

CASE #7

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This 18 year old, right handed, mentally delayed male with serious behavioral disturbances, was admitted to hospital on 5/23/77. He was the product of a full-term, uncomplicated pregnancy. At age 4, he developed measles with a fever of 105<sup>o</sup>F. One year later, he had his first major motor convulsion. This was accompanied by an abnormal EEG. The patient responded to medical management and there were no further major convulsions until the age of 10 when he developed status epilepticus. The EEG at this time showed "temporal lobe damage and spikes". At age 11, he began to develop aggressive behavior which resulted in his expulsion from several schools. His behavior continued to worsen and his seizures remained essentially uncontrolled. At age 16, his IQ was 45. He had a normal brain scan and CT scan. Neurologic examination was within normal limits. The liver and spleen were not enlarged. A right temporal lobectomy was performed without complications. The frequency of his seizures has diminished, and he is now under reasonable good medical control. Liver function tests and a bone marrow aspiration were both normal. No amino acid abnormalities were demonstrated in the urine and the fibroblast culture was also normal.

The resected specimen weighed 31 grams. Following fixation, the amygdaloid nucleus appeared white and firm. The slides (stained with PAS - Kluver) submitted are from this nucleus. The changes were confined to the amygdaloid nucleus. Electron microscopic studies were performed on material fixed by immersion in ordinary formalin.

POINTS FOR DISCUSSION:

1. Is this a storage disease?
2. Is this a residual effect of measles encephalitis?
3. Is this a congenital malformation or none of the above?