The Journal of Emergency Medicine, Vol. 38, No. 3, pp. e1–e4, 2010

0736-4679/07 $–see front matter

ELSEVIER

doi:10.1016/j.jemermed.2007.06.020

Clinical Communications: Adults

REVERSIBLE POSTERIOR LEUKOENCEPHALOPATHY SYNDROME

Jeong Mi Moon, MD and Byeong Jo Chun, MD

Department of Emergency Medicine, Chonnam National University Medical School, Gwangju, South Korea
Reprint Address: Byeong Jo Chun, MD, Department of Emergency Medicine, Chonnam National University Hospital, 8 Hak-Dong, Dong-Gu, Gwangju 501-757, Republic of Korea

□ **Abstract—Reversible posterior leukoencephalopathy syndrome (RPLS) is a neurological disorder characterized by signs of posterior cerebral edema upon radiographic examination. RPLS has been strongly associated with abrupt and significant elevations in blood pressure or the administration of immunosuppressive drugs. Here, we report a case of RPLS occurring in a 30-year-old-mother with a presumed amniotic fluid embolism subsequent to delivery by cesarean section. On the fifth day after the initial successful resuscitation from the cardiorespiratory collapse, she manifested generalized seizure activity and abnormal radiological findings, which were found to be consistent with RPLS. Through our experience, we have suggested that RPLS might occur more easily at lower blood pressures than has been previously recognized. It is important to consider the lower limit at which blood pressure is controlled, especially if accompanied with fever. © 2010 Elsevier Inc.**

□ **Keywords—reversible posterior leukoencephalopathy syndrome; cerebral edema; seizure; hypertension; amniotic fluid embolism**

CASE REPORT

A 30-year-old woman (gravida 1, para 1) delivered a female baby by elective cesarean section at 38 weeks gestation under epidural anesthesia at a local hospital. Eight minutes after the delivery she abruptly became cyanotic and developed bradycardia, followed immediately by the absence of a palpable pulse. She was intubated and cardiopulmonary resuscitation was immediately initiated. One milligram of epinephrine was infused and spontaneous circulation was successfully restored approximately 3 min after the detection of cardiac arrest. She was then transferred to our hospital in a comatose state.

Upon arrival, approximately 60 min after delivery, her vital signs were as follows: blood pressure 90/40 mm Hg, temperature 37°C, pulse 156 beats/min, and a respiratory rate of 32 breaths/min. Physical examination revealed bleeding from the surgical site as well as from the vagina, and we also noted some oozing from the peripheral intravenous cannula site. On neurological examination, she was comatose with no focal neurological signs and showed decorticate posture to pain. Pupils were equal and reactive to light. Electrocardiogram displayed sinus tachycardia without ischemic change. The analysis of cardiac enzymes was abnormal: creatine kinase 126 U/L, creatine kinase-MB 61 U/L, troponin I 0.52 ug/mL (0–0.5), troponin T 0.154 ug/mL (0–0.1), lactate dehydrogenase 709 U/L, and myoglobin 170 ug/mL. The analysis of complete blood cells was white blood cell count 18,100/mm^3, hemoglobin 9.7 g/dL, and platelet 152,000/ mm^3. Other parameters, including those associated with urine test, and liver and renal function tests, except hypoalbuminemia, were all normal. The initial activated partial thromboplastin time (aPTT) (42.5 s) and prothrombin time (PT) (1.25 international normalized ratio [INR]) were slightly prolonged and the level of fibrinogen was decreased to 119.7 mg/dL (180–350). The levels of fibrin degradation products and D-dimer

RECEIVED: 27 May 2005;
ACCEPTED: 17 February 2007

were increased to 77.4 ug/ml (0–5) and 16 mg/L (0–0.3), respectively. On repeat laboratory testing at 1 h after presentation, the white blood cell count changed to 17,200/mm^3, hemoglobin 7.1 g/dL, and platelet 86,000/mm^3. The aPTT and PT results were prolonged to 120 s and 3.51 INR, respectively. These changes were consistent with disseminated intravascular coagulopathy. There were no abnormal findings on her chest X-ray study and brain computed tomography (CT) scan. Mechanical ventilation was instituted.

One hour after presentation, the patient received intravenous injection with inotropic agents because she maintained a low blood pressure despite the administration of crystalloid, colloid, and blood products. After 14 h, with a central venous pressure of 20 mm Hg, the blood pressure was stabilized and the administration of an inotropic agent (dopamine 18 μg/kg/min) was gradually tapered off. Sixty units of blood product, including packed red cells, platelets, fresh frozen plasma, and cryoprecipitate, were administered to the patient within 24 h, after which her coagulation profiles were stabilized at aPTT 34.2 s, PT 1.13 INR, fibrinogen 228 ug/mL, D-dimer 1.03 mg/L, and platelet 107,000/ mm^3.

Over the next 2 days she showed neurological improvements and exhibited localized responses to pain. Her base blood pressure was 125/80 mm Hg. However, the radiographs indicated pulmonary edema throughout both lungs. She was then put on a regimen of furosemide, nitroglycerin, and a 10-μg/kg/min dosage of dobutamine.

Over the next 3 days, we observed an intermittent elevation of body temperature up to a maximum temperature of 38.6°C. We suspected a diagnosis of puerperal endometritis based on the history of operative delivery, the developed fever at third postpartum day, the vaginal packing with cotton during postpartum first 2 days, and the continuous bloody vaginal discharge. The patient was empirically administered aminoglycoside (Amikacin) and third-generation cephalosporin (Cefotaxime). For the next 4 days, she complied with a few simple recommendations. Her arterial blood pressure fluctuated from 183/78 mm Hg to 139/76 mm Hg, coupled with the intermittent fever over 38.0°C. She suddenly began to exhibit generalized seizure activity, which was immediately controlled by the administration of intravenous lorazepam. Her blood pressure was kept under control by hydralazine, an antipyretic agent, and the discontinuation of dobutamine. All laboratory results, including serum chemistry and urine analysis, were within normal limits. The seizure did not recur, and she seemed to recover neurologically to a preictal state. The subsequent clinical course was favorable. On the 10th day after presentation, the pulmonary edema was improved and she was weaned from mechanical ventilation. We then conducted a careful neurological examination. She had a Glasgow Coma Scale score of 15, and a score of 14 on the mini-mental state examination: the function for recall (0/3), calculation (0/5), and orientation (3/10) were all found to be abnormal. She had amnesia with regard to the delivery, and had no memory of her infant daughter. Magnetic resonance imaging revealed high signal intensity in the bilateral occipitoparietal lobe white matter on the T2-weighted image, as well as on fluid-attenuated inversion recovery image. This lesion demonstrated no restricted diffusion on the apparent diffusion coefficient maps (Figure 1). The remaining hospital course was uneventful, and she was discharged from the hospital on the 20th day after admission. At the time of discharge, her blood pressure was 110/80 mm Hg.

On the third month after delivery, we conducted a neuropsychological screening test. She scored a 29 on the mini-mental state examination. Calculation and orientation functions seemed to have recovered fully; however, recall deficits (2/3) were continually seen, especially in regard to the patient's visual memory.

DISCUSSION

We describe the case of a patient who survived despite the high mortality of the amniotic fluid embolism (AFE), but experienced reversible posterior leukoencephalopathy syndrome (RPLS).

AFE has an estimated maternal mortality of between 60% and 80% (1,2). Due to the high mortality rate of AFE, early diagnosis, coupled with prompt cardiopulmonary resuscitation, is required to obtain a favorable outcome. However, AFE is still diagnosed clinically, by a process of exclusion. Many previous studies have reported that supportive diagnostic characteristics of AFE may include measurements of the serum levels of tryptase and complements, as well as the detection of squamous cells and mucous strands in blood in the pulmonary vasculature (3). Our patient exhibited a low level complement: the C3 was 64.6 mg/dL (90–180), and C4 was 9.71 mg/dL (10–40). In this case, the diagnosis of AFE was predicated on both the typical pulmonary cardiovascular hematologic clinical features and the low level of complement.

Reversible posterior leukoencephalopathy syndrome is a rapidly evolving neurologic condition, characterized by signs of transient posterior cerebral edema on imaging studies, as well as associated clinical features, including headache, nausea, altered mental function, visual disturbances, seizure and, occasionally, focal neurologic signs (4). The causes of this condition are diverse, but common precipitants include the acute elevation of blood pressure (usually above the limits defining malignant hyperten-