42nd ANNUAL DIAGNOSTIC SLIDE SESSION 2001

CASE 2001 - 06

Submitted By: Philip J. Boyer, M.D., Ph.D., Justin Cates, M.D., Ph.D., Julia A. Castle, M.D., Peter A. Merkel, M.D., E. Tessa Hedley-Whyte, M.D. Penn State University and Massachusetts General Hospital

Case Reference Number: MGH S97-34585

Clinical History:
The patient was a 78 year old woman with a two month history of ataxia and mild memory loss with transient episodes of a "dead" feeling in her right leg associated with intermittent weakness. She was admitted following an episode of speech arrest with right leg weakness. The patient's past medical history included breast carcinoma, for which she underwent right mastectomy 40 years previously; total abdominal hysterectomy and bilateral salpingo-oophorectomy, for an unknown reason; cholecystectomy; rheumatoid arthritis, osteoporosis with a history of an L1 compression fracture; chronic obstructive pulmonary disease with a long history of smoking; and hypertension. Additional past medical history is withheld for sake of discussion. Medications at the time of admission included Cardizem, Vasotec, and prednisone. Family history was significant for a sister who presented with a very similar set of symptoms; her father died of a stroke.

General physical examination was remarkable for a grade II / VI systolic ejection murmur, crackles at the base of her lungs, 3+ pitting edema, along with other findings, to be discussed. Neurologic evaluation of the patient at the time of admission found that she was oriented to person and place but not time; she could not name presidents; her mentation appeared slowed; she refused other cognitive testing. Cranial nerves were grossly intact. Motor examination, limited by lack of cooperation, was, symmetrically, 5/5 throughout except for 4/5 in the hip extensors and flexors. Babinski reflexes were down-going bilaterally; deep tendon reflexes were 2+ in the biceps and patella and 0 at the ankles. Finger to nose testing was intact. Sensation was grossly intact. Gait was unsteady and shuffling, requiring assistance.

Cranial imaging revealed hydrocephalus, dural and leptomeningeal enhancement, and several superficial cortical lesions. CSF evaluation, performed twice, revealed a slight elevation of protein and a mild pleocytosis; both samples were negative for malignant cells; cultures were negative. Evaluation for neoplasm by imaging revealed a calcified 0.5 cm lesion in the left lung apex and left axillary adenopathy. She had a left axillary lymph node biopsy, which was negative for neoplasm. She then underwent neurosurgical biopsy of one of her enhancing intracranial lesions.

She underwent treatment for the diagnosis rendered on the neuropathologic biopsy, which reduced some but not all symptoms. She ultimately declined further treatment and her condition declined. She died 18 months following her initial admission. A complete autopsy was performed.

Material Submitted:
(1) Kodachrome showing gross appearance of a portion of the biopsy at the time of surgery and a microscopic photo from one area in the specimen
(2) H&E stained glass slide

Points for Discussion:
(1) Differential diagnosis
(2) Pathogenesis