CASE 6

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A white female patient was first admitted in 1959, 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, age 13, and the mother had cafe au lait spots with tumors, and another brother, age 8, had been operated upon for a neurofibroma of the chest. This sibling also had headaches and an abnormal EEG.

The patient in January 1962 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 1962, 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 on April 23, a right frontal craniotomy was performed, revealing a dilated third ventricle and compression of the optic nerves.

She was readmitted on Aug. 31, 1962 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 on Sept. 16 and on Sept. 19 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.