

Case 6

Submitted by: Drs. Clayton A. Wiley, Richard E. Safrin, Peter W. Lampert.
 Department of Pathology (Neuropathology)
 University of California, San Diego
 La Jolla, CA 92093

Reference No.:

Clinical Abstract:

The patient was a 34 year old male who had been in a good state of health until the age of 33 when he developed persistent diarrhea secondary to Giardia and cryptosporidium infection. At that time the patient was noted to have a decreased T4 to T8 lymphocyte ratio. His Giardiasis was irradiated by therapy however the cryptosporidium infection failed to resolve. The patient was discharged on hyper-alimentation. The subsequent year was marked by frequent episodes to nausea and vomiting and abdominal pain. One week prior to admission, the patient noticed the development of painful raised skin lesions. On the day of admission he was found to be alert and responsive except that voluntary motion was restricted to the left side. Initial neurology exam showed focal clonic movements in the right upper extremity with inability to voluntarily move the right side, however, the remainder of the neuro exam was normal. Physical exam was remarkable for increased liver size (2 fingerbreaths below the costal margin) and multiple firm 1 to 2 mm raised skin lesions. These lesions were violaceous and blanched upon pressure but no tenderness was noted. Laboratory findings were unremarkable except for a white blood cell count of 13,200 with a left shift (57 segs, 5 bands, 15 lymphs and 23 monos). The patient was started on Dilantin, however, a precipitous decline in mental status over the next 8 hours resulted in his requiring intubation. Emergency CT scan at that time showed 3 walnut sized lucent lesions in the left hemisphere. A lumbar puncture showed an opening pressure of 248 mm of water. Eighteen ccs of xanthochromic fluid containing a glucose of 89 and a protein of 145 were removed. There were 129 white blood cells per cubic millimeter (83% segs, 1 lymph, 16% monos). Gram stain on this material showed no evidence of organisms. Biopsy of the skin lesion on the right abdomen showed deep dermal and subcutaneous inflammation with early abscess formation and necrosis but no evidence of microbial infection or Kaposi. As the patient's neurologic status continued to decline, a left frontal brain biopsy was performed which demonstrated an acute necrotizing encephalitis but no evidence of organisms. The patient was treated with Septra, gentomycin, amphotericin and clindomycin but showed no improvement. Repeat CT scan demonstrated multiple moderate sized low attenuation lesions in both gray and white matter, and serpiginous enhancement of gyri. The patient died the following day. All culture results from skin and brain biopsies were negative.

Systemic Autopsy:

General postmortem exam demonstrated lymphocytic depletion of lymph nodes, spleen and thymus. Focal pneumonia showed no evidence of an organism. While chronic enteritis was present in the bowel, no cryptosporidium could be identified. Testicles were atrophic.

Neuropathology gross examination:

The fresh brain showed swollen gyri with multiple sites along the surface of the cerebrum and cerebellum showing gray discoloration. These were circular in shape and upon sectioning demonstrated a variable extension of softening into the brain parenchyma (see kodachrome). The overall topography of these lesions consist of a small sphere with one surface in the meninges and the remaining portion projecting to approximately the gray-white junction. Lesions were noted on coronal sectioning to be distributed predominantly in the depths of sulci. There was a single 2 cm. diameter spherical lesion in the centrum semi-ovale of the parieto-occipital region.

Material submitted: One 2x2 kodachrome of occipital lobe; One H&E slide from cortex;
 One unstained slide from cortex

Points for discussion: 1. Identification of pathogenesis of the organism
 2. Antemortem diagnosis of CNS infections in AIDS patients