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Case of successful treatment of acute respiratory distress syndrome in the parturient woman with systemic lupus erythematosus and cerebral cyst of the third ventricular complicated by posterior reversible encephalopathy syndrome

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Abstract

Pregnancy and the postpartum period provoke an exacerbation of systemic lupus erythematosus (SLE), which requires aggressive cytostatic therapy that can cause life-threatening conditions for the fetus and complications for the mother, one of which is posterior reversible encephalopathy syndrome (PRES-syndrome). The article describes a clinical case of successful treatment of acute respiratory distress syndrome (ARDS) in a 22-year-old woman in labor with high activity of SLE during pregnancy complicated by the development of PRES syndrome. Also, for the first time in the postpartum period, the patient was diagnosed with occlusive periventricular hydrocephalus, a giant cyst of the third ventricle. An emergency endoscopic cyst ventriculocisternostomy was performed. In the future, the development of severe ARDS against the background of sepsis, which required the setting of extracorporeal membrane oxygenation (ECMO). A multidisciplinary approach and timely therapy saved the life of a young mother.

Keywords: *ARDS, PRES-syndrome, systemic lupus erythematosus.*

Editor,

We report the case of successful treatment of acute respiratory distress syndrome (ARDS) in a 22-year-old woman with high activity of systemic lupus erythematosus (SLE) during pregnancy complicated by the development of posterior reversible encephalopathy syndrome (PRES).

Patient B., 22 years old, primiparous, was hospitalized to the Almazov National Medical Research Centre due to severe respiratory failure against the background of an increase in hydrothorax, and hydropericardium. She was receiving prednisolone 30 mg/day and rituximab 1000 mg for life in combination with Plaquenil at 28 weeks of gestation due to treatment of SLE. Conducted pulse therapy with methylprednisolone 1000 mg per day for 3 days, also conducted antibacterial, anticoagulant, magnesium, and antihypertensive therapy with a positive effect.

Due to the deterioration of the fetal condition, an emergency cesarean section was performed at 28 0/7 weeks, and cytostatic therapy with cyclophosphamide in a dose of 1000 mg was continued.

Generalized convulsive seizure suddenly was developed on the 1st day after delivery. Magnetic resonance imaging (MRI) of the brain revealed changes in the subcortical-cortical structures of the occipital and temporal lobes, verified as PRES; occlusive periventricular hydrocephalus, giant cyst of the third ventricle (see figure 1).

An emergency endoscopic cystoventriculocisternostomy was performed. On the first day after neurosurgical intervention, respiratory failure rapidly progressed, requiring intubation and mechanical ventilation. Chest computed tomography (CT) demonstrated massive zones of infiltration in the lungs, left-sided hydrothorax, and hydropericardium. In dynamics, after 3 days, extensive areas of ground glass have appeared in the upper lobes of both lungs, zones of interstitial edema were formed, and the manifestation of polyserositis was increased. We diagnosed severe ARDS of mixed etiology (sepsis, SLE - lupus pulmonitis, bilateral nosocomial pneumonia) (see figure 2).

The uterus was extirpated due to an increase in the infectious systemic inflammatory reaction (purulent-necrotic metroendometritis with signs of thrombophlebitis) at 12 days after an emergency caesarean section.

Further, severe hypoxemia due to severe ARDS was persisted. Antibiotic therapy, SLE therapy, and endoscopic administration of surfactant (Surfactant-BL endobronchially 150 mg twice) were performed.

Despite the ongoing therapy, we observed normalization of procalcitonin levels, decrease in body temperature to subfebrile, but increasing restrictive parameters of mechanical ventilation and oxygen requirement (P_aO_2/FiO_2- 44.5), also - negative X-ray dynamics, increasing in the volume of infiltration to subtotal (see graph 1).

An implantation of a veno-venous extracorporeal membrane oxygenation (VV-ECMO) system was performed, and we started protective lung ventilation. The surfactant was re-introduced endoscopically. The total duration of VV-ECMO therapy was 20 days. In the control chest CT, there was positive dynamics - decreasing volume of consolidation in both lungs (P_aO_2 /FiO_2 more than 300). The patient's recovery was complicated by development recurrence convulsive syndrome sue to sedation stopped. Anticonvulsant therapy with valproate was started with positive effect. Plasma concentration of valproic acid reached 63.10 µg/ml. Mechanical ventilation was continued for 46 days.

Critical polyneuropathy was diagnosed. After 50 days of treatment in the intensive care unit (ICU), she was transferred to the therapeutic unit. The rehabilitation period was continued for 1 month after ICU in the hospital. The total duration of hospitalization was 119 days, in ICU - 63 days. After 6 months, the state of health is satisfactory. The child develops according to age. Consent was obtained from the patient to publish this report (April-August 2019).

SLE predominantly affects women during their reproductive years, occurring in about 1/1000 women aged between 15 and 45 years [1]. Pregnancy and the postpartum period provoke an exacerbation of SLE [3], which requires aggressive cytostatic therapy that can cause life-threatening conditions for the fetus and complications for the mother [4], one of which is PRES [5], phased diagnosis, and timely treatment.

Pregnancy can aggravate SLE, induce an active stage of systemic lupus erythematosus, increase the incidence of complications and adverse outcomes, and pose a great threat to maternal and fetal health [1], as demonstrated by our clinical case. Although rare among SLE patients, PRES tends to be associated with high mortality rates and mainly concerns female gender [2]. The diagnosis is mainly radiological, CT may visualize hypodense lesions, but MRI remains the gold standard for diagnosis.

The pathophysiology of PRES in SLED is not yet completely explained [3]. However, there is theory of the reflex hypopefusion of the brain secondary to endothelial dysfunction and exacerbated by vasoconstriction, spasm and/or autoimmune activation, therefore resulting in vascular hyperpermeability and vasogenic edema [4].

Extracorporeal life support, in particular VV- ECMO, was used by us as a backup therapy in a patient with severe ARDS. The Eolia study (2018) failed to rigorously demonstrate an improvement in 60-day mortality in severe ARDS [5]. However, the results of our use of VV-ECMO showed improvement in life-threatening hypoxemia, respiratory acidosis.

The mechanism that triggers the development of respiratory distress syndrome is the embolization of small vessels of the lungs with blood microclots, particles of damaged tissues against the background of toxic biologically active substances formed in the tissues - prostaglandins, kinins and others [6]. Violation of gas exchange is the result of an increase in the amount of interstitial water in the lung tissue [7]. Hypoxemia reflects the volume of intrapulmonary shunting, which is observed against the background of filling the alveoli with fluid, is accompanied by a significant decrease in lung elasticity, and the inspiratory reserve volume decreases. Filling the alveoli with fluid leads to the destruction of the surfactant, and, as a result, to multiple atelectasis. Decreased lung compliance is the result of interstitial and alveolar edema, fibrosis, and surfactant deficiency [8]. It is possible that the use of surfactant in this patient also contributed to the successful treatment of severe ARDS.

In our opinion, the success of patient recovery is due to the continuity of the stages of treatment in a multidisciplinary hospital, the coherence of the work of a multidisciplinary team, and the use of modern high-tech methods of diagnosis and treatment.

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