Case 1999 - 6

Submitted by:

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Case Reference number:

Fletcher Allen Health Care (Burlington, Vermont)

Clinical History:

The patient was a 43 year old woman the product of an uneventful pregnancy, labor and delivery, who developed normally. Family history revealed dementia in her paternal grandfather and great grandfather that became apparent after the age of seventy. Despite being an average to below average student (first grade repeated), the patient graduated from high school and worked in a bakery where she had problems "counting money". She married, gave birth to two normal children, and functioned adequately as a mother and housewife. At age 31, the patient was diagnosed with Hodgkin's disease, Stage IIB. She was treated with chemotherapy and eventually total nodal radiation that resulted in remission for the remainder of her life. At age 32, she was noted to be cachectic weighing only 86 pounds (ideal weight 107-110 pounds) despite adequate food intake. Computerized tomographic (CT) scans of the abdomen performed at age 32 and 34 for Hodgkin's disease follow up showed "a paucity of intraabdominal and retroperitoneal fat and no hepatosplenomegaly". At age 37 she was described as of "borderline intellectual capacity", but still able to manage a household and live independently. Cachexia and poor nutrition remained a chronic problem. Beginning 3.5 years prior to death, the patient developed progressive short term memory loss. She expressed no interest in ADLs, was frequently tearful, showed disordered sleep patterns, and had trouble with simple calculations and instructions. Cranial CT scan showed mild cortical atrophy. The patient was evaluated in a memory clinic two years before death. History indicated rapid forgetting, severe language impairment, the need for assistance with dressing and hygiene, and cuing for swallowing to avoid choking. Premorbid verbal IQ was estimated at 82 (70, 14 months earlier); mini mental state examination was 16/30 (22/30, 14 months earlier). Deficits on neuropsychologic screening were profound and widespread. Neurologic examination showed athetoid movements of both hands, hesitant speech, and gait imbalance (NOS). The patient died two years later with suspected pneumonia.

Necropsy findings:

The general autopsy showed bilateral focal obstructive pneumonitis; pulmonary edema; extensive mediastinal, supraclavicular and pleural fibrosis; pericardial effusion; and, marked cachexia.There was no residual Hodgkin's disease. Examination of the brain showed moderately severe, bilateral frontal atrophy.

Material submitted:

H & E section of hippocampus; unstained section of hippocampus

Points for discussion:

1. Diagnosis
2. Pathogenesis