Case 1999 - 6

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Case Reference number: Fletcher Allen Health Care (Burlington, Vermont) A97-74

Clinical History: The patient was a 43 year old woman of French-Canadian ancestry, 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
IIIB. 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. Computed
surgographic (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 in July 1995 (two years before death).
History indicated rapid forgetting, severe language impairment, the need
for assistance with dressing and hygiene, and cues for swallowing to
avoid choking. Premorbid verbal IQ was estimated at 82 (70, January
1994); mini mental state examination was 16/30 (22/30, September
1994). 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