Case Report

Status spasticus and psoas muscle edema due to anti-GAD antibody associated stiff-man syndrome

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Abstract

Severe muscle rigidity and spasms are uncommon causes of Intensive Care Unit (ICU) admissions. Stiff-man syndrome (SMS) is a rare disorder characterized by continuous muscle spasms, axial muscle rigidity, “tin soldier gait,” and continuous motor unit activity on electromyography. There are three clinical variants of SMS: stiff-limb syndrome, classical SMS, and paraneoplastic encephalomyelitis with rigidity and myoclonus. Three types of antibodies have been associated with SMS; however, anti-glutamic acid decarboxylase (GAD) antibodies are the most frequent and are seen in the idiopathic type of SMS. The spasms of SMS can be very disabling and severe enough to cause muscle ruptures and skeletal fractures. We present a case of anti-GAD positive SMS with “status spasticus” causing bilateral psoas myoedema and rhabdomyolysis due to repeated axial muscle jerking in a 64-year-old man and discuss the differential diagnosis of a “jerking patient in the ICU.”

Keywords: Psoas myoedema, rhabdomyolysis, status spasticus, stiff-man syndrome

Introduction

Severe muscle rigidity and muscle spasms or “jerking” can necessitate Intensive Care Unit (ICU) admission. A large number of cases can present with these signs and delayed diagnosis can have catastrophic consequences. Stiff-man syndrome (SMS) is a rare autoimmune disorder of the central nervous system characterized by predominant axial rigidity and spasms along with continuous motor unit activity (CMUA) on electromyography (EMG). Four types of antibodies are associated with SMS. High titer IgG antibodies against glutamic acid decarboxylase (GAD65) are usually associated with idiopathic SMS, anti-amphiphysin antibodies are associated with paraneoplastic SMS and glycine receptor antibodies are associated with paraneoplastic encephalomyelitis with rigidity and myoclonus (PERM).[1] Gephyrin antibodies have also been associated with SMS.[2]

Normally, spinal interneurons inhibit spontaneous discharges from spinal motor neurons through the neurotransmitter glycine. Other inhibitory inputs for the motor pathway include GABA-mediated inhibition from the cortex, brain stem, and cerebellum. If GAD function is inhibited significantly as in SMS then disinhibition results in continuous muscle stimulation by the motor neurons leading to the characteristic pattern of CMUA on EMG.

In some patients with SMS, severe spasms can cause skeletal muscle rupture, bone fracture, and joint dislocations (status spasticus). We present a case of status spasticus with psoas myoedema and rhabdomyolysis in anti-GAD positive SMS.

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Case Report

A 64-year-old man with a 30 years history of diabetes mellitus presented with severe back pain and leg spasms of 5 months duration. On examination, he was diaphoretic and tachycardic. There was persistent lumbar hyperlordosis in the supine position with lumbar paraspinal muscle hypertonia. Periodic bilateral leg spasms and anterior abdominal muscle jerks (10/min) were observed [Video 1 in online version]. Limb muscle tone was increased, and reflexes were normal. Computed tomography scan of the LS spine done 3 days earlier (at an outside hospital) was normal [Figure 1]. He was started on oral diazepam (30 mg/day) and baclofen (20 mg/day). A repeat magnetic resonance imaging (MRI) of the spine and brain (short tau inversion recovery sequences) showed high signal suggestive of muscle edema in the iliopsoas muscles [Figure 2]. Creatinine phosphokinase levels were elevated [16,000 U/l], and urine was positive for myoglobin. He was hydrated with intravenous (IV) fluids and started on continuous diazepam IV infusion (up to 1–2 mg/h). Nevertheless, the jerks persisted and he developed exhaustion and respiratory depression, requiring intubation and mechanical ventilation [Figure 1]. A differential diagnosis of tetanus, rabies, poisoning, and SMS with status spastics were considered. Vecuronium was used at a dose of 1–2 mg/h to quell the spasms. IV immunoglobulin (IVIG) was then administered at a dose of 2 g/kg over 5 days. He received approximately 160 mg of vecuronium over 5 days after which it was gradually discontinued. He required a continuous diazepam infusion of 0.1–0.4 mg/h for 3 weeks, 30 mg oral diazepam/day, and Gabapentin 900 mg/day to control his spasms. Renal functions remained stable. At 4 weeks, serum anti-GAD 65 antibody titers were positive at >1000 μ/ml (Neuroimmunology Laboratory, Oxford, UK), substantiating the diagnosis of “jerking SMS.” He was gradually weaned off the ventilator after 3 weeks and he was transitioned to 20 mg of diazepam PO. At follow-up 6 months later, he was totally asymptomatic and off medications.

Discussion

The differential diagnosis of a patient who has muscle rigidity and “jerking” in the ICU includes tetanus, status dystonicus (dystonic storm), status spasticus due to SMS, serotonergic syndrome, neuroleptic malignant syndrome, etc. [Table 1]. A thorough evaluation is necessary to identify the cause early to prevent morbidity and mortality. In some cases, patients have a clear sensorium as in tetanus and strychnine poisoning. In others, the sensorium may be clouded as in the drug-induced conditions, encephalitis, paraneoplastic conditions, and envenomations.

SMS is an unusual neuromuscular disorder with striking clinical manifestations. The prominent clinical manifestations include proximent axial muscle rigidity, lumbar hyperlordosis, muscle spasms, and CMUA on EMG. Three clinical variants of SMS are described; stiff-limb syndrome (a focal variant of SMS), the jerking SMS (the generalized variant), and a PERM; a rapidly progressive condition characterized by fulminant rigidity and death over 4–6 months. The last 2 conditions can necessitate ICU admission. Many patients with SMS are quite disabled and require a combination of drugs to achieve disease control. Episodic spasms and falls remain a constant threat and a “tin soldier gait” is common. Spasms lasting for days are termed “status spasticus” and can lead to exhaustion and skeletal or muscle

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Table 1: Differential diagnosis of muscle jerking and rigidity in the ICU patient

| Condition                              |
|----------------------------------------|
| Tetanus                                |
| Strychnine poisoning                   |
| Serotonergic syndrome                  |
| Neuroleptic malignant syndrome         |
| Encephalitis                           |
| Status Spastics (Stiff man syndrome)   |
| Status dystonicus                      |
| Rabies                                 |
| Black Widow Spider Venom (Latrotoxin)  |
| Malignant catatonia                    |
| Intrathecal Baclofen withdrawal syndrome|
| Parkinsonism-dyskinesia-hyperpyrexia syndrome |

ICU: Intensive care unit

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Figure 1: Computed tomography abdomen; normal muscle intensities

Figure 2: Panel A; magnetic resonance imaging coronal images showing hyperintensities in both iliopsoas muscles suggestive of myoedema
injury as in status dystonicus. Neuromuscular blockers and mechanical ventilation may be required in status spasticus to prevent muscle rupture, muscle hematomas, rhabdomyolysis, or bone and joint injuries.[3,4]

The rigidity and “jerks” are treated with GABAergic drugs such as benzodiazepines, baclofen or vigabatrin, IVIG, and plasmapheresis or rituximab.[5] In exceptional cases, intrathecal baclofen pumps have been implanted.[6] In conclusion, our patient had a jerking SMS and “status spasticus” associated with anti-GAD antibodies and hitherto undescribed MRI features such as bilateral psoas muscle edema. The peculiar muscle involvement on MRI should be a pointer toward axial muscle predilection and guide evaluation and therapy.

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Conflicts of interest
There are no conflicts of interest.

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