

Case #5

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This 11-year-old boy was admitted because of drooling, muteness, hypersomnia, emotional lability, and involuntary movements. Twelve days previously, a young man had attempted to strangle him after luring him into a cellar. The symptoms had come on abruptly six days later, after an intervening period during which he appeared well. On examination, the child was alert and cooperative but appeared inappropriately unconcerned and made no verbal replies. He performed complex commands, copied designs, and wrote well, including answers to arithmetic problems. There were bilateral subconjunctival hemorrhages. Ocular movements were full without nystagmus. He drooled constantly and movements of the tongue were slow. The reflexes were brisk, and the plantar responses were flexor. Sensation was intact. Coordination was impaired because of ataxia, dysmetria, and myoclonic jerks. He made few postural adjustments when lying down and required assistance to walk. Volitional acts were interrupted by choreic movements. An EEG showed slowing in the posterior leads. The cerebrospinal fluid contained 2 white blood cells, 44 mg.% protein. During hospitalization, the child had frequently recurring torsion spasms of the head, neck, trunk and extremities associated with facial grimacing, hyperpyrexia, flushing and diaphoresis while awake. These episodes could not be controlled with medications. Gradually worsening, he died twelve weeks after admission.

Post-mortem examination: Apart from the changes in the central nervous system, the only significant abnormality was mild bronchopneumonia.

Neuropathological findings: The lesions exemplified by those in the submitted slide were found symmetrically in the caudate nucleus, putamen and the globus pallidus bilaterally. There was bilateral focal neuronal loss in Sommer's sector of the hippocampus. The cortex, cerebellum and brainstem were unremarkable.

Submitted section stained with hematoxylin and eosin.

Points for Discussion:

1. Pathogenesis of the lesions.
2. Significance of the regional localization of the lesions; differential vulnerability?