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LETTER TO THE EDITOR

A case of toxic epidermal necrolysis with refractory acute respiratory distress syndrome

Dear Editor,

Toxic epidermal necrolysis (TEN) is a life-threatening cutaneous reaction with several complications. Although acute respiratory distress syndrome (ARDS) is rare, it can cause rapid and potentially fatal pulmonary dysfunction. Here, we present a case of TEN with ARDS attributed to acetaminophen.

A 49-year-old woman with cerebral palsy was admitted to our hospital with a high fever and erythema of the face and trunk that developed 1 day earlier. She took acetaminophen twice at 4 and 1 day before admission. The erosion covered approximately 30% of the body surface area, and hemorrhagic erosions were observed on the lips (Figure 1A). TEN was diagnosed, with a severity-of-illness score (SCORTEN) of 3. Chest computed tomography (CT) revealed a bilateral infiltrative shadow in the lower lung fields (Figure 1B). Aspiration pneumonia was diagnosed. All medications were discontinued, and methylprednisolone pulse therapy (steroid pulse; 1000 mg/d for 3 days) was administered with antibiotics.

On Day 2, she developed acute-onset dyspnea (SpO2: 70%-80%). Chest CT revealed spreading of the bilateral diffuse infiltrative shadow (Figure 1C). ARDS induced by TEN was diagnosed, and mechanical ventilation was initiated. Intravenous immunoglobulin therapy (IVIg; 400 mg/kg/d for 5 days) was administered after steroid pulse therapy, leading to rapid improvements in the skin and mucosal involvement. The prednisolone dosage was tapered to 30 mg/d, and re-epithelialization was completed on Day 19. Nevertheless, the ARDS responded poorly, and steroid pulse therapy was repeated on Days 19-21. Thereafter, the chest shadow and dyspnea gradually improved, and prednisolone was slowly tapered to 5 mg/d on Day 100. However, she developed an ARDS relapse a few days later and received steroid pulse therapy. The chest CT findings improved, although she could not be weaned from mechanical ventilation upon discharge at Day 150. A drug-induced lymphocyte stimulation test was positive for acetaminophen (stimulation index: 803%), and a patch test was negative.

Reports describe an approximate respiratory complication rate of 40% during the acute phase of SJS/TEN.[1] Prost et al reported that approximately 25% of SJS/TEN patients required mechanical ventilation (MV); of these cases, 57% had a fatal outcome.[2] Lebargy et al reported that dyspnea, hypoxia, increased bronchial secretions, and normal chest X-rays within the first 3 days of illness indicated early bronchial involvement.[3] To date, however, few case reports have described the features of SJS/TEN with ARDS. Three of 138 patients with SJS/TEN who were treated at our hospitals from 2000 to 2019 developed ARDS, of whom two (including the present case)

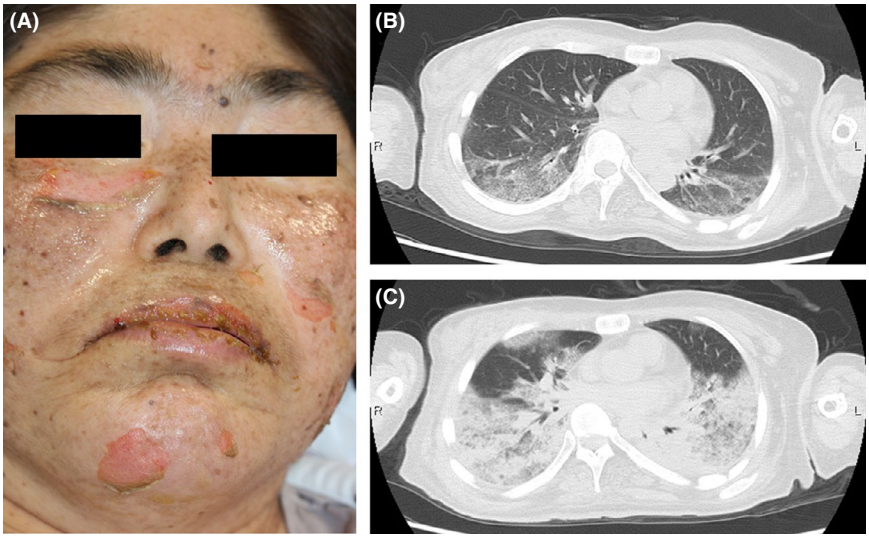

FIGURE 1 Clinical findings. A, Erythema and erosion on the face. B, A chest computed tomography (CT) on admission revealing a bilateral infiltrative shadow in the lower lung fields. C, A chest CT scan on hospital Day 2 revealing spread of the bilateral diffuse infiltrative shadow throughout the lung field

survived. The third and fatal case did not begin treatment until more than 1 week postonset.

Evidence regarding the effective treatment of pulmonary complications with SJS/TEN is lacking. Although antibiotics and steroids are commonly used,[1] the effectiveness of systemic steroid therapy for ARDS remains controversial; some studies reported effectiveness,[4,5] while others reported no survival benefit.[6,7] Moreover, steroid-induced infections tend to worsen prognosis. We conclude that steroid initiation promptly after onset, followed by IVIg, may have contributed to our patient's survival. Additional case studies are needed to support future treatment development.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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