

## 48<sup>th</sup> ANNUAL DIAGNOSTIC SLIDE SESSION 2007.

### Case 2007-1

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The patient was a 56-year-old female who presented initially with a six-month history of progressive visual loss and intermittent frontal headaches, fatigue, forgetfulness, and personality change. Magnetic resonance imaging revealed a 2 cm enhancing suprasellar mass that was biopsied and found to be a papillary craniopharyngioma. A bifrontal craniotomy was then performed that resulted in subtotal resection of the tumor. Postoperatively, her course was complicated by symptoms of panhypopituitarism including diabetes insipidus, hypothyroidism, and hypogonadism. 2 additional resections were performed for tumor recurrences three and seven years later as well as at least 2 gamma knife surgeries. Prior to her final resection in 12/05, an MRI showed recurrent tumor. 3 months after this resection, she was brought to the neurosurgery clinic by her family who stated she was experiencing difficulty walking and mild confusion. These complaints were in addition to chronic complaints of blurred vision, headaches, and occasional tinnitus. At that time it was noted that she was unable to recall 3 objects on exam after 5 minutes, and walked with a wide-based gait. Three months later she presented to the emergency department with new onset atrial fibrillation, peripheral edema and confusion, as well as episodes that resembled absence seizures. At that time her husband also stated that she had been “dizzy” and that she had been hypersensitive to vibratory sensations.

Over the course of her disease, 11 CSF samples were obtained, all of which showed xanthochromasia. CSF proteins ranged from 115 to 706, WBC's from 6 to 394, and RBC's from 2 to 5200. Despite extensive evaluations, her symptoms following her final resection defied explanation. Imaging revealed no definite change in her tumor. EEG's, including a 24-hour ambulatory EEG showed either focal slowing or no abnormality. During this time, her mental and functional status continued to deteriorate. MMSE scores ranged from 26/30 to 20/30. During her final neurologic exam four months following her last visit to the emergency department, the patient's caregiver reported that the patient suffered from difficulties in concentrating and poor recent and day-to-day memory. She did not complain of vision, hearing or swallowing difficulties. On examination, she was noted to have a slow latency of response when questioned. Her language was normal. She had a mask-like face, a smooth gait when walking with a walker, and intact reflexes.

Nine days later, the patient presented to the emergency department after a reported seizure. During evaluation, she experienced an additional grand-mal seizure. She was transferred to the intensive care unit where additional seizure activity was noted. Resuscitation measures and anti-seizure medications were administered. She continued to deteriorate in terms of her hemodynamic and neurologic status. The patient then showed signs of brain death and expired two days later. The eyes, kidneys, bladder, liver, adrenals, atria of the heart and aorta were harvested for organ donation.

A general autopsy revealed a recent hemorrhagic infarct of the lingula of the left lung and a focus of hemorrhagic mesenteric fat necrosis. The brain weighed 1400g and was markedly soft and friable, with distortion of the structures occupying the base of the brain and brainstem. Gross examination revealed cerebellar tonsillar herniation, as well as red-brown discoloration of the superficial portions of the inferior temporal lobes, anterior pons, cerebellum, and the entire spinal cord. Residual tumor was not identified.

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Material submitted: 1 H&E stained slide and 1 unstained slide of 3 spinal cord levels

Point for discussion: What is the diagnosis and etiology of her clinical symptoms?