist. In order to improve their chances of recovery and maintain their quality of life, patients with SPS would benefit from being managed by an interprofessional team that included a physical therapist, psychiatrist, and orthopedic surgeon. It is frequently associated with other autoimmune diseases like pernicious anemia, myasthenia gravis, Type 1 Diabetes mellitus, etc. It may be a paraneoplastic manifestation in breast cancer and Hodgin's lymphoma [5]. Antibodies against glutamic acid are found in 60-75% of patients [1].

Conclusion

A 57-year-old woman presented with diffuse painful muscle spasms; physical examination revealed diffuse stiffness of the limb muscles with shoulder spasms. On further evaluation, Anti-GAD antibodies confirmed her diagnosis.

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