

46th ANNUAL DIAGNOSTIC SLIDE SESSION 2005

CASE 2005-2

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Clinical History:

The patient was a 25 year old, highly functioning college student who developed progressive cognitive and motor decline beginning in 2003. After a successful initial college career, he dropped out of school and, while traveling in Europe, was hit by a car. Although the extent of injuries was described as minor, his grandmother reported that “he wasn’t the same after the accident”. He began to manifest a progressive language impairment, gait disturbance, and social isolation. His living situation deteriorated such that he became homeless, and shortly thereafter he was admitted to the Neuropsychiatric Service at Maine Medical Center (MMC) in October 2004. An MRI of the brain with and without gadolinium demonstrated low signal involving the caudate nucleus, globus pallidus, and putamen bilaterally. High T2 signal also was noted to “surround” the basal ganglia, and to involve the thalami. A subsequent CT scan of the brain was interpreted as “consistent with iron deposition within the basal ganglia”. The patient was discharged on medical treatment that included sertraline, carbidopa/levodopa, mirtazapine, and olanzapine. Because of a deteriorating neurologic condition and additional laboratory data, the patient was readmitted to MMC two months later and appropriate therapy was begun. Physical examination demonstrated no spontaneous language and choreiform movements of the extremities. Shortly after admission, the patient decompensated, developed an aspiration pneumonia, and required assisted ventilation. He died after his family made a decision to withdraw care.

Necropsy Findings:

The general autopsy showed hepatosplenomegaly with macronodular cirrhosis, focal acute pneumonia, and ascities. Gross and microscopic features of the brain will be presented at the slide session

Material submitted: One H&E section of the lesion.

Points for discussion: 1. Diagnosis
2. Prognosis