CASE 6

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A female patient was first admitted to hospital at the age of 4 years, because of abdominal enlargement for 4 months. At operation, a tumor of the right kidney measuring 10x8x8 cm. was removed. This was diagnosed as a nephroblastoma. One brother and the mother had cafe au lait spots with tumors, and another sibling had been operated upon for a neurofibroma of the chest. This sibling also had headaches and an abnormal EEG.

The patient in January complained of headaches for one month. Neurologic examination, including a spinal tap, was normal. The EEG, was abnormally slow with atypical sleep frequencies. Numerous cafe au lait spots were noted.

In March the patient showed ataxia and papilledema. A spinal tap revealed clear colorless fluid, pressure 300 mm.; protein 39mg.%; no cells. A skull plate revealed an abnormal sella turcica. The child was thought to have an optic glioma and radiation therapy was begun. Following this, her visual acuity deteriorated and a right frontal craniotomy was performed, revealing a dilated third ventricle and compression of the optic nerves.

She was readmitted at age 7, in a lethargic state. A ventriculogram revealed gross dilatation of both lateral ventricles and the third ventricle. Dye injected into the lateral ventricle was not recovered from the lumbar space. She began to have convulsions. A posterior fossa exploration was performed. The left cerebellar hemisphere was increased in size and very firm, with blunt folia. A portion of this hemisphere was resected. The material submitted is from this biopsy, stained by the hematoxylin-eosin and Masson trichrome techniques. The patient deteriorated and died on the 35th postoperative day.