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Case report

A case of childhood stiff-person syndrome with striatal lesions: A possible entity distinct from the classical adult form

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Abstract

Parainfectious or autoimmune striatal lesions have been repeatedly described in children. We report a 7-year-old girl with painful muscle spasms, leading to the diagnosis of childhood stiff-person syndrome (SPS). Striatal lesions were demonstrated by diffusion-weighted magnetic resonance imaging (MRI) and single-photon emission computed tomography but not by conventional MRI. Autoantibodies against glutamic acid decarboxylase (GAD) were absent. Steroid pulse therapy and high-dose intravenous immunoglobulin resolved all the symptoms with slight sequelae. Childhood SPS may be characterized by absent anti-GAD antibodies and a transient benign clinical course, and it may have a pathomechanism distinct from that in adult SPS.

Keywords: Child; Diffusion-weighted imaging; MRI; SPECT; Stiff-person syndrome; Striatum

1. Introduction

Parainfectious or autoimmune striatal lesions have been repeatedly described in children and are triggered by various infectious agents, including viruses and *Mycoplasma pneumoniae* [1]. This category includes some specific entities such as pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections [2] and encephalitis lethargica syndrome [3]. Patients show a wide range of neuropsychiatric symptoms: lethargy, chorea, bradykinesia, rigidity, mutism, depression, and obsessive–compulsive behaviors.

We report a girl exhibiting obsessive–compulsive behaviors, muscle rigidity, and lethargy, as well as painful episodic muscle spasms, leading to the diagnosis of childhood stiff-person syndrome (SPS). Striatal lesions were demonstrated by diffusion-weighted imaging (DWI) of magnetic resonance imaging (MRI) and single-photon emission computed tomography (SPECT) but not by conventional MRI such as T1- and T2-weighted images. We discuss the possible involvement of striatal lesions in childhood SPS.

2. Case report

A 7-year-old girl was admitted to a hospital because of intermittent pain in her left thigh and calf and diffi-

Abbreviations: DWI, diffusion-weighted imaging; GAD, glutamic acid decarboxylase; SPECT, single-photon emission computed tomography; SPS, stiff-person syndrome.

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culty in walking for 3 days. Around 10 days before admission, she coughed. Cerebral and spinal MRI and cerebrospinal fluid study revealed no abnormalities. She showed episodic painful muscle spasms lasting around 30–60 min in the back and the bilateral but left-predominant lower limbs several times a day. On day 4 of illness, she also exhibited repetitive behaviors: repeatedly grasping alternate sides of the bed guards with her right hand during wakefulness.

The patient was transferred to our hospital on day 7 of the illness. She was alert, and muscle tonus and deep tendon reflexes were normal with no pathological reflexes. The spasms occurred spontaneously or were precipitated by emotional upset or physical stress such as blood puncture. Except during the spasm attacks, most of the time she showed the repetitive grasping behavior, which was relieved transiently by distracting stimuli such as commands during neurological examination. When she was asked, "Why do you grasp the bed guards again and again?" she said, "I feel the urge to do so". All the symptoms and behaviors ceased during sleep.

Routine hematological tests, serum chemistry, and a cerebrospinal fluid test were unremarkable. Serum autoantibodies including antinuclear and anti-glutamic acid decarboxylase (GAD) antibodies were all negative. Infection of Group A beta-hemolytic streptococcus was excluded by rapid antigen testing, throat culture, and serum antibodies. T1- and T2-weighted and fluid-attenuated inversion recovery MRI of the brain and spinal cord did not disclose any abnormalities. However, DWI of MRI and SPECT revealed bilateral lesions in the striatum (Fig. 1A, C). Electroencephalography revealed an occipital dominant alpha wave with intermittent, generalized, bilaterally synchronous bursts of 2–3 Hz delta waves with a duration of 1 s and identified no specific alteration during the spasm.

Each episode of the painful spasms was treated by intravenous diazepam (0.5–1 mg/kg) with considerable

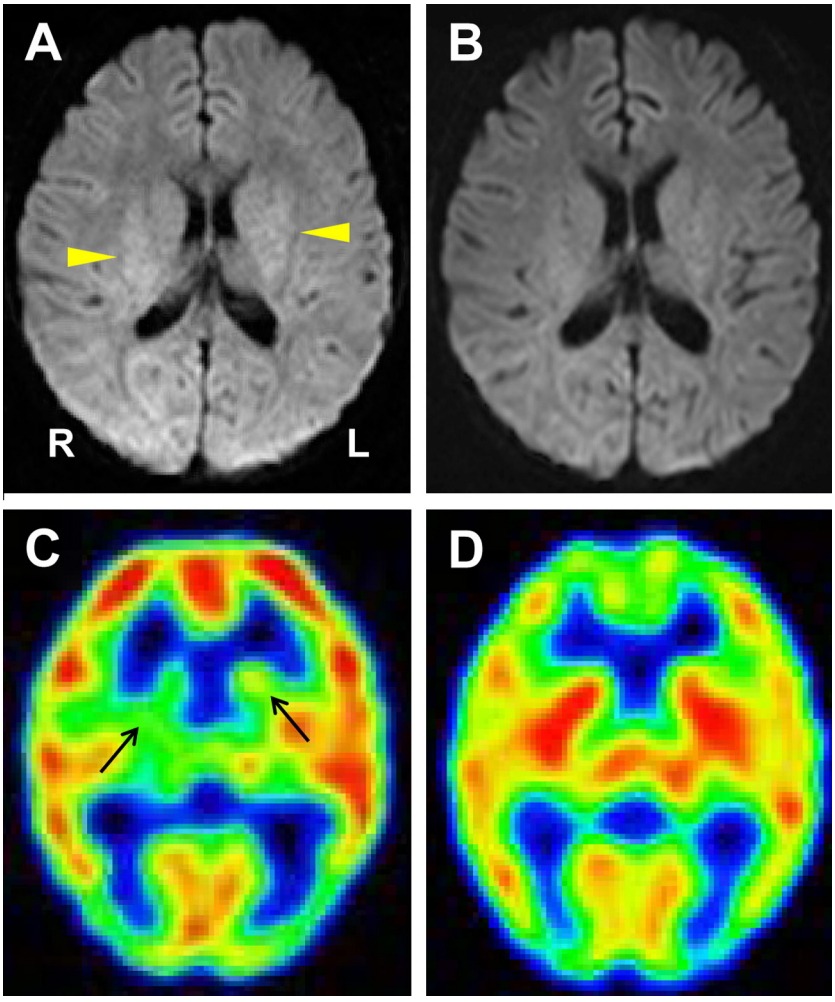

Fig. 1. Neuroimaging studies on day 11 (A, C) and around a month (B, D) of illness. Diffusion-weighted imaging (b value = 2000 s/mm^2) demonstrates right-dominant bilateral lesions in the striatum (A), which resolves thereafter (B). Single-photon emission computed tomography reveals hypoactivity in right-dominant bilateral lesions in the striatum (C). The lesions improve, but diffuse hypoactive areas emerge in the bilateral frontal lobes (D).