Status dystonicus and rhabdomyolysis in a patient with subacute sclerosing panencephalitis

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To the Editor,

Subacute sclerosing panencephalitis (SSPE) is an uncommon presentation of measles virus infection in the central nervous system[1]. The condition appears to result from mutations in the matrix protein, fusion protein, or hemagglutinin of the measles virus, which allow the virus to spread throughout the brain[2]. Dystonia is a neurological movement disorder, in which sustained muscle contractions cause twisting and repetitive movements or abnormal postures. Patients with dystonia sometimes develop increasingly frequent and relentless episodes of devastating generalized dystonia, which is called status dystonicus or dystonic storm[3]. Dystonia has been described in a few cases with SSPE, but status dystonicus has not been reported before[4,5]. On the other hand, there are a few cases of rhabdomyolysis associated with dystonia[6,7]. Here, we describe a patient with SSPE who had rhabdomyolysis due to status dystonicus.

A 14-year-old boy with SSPE presented with rapidly progressive dystonia, hyperpyrexia and myoglobinuria during the course of a lower respiratory tract infection. SSPE was diagnosed two years ago when he presented with frequent drop attacks and behavioral changes. The course of the disease was catastrophic and the patient rapidly progressed to stage 3 in one year. He frequently had paroxysmal dystonic attacks. He was treated with isoprinosine (1500 mg/day), carbamazepine (600 mg/day) and baclofen (15 mg/day). Five days before presentation, fever with productive cough started and he was diagnosed as pneumonia. Ceftriaxone and salbutamol were started. During this period, dystonic spasms increased and became extremely intense and generalized. At presentation, his temperature was 41°C and he voided "Coke-like" urine. The urine sample was hemopositive without red blood cells. Serum creatine phosphokinase was 30,0000 IU/L (normal: 40-170 IU/L). Renal function tests were in normal limits. After cooling and intravenous hydration, midazolam infusion was started at a dosage of 0.1 mg/kg/hour. Baclofen dosage was increased to 30 mg/day and clonazepam was added (1 mg/day). Bronchodilator and antibiotic treatment with ceftriaxone were continued. The intensity of dystonia decreased and serum creatine phosphokinase values returned to normal values in two weeks. Renal failure did not develop during the follow-up.

Rhabdomyolysis is a clinical syndrome that is an important cause of acute renal failure. It results from injury to muscle tissue, which may be caused by physical, chemical or biological factors. Rhabdomyolysis may result from a wide variety of disorders including trauma, infections, intoxications, malignant neuroleptic syndrome, and hereditary metabolic and connective tissue disorders[8]. Status dystonicus is a rare cause of rhabdomyolysis, and a few cases with hereditary dystonia who developed rhabdomyolysis during status dystonicus have been described[6,7]. The most common causes of acute dystonia in children are encephalitis, postinfectious central nervous system disorders, stroke, intoxications, adverse effects of antiemetic and antipsychotic drugs, inherited metabolic disorders, and acute exacerbations of chronic neurologic disorders[9]. Patients with dystonia sometimes develop severe episodes of generalized dystonia and rigidity (status dystonicus-dystonic storm). The patients have intense muscle activity during status dystonicus and are liable to develop metabolic complications like rhabdomyolysis. Dystonic movements in SSPE have been described rarely[4,5]. One of the patients with dystonia and SSPE showed dystonic posturing coinciding with the periods of delta activity on electroencephalography[5]. The ictal SPECT of the patient revealed marked increased activity in the bilateral caudate, and the authors suggested that dystonic movements in SSPE may represent basal ganglia ictal activity[5]. Status dystonicus in patients with chronic

neurologic disorder is generally triggered by an infection. Status dystonicus in our patient also occurred during a lower respiratory tract infection. In the differential diagnosis, other causes of rhabdomyolysis like malignant neuroleptic syndrome or hereditary causes like carnitine metabolism disorders and glycogen storage disorders were not considered because the attacks were not recurrent and there was no history of exposure to neuroleptic medication or anesthetic agent. Most of the cases with status dystonicus and rhabdomyolysis do not respond to drug therapy like tetrabenazine, trihexyphenidyl, baclofen, clonazepam, and levodopa-carbidopa, and sedation, curarization or surgical procedures like bilateral thalamotomy may be needed[6,7]. Our patient responded well to midazolam infusion and clonazepam; the intensity of the dystonia decreased and rhabdomyolysis resolved in 14 days.

In conclusion, intercurrent infection in SSPE may result in continuous and intense muscle contraction of dystonia. As a result of generalized dystonia, hyperpyrexia and rhabdomyolysis may occur. Serum creatine phosphokinase measurements should be routinely monitored in these patients to prevent renal complications.

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