



Pathology Biology Section – 2007

G5 Acute Hemorrhagic Leukoencephalopathy (Weston-Hurst Disease): A Natural Disease Presents as Head Trauma

Kenneth D. Hutchins, MD, Miami-Dade County Medical Examiner Department, Number 1 Bob Hope Road, Miami, FL 33136; and Lyla E. Perez, MD, Southern Regional Medical Examiner Office, 1175 DeHirsch Avenue, Woodbine, NJ 08270*

The goal of this presentation is to describe a case of death due to acute hemorrhagic leukoencephalopathy (AHL), a natural disease masquerading as head trauma.

This presentation will impact the forensic community and/or humanity by demonstrating how AHL, a natural occurring brain disease which may mimic brain trauma and must be considered in the differential diagnosis of cerebral hemorrhage.

Naturally occurring neurologic disease may occasionally mimic traumatic injury.

A 19-year-old man complained of headache after bouncing a soccer ball on his head. He was admitted to a hospital where he became lethargic, then comatose. A computed tomographic (CT) scan revealed dense, bilateral, frontal lobe hematomas. Angiography did not demonstrate vascular abnormalities. A craniotomy was performed to evacuate the hematomas; however the patient died after a four day hospitalization. Further history revealed that the patient may have sustained head trauma during an altercation in the days previous to the hospital admission. Because of the possibility of a trauma related death, jurisdiction was assumed by the medical examiner and an autopsy was performed. Gross neuropathologic examination revealed a swollen, soft brain that exhibited evidence of surgical intervention and transtentorial herniation. The frontal lobes had dense, confluent hemorrhages, located predominantly in the white matter, extending across the genu of the corpus callosum. Additional foci of hemorrhage were in the convolutional white matter of the cingulate gyrus and the internal and external capsules. There were no cortical contusions or other traumatic pathology. Histologic examination demonstrated perivascular hemorrhage surrounding necrotic blood vessels with fibrinous exudates within the Virchow Robin spaces, perivascular inflammation, and demyelination. These findings were diagnostic of AHL. AHL, or Weston-Hurst disease, is a rare inflammatory, demyelinating disease of the brain characterized clinically by an abrupt onset of neurologic symptoms and signs with rapid progression. Its onset is frequently associated with an antecedent viral illness. The disease is usually fatal, although recovery has been described with medical treatment. The clinical differential diagnosis includes herpes simplex virus type 1 encephalitis, meningoencephalitis, encephalomyelitis, sagittal sinus thrombosis, central nervous system vasculitis, and in the elderly, congophilic angiopathy. Traumatic injuries are usually not entertained in the differential diagnosis, although the gross findings are similar to those associated with fat embolism following skeletal fractures. In this case, the unusual CT scan findings and the history of possible antecedent head injuries led to the suspicion that the lesions resulted from head trauma. This case illustrates the importance for forensic pathologists to recognize AHL as a natural disease process that may rarely mimic traumatic brain injury.

Head Trauma, Acute Hemorrhagic Leukoencephalopathy, Demyelinating Disease