

CASE #5

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Since the age of 6 months, this girl suffered periodic attacks of abdominal pain, anorexia and vomiting and poor muscular tone, followed by lethargy and stupor. At the age of 8 years, she had a severe bout of abdominal pain, dazedness, restlessness and delusions. She was first given 1/4 grain Phenobarbital 3 times daily, and when this had no effect, the dose was increased to 1 grain with 1 tablespoon Phenergan t.i.d. The following day, she became unconscious and was admitted to hospital. After 4-5 days of coma, she developed convulsions. Bilateral extensive decompressive craniotomies were then performed and an arachnoid cyst excised. The patient remained unconscious and was admitted to hospital. After 4-5 days of coma, she developed convulsions. Bilateral extensive decompressive craniotomies were then performed and an arachnoid cyst excised. The patient remained unconscious and was transferred to the Vancouver General Hospital. There was massive bulging at the sites of decompression, flaccid limbs but bilateral Babinski responses. Later she became decerebrate. E.E.G. demonstrated widespread severe cerebral depression. She was treated with Dilantin to control recurrent convulsions, and a low protein diet. She remained unchanged until her death 6 months later.

Necropsy showed fatty degeneration of the liver. The dura was markedly thickened under the craniectomies and contained thin subdural membranes. The brain weighed 950 gms. The cerebral cortex was markedly atrophic and the lateral ventricles dilated.

Section of brain is stained with PAS-Klüver. Black and white photo of brain included.

Points for discussion:

1. Can disease be diagnosed from morphological changes?
2. If not, what types of biochemical studies would be appropriate while patient is still alive and/or on neural tissues at necropsy?