CASE 1993-5

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CLINICAL SUMMARY

Clinical History: This 34-year-old female was admitted to the hospital 3 weeks after developing left eye pain. She was diagnosed as having optic neuritis. The pain did not respond to Naproxen, but did respond to Sudafed and Tylenol. One week later, she developed headache and fever and was admitted to another hospital.

Her past medical history was non-contributory. A lumbar puncture was performed and revealed 40 RBCs; 11,730 WBCs with 86 percent polys; total protein, 131; glucose, 37. She was given Ceftriaxone. CAT & MRI scans of the head showed a lesion in the right medial frontal cortex with thickening of the anterior corpus callosum. Cultures of her cerebrospinal fluid were negative. She was discharged three days later.

Within the next few days the patient became very confused, had brisk reflexes, left ankle clonus and bilateral Babinski's; and was subsequently admitted to another hospital. She was treated with Decadron and Acyclovir. A repeat CAT scan showed increase in the bifrontal lesions and an emergent left frontal white matter biopsy, by drill needle was performed. The biopsy showed sheets of histiocytes, but no polymorphs or viral inclusions. The patient progressively deteriorated and was subsequently intubated. She was given Mannitol, but remained unresponsive. The family requested that the patient be transferred to this hospital for further medical coverage.

The patient arrived at this hospital intubated and unresponsive to stimuli. Blood pressure, 101/55; temperature, 101.9; respiration rate, 18; pulse, 92. Physical examination was remarkable for non-reactive pupils and no response to deep pain. Pertinent laboratory data included a white blood cell count of 12.1 (77 polys, 16 segs, 7 monos). Her EEG was flat line and the patient was deemed brain dead. The family agreed to withdraw supportive measures. The patient as pronounced dead at 7:50 p.m. on the day of admission.

Necropsy Findings: A brain only autopsy was performed. The brain was markedly but symmetrically edematous with evidence of transtentorial and foramen magnum herniation. On coronal sections there were several large greyish plaques in the white matter at several levels which spared the cortex. Secondary brain stem hemorrhage were present.

Material Submitted:
1. H&E section of cerebrum.
2. 1 Kodachrome of inclusions in macrophages from biopsy.

Points for Discussion:
1. What is the diagnosis?
2. What are the elongate structures in the macrophages from the biopsy?