CASE #6

SUBMITTED BY: William G. Ellis, M.D., Department of Pathology, University of California-Davis, Davis, California 95616

The patient is a 16 year old male who was referred for evaluation of headaches and intermittent vomiting of 5 months duration, anemia, fatigue, lack of sexual maturity, and growth arrest which began at age 12 years. Examination: height 150.9 cm. and weight 37.5 kg. (each less than the second percentile), head circumference 55 cm. (50th percentile), right congruous homonymous hemianopsia, bilateral papilledema, slightly decreased visual acuity, spleen and liver enlarged to 5 cm. below the costal margins, immature genitalia with sparse pubic hair, but no neurologic deficits.

Laboratory data: hypochromic microcytic anemia (hematocrit 27%, hemoglobin 8.16) polyclonal hyperglobulinemia, sedimentation rate 140, prolonged partial thromboplastin time, increased numbers of plasma cells and absence of stainable iron in bone marrow, and normal growth hormone. Computerized tomography showed a large intracranial mass extending above and below the left tentorium and adjacent to falx cerebri, falx cerebelli, straight sinus and left transverse sinus. Angiography showed a moderately vascular neoplasm, an enlarged tentorial artery and vein of Labbé.

Left parieto-occipital craniotomy revealed a large, spherical, highly vascular extra-cerebral neoplasm which extended at least 7 cm. anteroposteriorly from occipital bone to near the quadrigeminal plate and was solidly attached or merged with the diffusely thickened tentorium as well as other dural reflections. Approximately 85% of the neoplasm was resected. After surgery, the patient had a transient right homonymous hemianopsia, several generalized seizures, brief inappropriate ADH secretion, and a poor plasma cortisol response to insulin, but no permanent neurologic deficit.

In the six months following surgery, his growth rate has increased, sexual maturation has begun, his neurologic status has remained normal, but his hypochromic microcytic anemia and hyperglobulinemia have improved only to a mild to moderate degree.

MATERIAL SUBMITTED: One (1) H & E stained slide and one (1) unstained slide are submitted.

POINTS FOR DISCUSSION:

1. Diagnosis of this neoplasm.
2. Is it benign or malignant?
3. Is there a relationship between the endocrine dysfunction, hepatosplenomegaly, hyperglobulinemia and coagulopathy and the intracranial neoplasm?