50th ANNUAL DIAGNOSTIC SLIDE SESSION 2009

CASE 2009-8

Submitted by: C. Harker Rhodes, M.D., Ph.D., Dartmouth-Hitchcock Medical Center, 1 Medical Center Dr., Lebanon, NH 03756

Clinical History: The patient was a previously healthy five-year-old male with up-to-date immunizations who initially sought care for headache, low-grade fever, sore throat and unilateral neck tenderness. He had no tick exposure and no recent travel. A number of classmates had cold-like illnesses. Over the following 2 days he experienced myalgia and progressive weakness of his arms (right arm greater than left) and a change in the timbre of his voice. Four days after onset he visited his primary care provider who diagnosed a nonspecific viral illness. He had a white blood cell count of 10,900/mm3 (80% neutrophils, 5% bands, and 12% lymphocytes). Soon after his appointment the patient developed bowel and bladder incontinence as well as the inability to walk. Later that evening he was found in bed apneic and unresponsive with intermittent tonic posturing of the upper extremities. He suffered a cardiac arrest en route to the hospital and on admission to the pediatric intensive care unit was severely hypothermic with an arterial blood pH of 6.5 and a pCO2 of 102 mmHg. A CT scan of the brain showed cerebral edema with evidence of tentorial herniation. A chest x-ray showed a hazy opacity of the right lower lobe. Once criteria for brain death were met, life support was withdrawn and a full autopsy performed.

Autopsy findings: Cerebrospinal fluid obtained by cisternal puncture showed slight xanthochromia, a nucleated cell count 2094 per mcL, with 54% lymphocytes, a glucose of 2 mg/dL, and a protein 368 of mg/dL).

The patient's lungs were three times the normal weight and diffusely firm. Microscopic sections show acute mixed pneumonia with associated hemorrhage. The spleen was twice the normal weight and microscopically showed a florid follicular hyperplasia.

Post-mortem gross examination of the brain reveals profound edema and normal appearing leptomeninges. The brain stem was grossly unremarkable though multiple red-brown discolorations averaging 7 mm in greatest diameter were identified in the dentate nucleus bilaterally.

Material submitted: An H&E section of spinal cord.

Points for discussion:

- 1. Diagnosis.
- 2. In vitro testing.
- 3. Clinical implications.