CASE 1

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Clinical Abstract:

The patient was a 32-year-old male with Kleinfelter's syndrome and a long history of schizoaffective disorder. He had been treated with a number of antipsychotic medications, including phenothiazines and butyrophenones. He developed tardive dyskinesia that was refractory to treatment. In attempts to control complications of the neuroleptic therapy, he had recently been given tetrabenazine, an experimental presynaptic dopamine blocking agent, and was being followed in the Movement Disorder Clinic of Columbia University.

On the day of admission, he was found wandering in the street with bizarre body movements and shortness of breath. He was brought to the emergency room where he was noted to be tremulous, tachypneic, tachycardic, and unable to respond to questioning. The physical examination revealed a tall, thin male in respiratory distress with continuous lip smacking and body movement. The temperature was 104^{O} F, BP 190/98, and RR 36. He was unable to loosen his jaw. The neurologic exam was otherwise unremarkable. The general physical examination, except for small testes, was unremarkable.

The patient met the criteria for neuroleptic malignant syndrome (NMS) and was given Dantrolene and Bromocriptine on an emergency basis. Despite aggressive therapy, including a cooling blanket, it was not possible to lower his temperature. Two days later he had a witnessed respiratory arrest from which he was immediately resuscitated. Throughout his course his blood sugar and oxygenation were within normal limits. He, however, remained unresponsive to verbal commands and painful stimuli. A non-contrast CT scan was negative. The EEG showed diffuse gray matter dysfunction. He developed laboratory evidence compatible with rhabdomyolysis with CPK of 32,060 (0% MB fractions) and elevated LDH (1500), SGOT (1,550), and SGPT (11,020). Hyperpyrexia continued throughout his hospital course.

Despite broad-spectrum antibiotic therapy, his fever spiked to as high as 106^{O}F. He developed flexion contractures and later septicemia by Candida and Proteus. A feeding gastrostomy was placed and he received parenteral hyperalimentation. Four months after his admission, his family requested that he receive no further diagnostic procedures, no antibiotic therapy, and no resuscitation in the event of cardiopulmonary arrest. One week later he died.

Autopsy findings:

The brain was grossly unremarkable. There were multiple microabscesses, some of which contained yeast with pseudohyphae, in the viscera and brain. The cerebral cortex and hippocampus were free of anoxic-ischemic changes.

Material submitted: One H&E slide of cerebellum.

Points for discussion:

What is the nature of the changes in the cerebellum?