CASE 9

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Reference No.

Clinical Abstract:

The patient was a 47 year-old bisexual male who presented with a few months' history of fatigue, weight loss and intermittent episodes of diplopia, dysarthria and right facial numbness. General physical examination showed generalized lymphadenopathy, diffuse pulmonary rhonchi and hepatomegaly. Neurological examination was unremarkable. Laboratory evaluation revealed elevated liver enzymes, hypergammaglobulinemia, lymphopenia and inverted helper-suppressor T-cell ratio ($T_4/T_8 = 0.17$). CSF analysis was negative, and lymph node biopsy was non-specific. Extensive bacterial, viral, fungal and AFB cultures of blood, urine, sputum, CSF and lymph node tissue were all negative. Chest x-ray was normal, and CT scan of the head showed mild lateral ventricular enlargement.

Two months later the patient developed gait disorder, increasing somnolence, slurred speech, diplopia and numbness of the right side of the body. Neurological examination showed bilateral internuclear ophthalmoplegia with absent convergence, right-sided sensory loss and ataxia, diffusely increased tone with hyperactive deep tendon reflexes and Babinski's signs. Visual and auditory brain stem evoked potentials were bilaterally abnormal.

Four weeks antemortem the patient was readmitted for evaluation of fever and productive cough. In addition to the above deficits, his neurological examination at this time showed bilateral optic atrophy, vertical nystagmus and right-sided hemiparesis. CSF IgG was elevated with oligoclonal bands, and CT scan of head showed ill-defined non-enhancing pontine lesion without mass effect. Multiple blood, urine, sputum and CSF cultures for bacterial and viral agents were negative, except for one sputum culture that grew Herpes simplex and one urine culture positive for CMV. Bronchoscopy with transbronchial lung biopsy and culture were unremarkable. HTLV-III ELISA antibody was positive in both serum and CSF.

The patient remained febrile, developed diffuse maculopapular rash and became lethargic and disoriented. Over the next week, he progressed into a state of akinetic mutism which proceeded to complete unresponsiveness. He then developed bilateral pneumonia and died of respiratory failure.

Autopsy findings included bilateral bronchopneumonia, diffuse moderate lymphadenopathy without evidence of infectious processes or neoplasms, and hemorrhagic cystitis. The liver had multiple firm, irregularly shaped nodules which on microscopic examination showed coagulation necrosis of hepatocytes with lymphocytes and plasma cells. The brain weighed 1560 gm. Grossly, there were numerous grayish lesions of the subcortical and deep white matter throughout the cerebral hemispheres, midbrain, pons and spinal cord.

Material submitted: One H&E slide of the cerebrum
One 2x2 Kodachrome of the cerebrum

Points for discussion: 1) Diagnosis
2) Pathogenesis