

CASE 8

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The patient was graduated from M.I.T. at the age of 21 as a metallurgist. He first developed convulsions at the age of 29. These were controlled by phenobarbital. A complete work-up revealed only evidence of a thyroid deficiency which was treated with thyroid extract. At the age of 41 he began to show memory deficits. An EEG at this time showed slow waves in both temporal regions. Control of seizures at age 52 required both Mesantoin and phenobarbital. There were no further seizures until age 60.

The following year he showed a greater memory loss than previously. During this period he was demoted from plant engineer to assistant plant engineer.

The patient was first admitted to our hospital the following January complaining of angina and exertional dyspnea. He was found to have aortic stenosis and anemia (Hb. 9.2 gm.%). His bone marrow was fatty with increased stainable iron. The PBI measured 5.7 mcg. and the leucineaminopeptidase was reduced to 40.7 U. The urinary triglycerides were increased to 209 mg.%. A Hufnagel valve was placed under Inovar anesthesia. The patient expired on the first post operative day.

At autopsy, he was found to have pulmonary hemorrhage and edema with massive hepatic necrosis.

The slides submitted are a paraffin section stained by the Luxol Fast Blue-PAS-Hematoxylin technique and an epon-embedded section of tissue postfixed in osmium and stained by the PAS technique. Neurons like those seen in these slides were found scattered throughout the cerebral cortex. Rare Purkinje cells were involved. There was essentially no involvement of the basal ganglia, brainstem, or spinal cord.