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Stiff-person syndrome: Commonly mistaken for hysterical paralysis

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Dear Sir

Stiff-person syndrome (SPS) is a rare neurologic disease with dramatic manifestations. The condition is thought to have an autoimmune etiology. Associated neuropsychiatric symptoms of SPS cause it to be mistaken for hysterical paralysis, a conversion disorder which presents with voluntary motor deficits that suggest a neurological or general medical condition. However, after thorough evaluation, the symptoms are found to be unrelated to a physical cause [1].

Important differences between the presentation of SPS and hysterical paralysis must be identified to initiate appropriate therapy. SPS generally has a progressive course, while conversion disorders rarely persist after 1 year [2]. Additionally, the presence of autoantibodies against glutamic acid decarboxylase (GAD) is commonly used as a diagnostic marker for SPS [3]. Following diagnosis with SPS, a patient may be treated with intravenous immunoglobulin (IVIg) therapy, baclofen, and benzodiazepines [4].

We present a case of a 29-year-old black woman who presented with progressive rigidity of the lower extremities that had occurred over the course of a year. She was previously diagnosed with hysterical paralysis of the lower extremities. Her rigidity worsened during several months of observation, eventually encompassing her trunk, neck and upper extremities. Tests measuring creatine kinase, toxins, and heavy metals yielded negative results. Physical exam revealed 1+ reflexes throughout, and gross rigidity with slight lordosis. Bladder and bowel functions were normal. Results of multiple imaging studies of the brain and lower spine were negative. A psychiatric consultation concluded that there was little evidence supporting a functional origin for her symptoms.

At this point, a reevaluation of her symptoms led to a suspected case of SPS. A test measuring GAD antibodies was elevated at 62 U/ml (normal: <1). The diagnosis was confirmed by neurology consult. The patient began treatment with baclofen, diazepam and bi-annual IVIg therapy. Over several months, the patient showed marked improvement.

The rarity of SPS contributes to its common misdiagnosis as a functional disorder. On the other hand, many case studies of patients with SPS have been released recently, some including groups of over 20 patients [5]. For this reason, SPS is an important diagnostic consideration for a patient thought to have hysterical paralysis.

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Stiff-person syndrome in a female patient with type 1 diabetes, dermatitis herpetiformis, celiac disease, microcytic anemia and copper deficiency
Just a coincidence or an additional shared pathophysiological mechanism?

Sir,

We have read with great interest a recent paper published in your journal of a patient suffering from stiff-person syndrome (SPS), type 1 diabetes mellitus (DM), dermatitis herpetiformis (DH) and celiac disease (CD). We want to report a patient with the same clinical presentation, also diagnosed with microcytic anemia with normal iron level and decreased copper level in serum [1].

Our patient with SPS was admitted because of pain and spasms in back and right leg. The serum GAD IgG titer was 1548 μg/mL. She had been suffering from type 1 DM and from frequent diar-