Brain & Development 39 (2017) 252–255

BRAIN & DEVELOPMENT
Official Journal of the Japanese Society of Child Neurology

www.elsevier.com/locate/braindev

Case Report

A pediatric patient of hemorrhagic acute transverse myelitis

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Received 21 April 2016; received in revised form 12 September 2016; accepted 12 September 2016

Abstract

An 11-year-old boy presented with progressive leg hypesthesia but no history of trauma. Dysuria and constipation appeared subsequent to gait difficulty. He was admitted 8 days after onset. Spinal magnetic resonance imaging (MRI) revealed longitudinal hyperintensity with cord swelling and hypointensity on T2-weighted images, suggesting severe inflammation and microbleeding change, respectively. Gadolinium contrast-enhanced MRI demonstrated mild enhancement in the lesions. Platelet count and coagulation findings were normal, and cerebrospinal fluid analysis showed no pleocytosis. He was diagnosed with idiopathic acute transverse myelitis (ATM), and intravenous methylprednisolone pulse therapy and plasmapheresis were initiated. On day 14, motor dysfunction aggravated suddenly, accompanied by expanding hemorrhagic lesions. Thereafter, administration of intravenous immunoglobulin, repeated intravenous methylprednisolone pulse therapy and prednisolone for one month resulted in complete recovery four months later. Both anti-aquaporin-4 and anti-myelin oligodendrocyte glycoprotein antibodies were negative. We presented the first pediatric case showing hemorrhagic spinal lesions in the clinical course of ATM. This severe complication should be recognized in the management of ATM.

Keywords: Spinal hemorrhage; Myelitis; Treatment; Children; Plasmapheresis

1. Introduction

Acute transverse myelitis (ATM) is caused by local inflammation of the spinal cord, and it manifests motor paralysis, sensory disturbance, and dysautonomia corresponding to the location of the lesion [1,2]. It is diagnosed by excluding various other diseases that may cause spinal cord symptoms based on the time course of clinical symptoms, cerebrospinal fluid test findings, and contrast magnetic resonance imaging (MRI) findings [2]. On the other hand, the causes of spinal hemorrhage include vascular malformation, spinal cord infarction, and tumors, but it is atypical in inflammatory and demyelinating diseases.

2. Case report

The patient was an 11-year-old boy. He had a medical history of idiopathic thrombocytopenic purpura at 6 years old. His mother had a medical history of Sjogren's syndrome. He had received no vaccination within

Abbreviations: ATM, acute transverse myelitis; MRI, magnetic resonance imaging; MMT, manual muscle test.

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http://dx.doi.org/10.1016/j.braindev.2016.09.007

the 3 months prior to the onset of the disease, nor was there any preceding infection or trauma. He presented with muscle weakness of the bilateral lower limbs and gradually progressed to walking difficulty. On the 8th day of illness, difficulty in urination and constipation appeared, and the patient was admitted. On physical examination, consciousness was clear; body temperature, 36.1 °C; heart rate, 80/min; blood pressure, 110/69 mmHg; and SpO_2, 100% (room air) without respiratory disturbance. No abnormality was detected in the cranial nerve system nor fundus. No motor paralysis, sensory disturbance, nor abnormal deep tendon reflex was noted in either upper limb. Motor paralysis (manual muscle test: MMT, 3/5) was observed in the bilateral lower limbs and he could not walk independently. The deep tendon reflex was weak in the bilateral lower limbs. There were sensory disturbance of the 6th thoracic spinal cord or lower as well as bladder and rectal disturbance. On plain head MRI, there were no clear abnormal findings. Whole spine MRI revealed swelling of the C5-Th2 spinal cord, and there were laterally symmetric high-intensity regions corresponding to the gray matter in C4-Th10 on T2-weighted imaging (Figs. 1 and 2; A). Gadolinium contrast imaging revealed mild contrast enhancement. In addition, low-intensity regions were scattered in the Th3-Th4 high-intensity regions on T2-weighted imaging (Fig. 2-A; a–c). There was no clear flow void. On blood testing on admission, the platelet count and coagulation function were within the normal ranges, and anti-nuclear, anti-ds-DNA, anti-ss-A, and ss-B antibodies were negative. On cerebrospinal fluid testing, the cell count was 2/uL; protein, 26 mg/dL; lactic acid, 15 mg/dL; and pyruvic acid, 0.8 mg/dL; showing no elevation. Myelin basic protein was 475.8 pg/mL (normal range 102.0>) and neopterin was 22 ng/mL (normal range 20>), showing mild elevations, and oligoclonal bands were negative. No atypical cells were noted on cytology of the cerebrospinal fluid. Polymerase chain reaction tests for herpes simplex virus type 1and 2, human herpes virus 6 and 7, cytomegalovirus, Epstein-Barr virus and varicella-zoster virus in the plasma and cerebrospinal fluid were negative.

He was diagnosed as acute idiopathic transverse myelitis. Intravenous methylprednisolone pulse therapy (30 mg/kg over 3 days) was initiated, but motor paralysis did not improve, and sensory disturbance progressed to the 4th thoracic spinal cord. Plasmapheresis was started from the 13th day of illness. He received albumin replacement for 2 consecutive days, but motor paralysis of the lower limbs aggravated (MMT 2/5) on the 14th day. As he exhibited hypofibrinogenemia (<50 mg/dL or lower), the replacement solution was changed to fresh

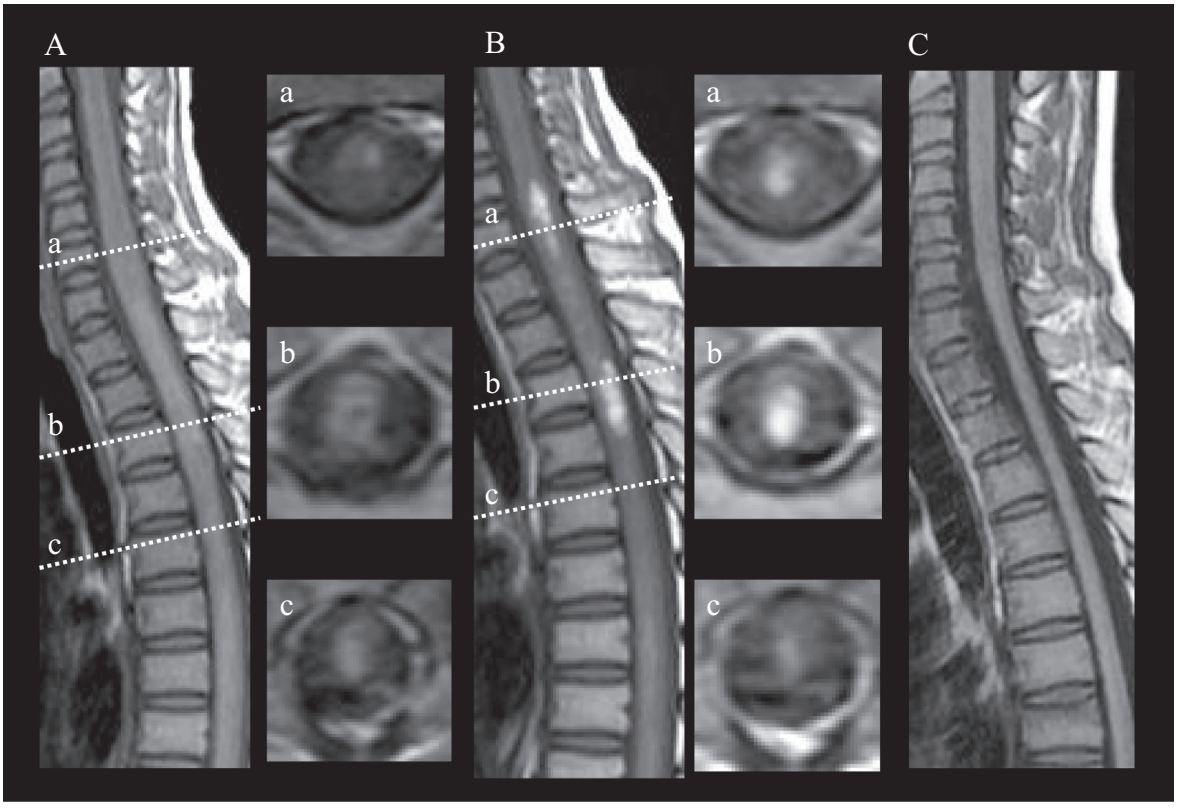

Fig. 1. Serial MR images of T1-weighted images. A: At admission, longitudinally extensive cord lesions mainly involving the gray matter with mild hyperintensity (a–c). B: On day 20, very high intensity lesions showing expanding hemorrhagic lesions (a, b). C: 6 months later after the onset, no signal changes and mild atrophy.