ELSEVIER

Brain & Development 34 (2012) 380–383

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

www.elsevier.com/locate/braindev

Case report

A case of ADEM with atypical MRI findings of a centrally-located long spinal cord lesion

Yukifumi Monden [a], Takanori Yamagata [a,*], Yuri Kuroiwa [a], Toshiyuki Takahashi [b], Masato Mori [a], Tokiko Fukuda [a], Hideo Sugie [a], Mariko-Yoshida Momoi [a]

[a] Department of Pediatrics, Jichi Medical University, Japan
[b] Department of Neurology, Tohoku University School of Medicine, Japan

Received 18 February 2011; received in revised form 30 May 2011; accepted 20 June 2011

Abstract

The patient was a 14-year-old male diagnosed with acute disseminated encephalomyelitis (ADEM) with acute onset of multifocal central nervous system symptoms. He showed increased cerebrospinal fluid cell counts and high myelin basic protein levels, which responded well to steroid pulse therapy. Spinal MRI showed a centrally-located long spinal cord lesion (LCL) involving 17 vertebral bodies from C2 to T11 that later expanded into the white matter, and lesions on the ventral side of the medulla. The cause of LCL has been reported to be heterogeneous. In this case, LCL is considered to be associated with ADEM, an acute autoimmune response to myelin, and vascular inflammation of the gray matter of the spinal cord.

Keywords: ADEM; Centrally located long spinal cord lesion (LCL); NMO

1. Introduction

A centrally-located long spinal cord lesion (LCL) is defined as a lesion that involves more than three vertebral bodies, located in the central area of the spinal cord on MRI images. LCL has attracted attention since being reported as a characteristic MRI finding in cases of neuromyelitis optica (NMO) [1–3]. The underlying diseases associated with it vary widely, however, and include not only NMO but also infections, tumors, vascular diseases, and autoimmune diseases. LCL without white matter lesions in patients with acute disseminated encephalomyelitis (ADEM) has never been reported. Lesions are typically asymmetric and variable in number and size in ADEM [4]. We report a pediatric case of a male with an ADEM who showed LCL and bilateral lesions on the ventral side of the medulla after an infection.

2. Case report (Fig. 1)

The patient was a 14-year-old male. A few days after an upper respiratory infection, he developed acute lower back pain, weakness and numbness of both legs, and a feeling of residual urine, resulting in difficulty in walking and urinary retention over 7 days. At 7 days from onset, his height was 168 cm (+1.0 SD), and weight was 50.0 kg (−0.2 SD). Vital signs were normal. He was fully conscious. He presented no nuchal rigidity. On neurological examination, cranial nerves were intact. A manual muscle test (MMT) of the four limbs revealed grade 5/5. Grasping power had bilaterally decreased (25/25 kg) compared with 3 months earlier (32/32 kg). Deep tendon reflexes were normal in the upper and lower limbs. Cerebellar sign was not observed, but

* Corresponding author. Address: Department of Pediatrics, Jichi Medical University, 3311-1 Yakushiji, Shimotsuke-city, Tochigi, Japan. Tel.: +81 285 58 7710; fax: +81 285 44 8329.
E-mail address: takanori@jichi.ac.jp (T. Yamagata).

0387-7604/$ - see front matter © 2011 The Japanese Society of Child Neurology. Published by Elsevier B.V. All rights reserved.
doi:10.1016/j.braindev.2011.06.010

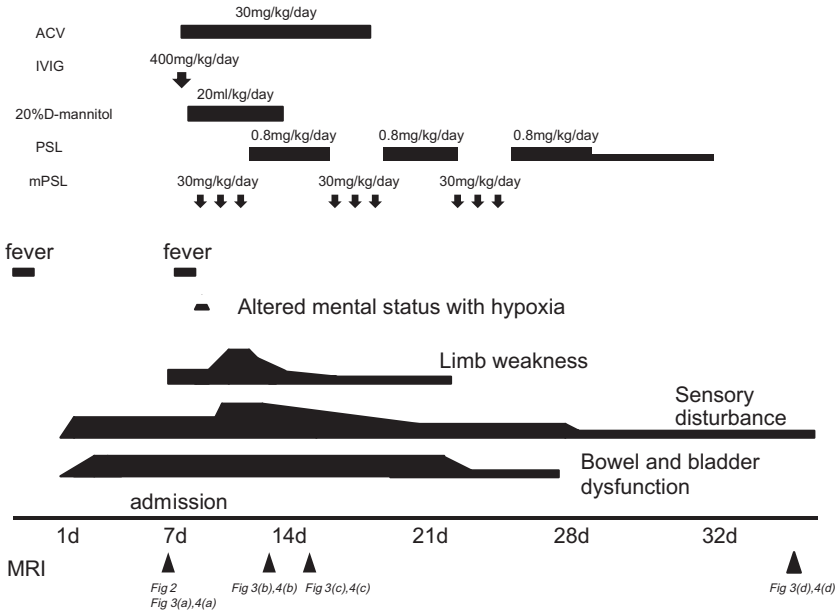

Fig. 1. Clinical course of the patient.

tactile sensation and proprioception were reduced in the lower bilateral extremities under the T10 level. He presented urinary retention and bowel dysfunction, and the cremasteric reflex was not detected.

Laboratory results were as follows: white blood cell count, 16,200/μl; C-reactive protein, 0.24 mg/dl; erythrocyte sedimentation rate, 27 mm/h. Anti-double-stranded DNA IgG antibody was 4.7 IU/ml (<10 IU/ml). Serum anti-aquaporin 4 (AQP4) antibody was negative. In the cerebrospinal fluid (CSF), cell count was 109/mm^3 (polycyte 6 mm^3, monocyte 103 mm^3), protein was elevated to 130 mg/dl, glucose was 66 mg/dl (serum glucose was 138 mg/dl), myelin basic protein (MBP) was over 2000 pg/ml (>102 pg/ml), and oligoclonal bands were negative, as were IgG index and AQP4. CSF culture was negative. Serum anti-*Mycoplasma* antibody and viral antibodies to *Human immunodeficiency virus*, *polio*, and *Varicella zoster virus* were all negative. *Herpes simplex virus* (HSV)-DNA was negative in the CSF by PCR, and *Epstein–Barr virus* was identified as having been a past infection. T2-weighted MRI (Fig. 2) showed high signal intensity in the central gray matter from C2 to T11 and lesions on the ventral side of the medulla and the pons that were not continuous with the spinal cord.

Intravenous acyclovir (ACV) injection (30 mg/kg/day for 7 days) and γ globulin (IVIG) administration (400 mg/kg/day for 2 days) were performed. Despite

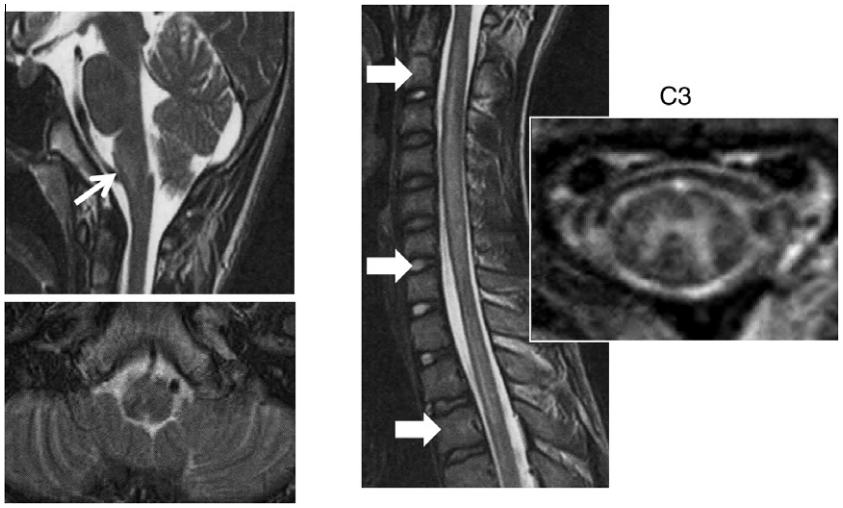

Fig. 2. T2-weighted MRI at 7 days from onset: high spinal lesion from C2 to T11, centering on the gray matter and lesions, not continuous with the spinal cord on the ventral side of the medulla.