

Case #3

Submitted by: Margaret Grunnet, M.D.
University of Utah, Salt Lake City, Utah, 84132

The patient was a 49 year old game warden who was in good health until one year prior to admission, when he developed the gradual onset of vertigo, falling to the right, diplopia and bizarre behavior. On admission to the V.A. Hospital, he was oriented to name only. He was able to repeat 3-4 digits forward and none backward. He was emotionally labile, distractable and poor at math. However, he was able to carry out verbal commands and use simple objects accurately. Cranial nerves were intact, and motor and sensory examination revealed only somewhat hyperactive DTRs on the right. CSF exam revealed 156 small lymphs, 256 RBCs/mm³ and a protein of 75 mg%.

One month after admission, the patient became increasingly obtunded, and three months later, he was unresponsive and showed decerebrate and decorticate rigidity. This continued three months, following which he became more responsive and after several months, he could talk in sentences and oral feedings were begun. He then again became less and less responsive over a period of three months and was virtually unresponsive for the last eight months before his death.

At autopsy, his brain showed marked atrophy. The major lesions consisted of demyelinated areas in the centrum semiovale, and periventricularly. Similar lesions were seen in the brain stem and cerebellum. Representative microscopic sections are included. On electron microscopic examination, apparently intranuclear filamentous structures somewhat resembling paramyxovirus are seen.

Two slides are distributed: one stained with H & E and one stained with LFB-CV.

Point for Discussion:

1. Is this a case of multiple sclerosis, progressive multifocal leukoencephalopathy, adult SSPE or some new viral demyelinating disease?
2. Are the filamentous structures seen on E/M viral particles, viral-induced nucleoprotein strands or unrelated to virus?