CASE 1999-3

Submitted by: Martha Simmons, MD PHD and Richard L. Davis, MD Neuropathology Unit, HSW 430

UCSF

San Francisco, CA 94143-0506

Clinical History: The patient is a 28 year old right handed woman who was well until she was hit by a bicycle and lost consciousness for several minutes. In the following weeks, she developed new onset headaches associated with nausea and vomiting. The headaches were worst in the early morning, and frequently woke her up at night. Over the next four months, these episodes became as frequent as three times a week, and the patient lost 10 pounds. She felt relatively well in between headaches, until one week prior to admission when she began complaining of difficulty concentrating at work. At this time, her boyfriend also noticed clumsiness in the use of her hands while eating.

The patient had never had significant headaches requiring analgesic use. Her mental status appeared unchanged according to her family and boyfriend. There was no history of seizures or visual changes. She had an unremarkable past medical history, and normal development. Her mother noted that she exhibited frequent head banging as a child. The patient's mother has migraines, and several maternal great aunts died of cancer in old age. The patient is a college graduate, and works as a graphic artist.

Four months later, the patient went to the emergency room because of a particularly severe headache and vomiting. Her vital signs were normal, with BP 102/61 and HR 78. Her general examination was normal. Her head circumference was 55 cm. On neurologic testing, mental status was grossly intact. She had bilateral mild papilledema, but otherwise normal cranial nerve testing. Motor exam revealed a right pronator drift, and was otherwise normal. She had diffuse symmetrical hyperreflexia. Plantar responses were flexor. Sensation was grossly normal. Her gait was normal and there was no dysmetria, but she fell to the right when attempting to perform tandem gait.

On admission, a head MRI showed a 5 x 6 x 8 cm multicystic heterogeneous mass centered in the region of the septum pellucidum, extending into the lateral ventricles with compression of the foramina of Monroe bilaterally, resulting in obstructive hydrocephalus. The mass partially enhanced with gadolinium. There was also bilateral uncal hemiation, and downward brainstem hemiation. The patient was started on decadron, and received intermittent mannitol for severe headaches and vomiting. On the fifth hospital day, a craniotomy was performed.

Gross description: Soft red and tan tissue.

Material submitted: one H&E section

one unstained section

Points for discussion:

Diagnosis?

Does location alter diagnosis?

What special stain(s) used and why?

Prognosis and therapy?