Brain & Development 38 (2016) 875–879

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

www.elsevier.com/locate/braindev

Case Report

Acute encephalopathy with biphasic seizures and late reduced diffusion associated with staphylococcal toxic shock syndrome caused by burns

Takaoki Yokochi, Shinpei Sakanishi, Yuuki Ishidou, Go Kawano, Toyojiro Matsuishi *, Yukihiro Akita, Keizo Obu

Department of Pediatrics, St Mary's Hospital, Fukuoka, Japan

Received 5 January 2016; received in revised form 5 April 2016; accepted 7 April 2016

Abstract

We report a case of acute encephalopathy with biphasic seizures and late reduced diffusion (AESD) associated with toxic shock syndrome caused by burns.

A one-year-old girl was admitted to our hospital for treatment of severe burns. On day 3, she exhibited a fever, generalized rash and multiple organ failure. She was diagnosed with toxic shock syndrome after burns. She had seizures with fever twice on the same day, followed by secondary seizures on day 8 and transient deterioration of the gross motor functions involved in sitting alone and rolling over. On day 9, MRI diffusion-weighted images showed bright tree appearance (BTA).

We conclude that she developed AESD.

Keywords: Bright tree appearance (BTA); Acute encephalopathy with biphasic seizures and late reduced diffusion (AESD); Toxic shock syndrome (TSS); Burn; Methicillin-resistant *Staphylococcus aureus* (MRSA)

1. Introduction

The bright tree appearance (BTA) is evidence of acute encephalopathy with biphasic seizures and late reduced diffusion (AESD) in pathognomonic magnetic resonance imaging (MRI). BTA is a term for high-intensity lesions in the subcortical white matter shown on diffusion-weighted images. Most cases of AESD have prodromal virus infections and are characterized by febrile status epilepticus (FSE) on the first day, followed by transient recovery of consciousness. The secondary cluster of complex seizures occurs on days 3–6. At this stage, MRI shows BTA. Affected brains gradually atrophy, and the patients have various levels of neurological sequelae [1]. The pathogenesis of AESD has been suggested to be related to excitotoxic injuries, and BTA corresponded to enhancement of the delayed brain edema [2,3]. Recently, BTA was found to be associated with head injury [4]. However, no cases of BTA associated with staphylococcal toxic shock syndrome (TSS) caused by burns have been reported until now.

Here, we report a case in which burns caused complicating TSS and which consequently developed AESD.

* Corresponding author at: Department of Pediatrics, St. Mary's Hospital, 422 Tsubukuhonmachi, Kurume City, Fukuoka 830-8543, Japan. Tel.: +81 942 35 3322; fax: +81 942 34 3115.

E-mail address: tmatsu@st-mary-med.or.jp (T. Matsuishi).

http://dx.doi.org/10.1016/j.braindev.2016.04.003

2. Case report

A one-year-old girl was admitted to our hospital for treatment of severe burns. She had no previous illness, and her developmental milestones were normal. On day 1, a pot fell from a table, and boiled water splashed on the body. Water was used to cool her at home, and she was brought to our hospital by ambulance.

When she arrived at our hospital, her consciousness was clear; she exhibited a Glasgow Coma Scale of 15 (E4V5M6) without any neurological abnormalities. Her pulse rate was 124 per minute, her blood pressure was 110/72 mmHg, and her body temperature was 36.3 °C. Second-degree burns (partly deep dermal burns) were observed on her body and on her right upper and lower limbs. The burns covered 20% of her total body surface area. There were no signs suggesting abuse such as old or new intermingled traumas or fractures other than the burns.

Laboratory examination on admission was normal.

Because the area covered by second-degree burns was extensive, she was administered Ringer's solution according to the Parkland formula and her injured skin was treated with ointment. An H_2 blocker was also administered to prevent Curling ulcer. Her general condition was stable until day 2. However, on day 3, she exhibited a fever, watery diarrhea and generalized morbilliform rash (Fig. 1). She also exhibited tachycardia and peripheral circulatory failure, and rapidly developed pre-shock although her blood pressure was stable. Therefore, TSS subsequent to the burns was suspected. The administration of vancomycin was started because the participation of *Staphylococcus aureus* was considered. Gabexate mesilate was also started, because the patient fell into a state of disseminated intravascular coagulation (DIC). On the same day, she twice had generalized tonic–clonic seizures, each lasting for 10 min. The visible seizures were controlled rapidly by diazepam; however, she took 6 h to completely awaken after the first seizure and 10 h after the second seizure. Cranial computed tomography (CT) was performed and showed normal results. Cerebrospinal fluid examination revealed normal findings. Therefore, her seizures were judged to be a transient event associated with her febrile condition. Her fever continued and laboratory results on day 4 revealed lymphopenia, elevation of C reactive protein, anemia, thrombocytopenia, elevation of hepatic enzyme, coagulation disorder, metabolic acidosis, and elevation of cytokines such as interleukin-6 (IL-6) and interleukin-2 receptor (IL2-R) (Table 1). Methicillin-resistant *S. aureus* (MRSA) was also detected a few days later in a bacterial cultivation from the injured skin, and TSS toxin-1 (TSST-1)-producing bacteria were identified. The ratio of T-cell-receptor Vβ2 (TCRVβ2)-positive T cells to CD4-positive T cells as determined by a flow cytometer was reduced to 0.4% on day 4. The proportion expanded to 19.7% on day 26 (usually a ratio over 14.9% is considered an expansion). The ratio of CD45RO-positive T cells to TCR Vβ2-positive T cells was also elevated to 75% (usually a ratio over 11.1% is considered an elevation). Therefore, TSST-1 was considered to play an important role in her condition. Bacterial cultivations from sources other than the injured skin were negative.

Thereafter, her fever eased, and her rash started to disappear on day 6. Her symptoms and laboratory results then gradually improved. However, she was continuously drowsy and displeased after the seizures.

On day 8, she had generalized seizures three times, with each seizure lasting for 30 s to 1 min. Her consciousness level rapidly recovered each time, and the cranial CT findings were normal. However, her gross motor function was deteriorated, and she could not maintain a sitting position. On day 9, MRI diffusion-weighted images showed high intensity in the subcortical white matter of the bilateral parietal lobes and in some parts of the cerebral cortex of the bilateral temporal and occipital lobes, and the apparent diffusion coefficients (ADC) in the lesions were reduced; in other words, the cranial MRI findings showed BTA (Fig. 2). The sleep electroencephalograph findings on the same

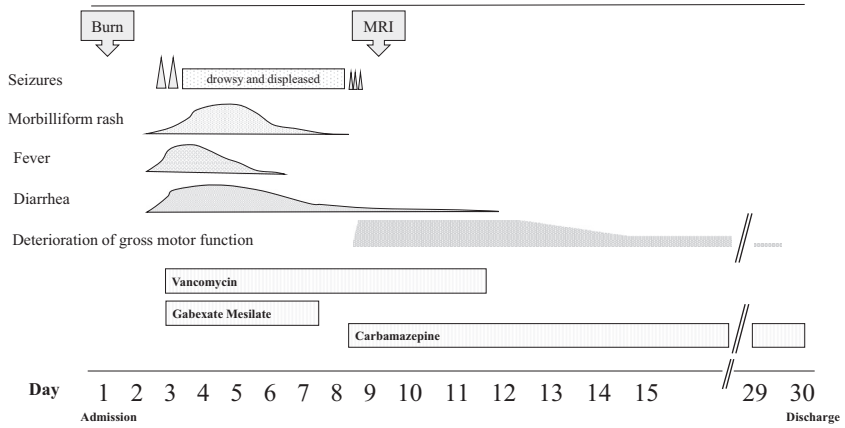

Fig. 1. Clinical course.