CASE 1996-2

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CLINICAL HISTORY: This 51 year old man was in his usual state of health when he became acutely disoriented while on a boat trip. He was taken to a local hospital and found to be hypothermic and hyponatremic. Cranial magnetic resonance imaging was unremarkable. He was treated for the syndrome of inappropriate anti-diuretic hormone with an increase in his serum sodium but his mental status continued to be poor with disorientation and increasing somnolence. Neurology was consulted and felt that the patient was suffering from narcolepsy. He was treated with Ritalin with improvement in his somnolence but with continued disorientation. He was given the diagnosis of a possible midbrain infarction and discharged. However, his mental status did not improve, and 7 months later he developed worsening dementia with hallucinations requiring an admission to a local psychiatric hospital. Treatment with various psychotropic medications did not improve his status. Six months later, he was admitted to a local hospital for a evaluation of worsening dementia. Work up included lumbar puncture which revealed normal protein, glucose, 0 WBC's and negative VDRL; ESR 74 with negative RF and ANA, normal B12, normal TFT's, normal cortisol levels, urine drug screen negative, HIV negative, and lyme titers negative. An admission chest x-ray revealed a small right pleural effusion with analysis consistent with an exudate, but all cultures were negative. A chest computed tomography revealed small bilateral pleural effusions but no masses or adenopathy. During his hospitalization, a mass of the left jaw was also noted. He was transferred to our Medical Center.

PHYSICAL EXAM: VITAL SIGNS: T-97.5, BP-115/60, P-70, R-16. GEN: Slightly obese, awake, and responsive. NECK: approximately 6 x 4 cm firm mobile, nontender mass in left submandibular region; no carotid bruits. HEART: RRR without M, R, or G. NEURO: Alert, oriented to name and place, fully awake but confabulating with a decreased attention span, and poor cooperation. He could repeat well, but had 0/3 recall at 3 minutes. Serial sevens were poorly performed. A left Horner's syndrome was present but other cranial nerves were intact. Motor strength was 5/5 throughout with increased tone and occasional myoclonic jerks. DTR's were 2 + and symmetric Babinski signs were absent. There were no cerebellar deficits and the sensory exam was normal.

HOSPITAL COURSE: A repeat cranial MRI revealed global atrophy of the cerebellum and cortex. EEG showed diffuse slowing and disorganization into the delta and theta frequency range consistent with a moderate to severe encephalopathic process. Fine needle aspiration of the submandibular mass was performed with cytology consistent with a squamous cell carcinoma. Neck and chest CT showed a 2 x 3 cm well circumscribed mass located behind left sternocleiodomastoid muscle and right lower lobe atelectasis with mid-left pleural thickening but no masses or adenopathy. Oncology was consulted and felt that due to the patient's advanced dementia, chemotherapy or radiation therapy was not indicated. The patient was discharged to a nursing home and expired approximately 2 months later. An autopsy was performed and a diagnosis confirmed.

Material Submitted: H&E stained section of mesial temporal region.