


Stiff-person syndrome with rhabdomyolysis

Toshiyuki Nakanishi,¹ Tsuneaki Kenzaka ^{2,3}

¹Department of Internal Medicine, Nerima Hikarigaoka Hospital, Nerima-ku, Tokyo, Japan

²Division of Community Medicine and Career Development, Kobe University Graduate School of Medicine, Kobe, Hyogo, Japan

³Department of Internal Medicine, Hyogo Prefectural Tamba Medical Center, Tamba, Hyogo, Japan

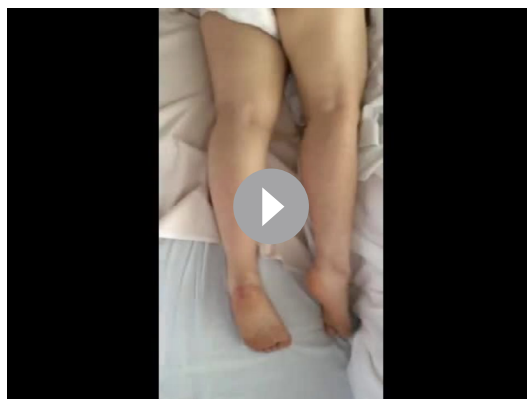
Correspondence to
Dr Toshiyuki Nakanishi;
r060eb@gmail.com

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DESCRIPTION

A 52-year-old woman presented with episodic stiffening of her back and thighs every 3 months for 5 years. The sudden stiffness had worsened and resulted in multiple falls 3 years before admission. She became shut-in due to fear of falling and being unable to walk without using a cane. She was alert, although episodic spasms were induced repeatedly by sudden touch, noise and light. As she was on a selective serotonin reuptake inhibitor (SSRI) regimen and had a fever, diaphoresis and elevated serum creatine kinase (CK) (16 200 U/L), a working diagnosis of neuroleptic malignant syndrome (NMS) complicated by rhabdomyolysis was made, necessitating referral to our care facility for further examination. Physical examination revealed severe lumbar lordosis and muscle stiffness in her lower limbs, bilaterally ([video 1](#)). Blood tests revealed an extremely high level (>2000 U/mL) of anti-glutamic acid decarboxylase antibodies, and her cerebrospinal fluid contained oligoclonal bands. These findings led to the final diagnosis of stiff-person syndrome (SPS). Initially, under the tentative diagnosis of NMS, the patient had not showed any improvement with SSRI cessation and intravenous dantrium. However, after the SPS diagnosis was made, she was given plasma exchange and intravenous immune globulin, and subsequently commenced on oral diazepam, which was tapered gradually. Finally, she was able to walk without any difficulty or anxiety.

SPS is an adult-onset central nervous system disorder caused by an autoimmune reaction.¹ In SPS, episodic spasms are induced by sudden stimuli, which cause progressive muscle stiffness of the limbs and trunk, resulting in impaired ambulation.² In the early stages of SPS, the spasms can cause lumbar lordosis and falls, which often lead to social phobia and anxiety.³ If the patient has been taking psychiatric medications, clinicians may misdiagnose the



Video 1 Episodic spasms in stiff-person syndrome.

Patient's perspective

I appreciate the fact that I was finally diagnosed and received definitive treatment. I asked the doctor, Toshiyuki Nakanishi, to spread the clinical knowledge of this rare disease, stiff-person syndrome, and strongly believe that many of those who are suffering from stiff-person syndrome would also be diagnosed and treated properly.

Learning points

- ▶ Stiff-person syndrome (SPS) is often misdiagnosed as a psychiatric disorder, such as neuroleptic malignant syndrome or serotonergic syndrome, if an affected patient has been taking psychiatric medication.
- ▶ SPS should be considered for patients who have muscle spasms, in the presence of anti-glutamic acid decarboxylase antibodies.
- ▶ This case illustrates the importance of checking other parameters in addition to creatine kinase levels.

condition as a psychiatric disorder, such as NMS or serotonin syndrome (SS). Although serum CK levels can be elevated in the late stages of SPS, because the muscle spasms can be severe enough to cause muscle breakdown,⁴ a distinguishing indicator for SPS diagnosis is that serum CK levels are usually lower than that in other aforementioned disorders. To the best of our knowledge, only two other cases of SPS with concurrent rhabdomyolysis have been reported.^{4,5}

However, as in this case, SPS can mimic NMS and SS in that serum CK levels may be highly elevated. Based on a literature review, the median peak CK level was 10 888 U/L in patients with SS (n=60) and 15 059 U/L in patients with NMS (n=74), compared with 16 200 U/L in our patient with SPS.⁶

Even in such cases, SPS should be considered when patients present with the characteristic muscle stiffness and spasms.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

ORCID iD

Tsuneaki Kenzaka <http://orcid.org/0000-0002-3120-6605>

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