Case #4

Submitted by: J. G. Chi, M.D. and E. C. Dooling, M.D. The Children's Hospital Medical Center Boston, Massachusetts

This 5-1/2 year old boy was admitted because of headache and high fever of one day's duration. One week earlier, he had travelled through an endemic eastern equine encephalitis area in Rhode Island. On the day prior to admission, he was given 2 doses of ampicillin. The child continued to be febrile (40° C.), and complained of headache, stiff neck and dizziness. He vomited several times and had episodes during which his eyes were said to "roll back." On examination, he was confused and disoriented but he could follow simple commands. There were twitches of the eyelids and mouth and occasional jerks of the extremities. Petechiae were present on the neck, the throat was hyperemic and there was nuchal rigidity. Except for altered sensorium and equivocal plantar responses, the neurological findings were unremarkable. A lumbar puncture yielded clear fluid under an elevated pressure (300 mm H20). It contained 290 white blood cells (50% polymorphs and 50% monocytes), no red blood cells, 245 mg% protein and 95 mg% sugar. The peripheral white blood cell count was 19,400 with a shift to the left; platelets, hemoglobin, prothrombin time and electrolytes were normal.

Eight hours after admission, the child was stuporous but still responded to his name. Twelve hours after admission, he had a generalized seizure and subsequently became decerebrate and unresponsive. The fundi showed blurred disc margins, flame hemorrhages, and absence of venous pulsations. He was treated with Mannitol and Glycerol. Within six hours of the seizure, the pupils were dilated and fixed. He required assisted ventilation two hours later, and when carotid arteriography was performed, there was no significant passage of dye into the brain. Two electroencephalograms within the next 24 hours were isoelectric, and he was declared dead approximately 43 hours after admission and 3-1/2 days after first becoming ill.

Past history was unremarkable except for DPT vaccination two months earlier, two oral polio vaccinations 2 months and one week earlier, rubella and measles vaccinations six weeks earlier, and mumps vaccination one week earlier, in readiness for school.

All bacterial cultures, including blood and cerebrospinal fluid, were negative. Viral titers were not significantly elevated.

Visceral Pathology: General postmortem examination revealed no significant lesions in any organs, including the spleen, kidneys and lymph nodes.

CNS Pathology: The lesions exemplified by those in the submitted slide were scattered throughout the neuraxis without topographic specificity. The slide submitted is stained with Luxol fast blue - Hematoxylin and Eosin.

Post for Discussion:

What is the disease ?