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LETTER TO THE EDITOR

Fatal Varicella Infections in a Young Patient with Systemic Lupus Erythematosus

Varicella (chicken pox) is usually a self-limited disease, but sometimes it can cause severe complications and death. Here, we report a case of a 19-year-old male patient with systemic lupus erythematosus (SLE), who presented initially with abdominal pain, skin rash with disseminated vesicles formation, interstitial pneumonitis, fulminant hepatitis, and disseminated intravascular coagulopathy (DIC). His clinical condition advanced to multiple organ failure and death, despite acyclovir therapy.

A 19-year-old male patient had a history of SLE with relatively low C3, C4, normal anti-dsDNA, and he was well controlled medically. The patient had stopped steroid therapy 6 months previously, when he was in a stable condition. He was brought to the emergency department due to diffuse abdominal cramping pain. The results of a physical examination showed whole abdominal tenderness. The findings of laboratory tests revealed a normal hemogram, but with a slightly increased portion of monocyte, 9.6% and D-dimer, 3.41 mg/L. The results of hepatic and renal function tests were normal. Because the patient had a history of imperforate anus after an operation in childhood, he immediately received abdominal computed tomography with an angiogram, which showed gallstones, mild intrahepatic ducts dilatation, and nonspecific wall thickening in the rectum and sigmoid colon. Because of persistent acute abdominal pain, he received an explorative laparotomy. In the operation, some adhesion between the mesentery was found. His abdominal pain resolved after an adhesion lysis. On the following day, the patient had some vesicle eruption and a skin rash from his face to his trunk, spreading to the extremities. The finding of a Tzanck smear showed multinucleated giant cells, which supported primary varicella zoster viral infection (chicken pox). He received acyclovir at a daily dosage of 10 mg/kg in three divided doses intravenously. The results of laboratory tests showed D-dimer > 35.2 mg/L, fibrin degradation product >5 μg/mL, fibrinogen 67 mg/dL, platelet count 42,000/μL, a prolonged thrombin time, and a prolonged activated partial thromboplastin time. These findings suggested that he was in DIC status, and he had persistent bleeding from a surgical wound, with massive blood loss.

The patient's condition worsened within the first 24 hours. He had an accelerated heart beat up to 180 beats/min and acute respiratory distress syndrome (ARDS), which was refractory to oxygen therapy. Chest X-ray films revealed infiltration of bilateral lower lung fields (Figure 1). Having acute hypoxic respiratory failure, he received endotracheal tube intubation with mechanical ventilator support. He had pulseless electrical activity, and regained spontaneous circulation after cardiopulmonary cerebral resuscitation (CPCR) in the intensive care unit. A chest X-ray revealed progressively bilateral patchy opacities without cardiomegaly, and a transthoracic echocardiogram showed findings of preserved heart function, suggesting that the manifestations of the chest X-ray film were more like ARDS. The patient was diagnosed as having varicella pneumonia complicated with ARDS and DIC. Within 24 hours, he also had fulminant hepatitis (glutamate oxaloacetate transaminase = 6,343 U/L; glutamine-pyruvic acid-transaminase = 2,692 U/L, and total bilirubin level elevated from 1.03 mg/dL to 2.86 mg/dL). Although he was receiving extracorporeal membrane oxygenation (ECMO) for life support, the septic shock progressed, with multiple organ failure. The patient had refractory metabolic acidosis and hyperkalemia, even though he was receiving continuous venous hemofiltration. The patient died 96 hours after varicella skin manifestations and onset of DIC.

Varicella zoster viral infection is generally a benign, self-limited disease in an immunocompetent host. The incidence of varicella is markedly decreasing as the varicella vaccine becomes more widely used. For a normal unimmunized child, chicken pox-associated mortality is <2/100,000 cases. However, the risk is increased by >15-fold for adult patients. Before the varicella vaccine was introduced in 1995, many hospitalizations and deaths occurred among healthy persons younger than 20 years of age.[1] In Taiwan, the varicella immunization rate for 1-year-old children was 94% in the 2003 birth cohort, 95% in 2004, and 97% in 2005, 2006, and 2007 (http://www.cdc.gov.tw/public/data). In Taiwan, the number of cases of varicella significantly declined after nationwide immunization since 2004.[2] Varicella infection can become a life-threatening disease in immunocompromised hosts. Interstitial pneumonitis is the most common complication of primary varicella infection in adults.[3] The use of ECMO/extracorporeal life support (ECMO/ECLS) has been shown to be beneficial for ARDS patients.[4] However, in our patient, DIC made it difficult to maintain an effective clotting time in the therapeutic range, and DIC did not improve after receiving acyclovir therapy. Varicella infections have been found to have a predictive significance of acute DIC in SLE patients.[5] In the literature, only one case about fulminant varicella infection complicated with ARDS and DIC has been reported in an immunocompetent young adult, who was successfully treated with steroid pulse therapy, hemofiltration, and a mechanical respiratory support with a positive end-expiratory pressure.[6,7] Steroid pulse therapy may be a valuable management for our

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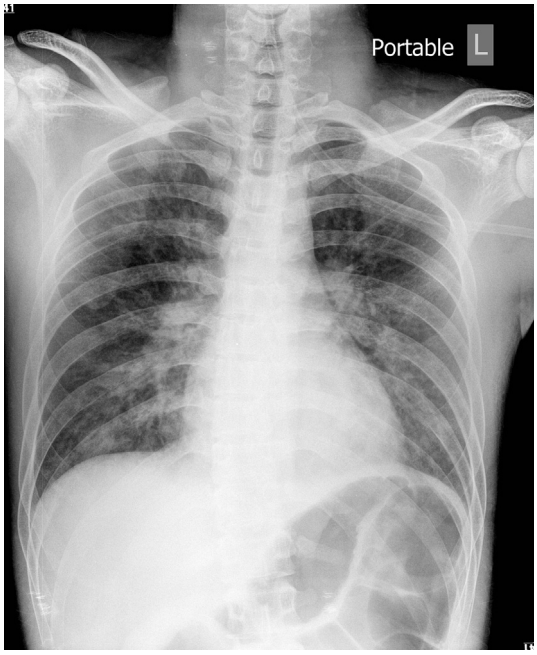

Figure 1 The chest X-ray revealed interstitial infiltration of the bilateral lower lobe of the lungs.

patient, although secondary bacterial infection would be inevitable. To correct our patient's DIC and major surgical wound bleeding with hypovolemic shock, he would need to receive massive blood transfusion including platelets, fresh frozen plasma, and packed red blood cells. Transfusion-related acute lung injury cannot be completely excluded. It is difficult to decide if other blood products, such as intravenous immunoglobulin (IVIG) or varicella zoster immune globulins (VZIG), should be given.[8] Early diagnosis and treatment with acyclovir, and combination therapy with IVIG or VZIG for varicella infections may be an alternative management in such cases.

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