CASE 8

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Clinical Summary: The patient was a 9-month-old girl who developed an acute febrile illness associated with vomiting, bloody diarrhea and renal failure. On the third day of illness she had a seizure which consisted of rigidity and unconsciousness but without clonic or tonic movements. Phenobarbital relieved the rigidity but she remained comatose. Serum sodium was 102 meq/l. A spinal tap done shortly after the seizure showed pink fluid in all 3 tubes but only 9 RBC. Protein was 29 mg% and sugar 28 mg%. Organisms were not seen or cultured. She was admitted to the Children's Hospital in extremis for peritoneal dialysis. Anemia, thrombocytopenia with diffuse petechiae and edema in association with uremia completed the clinical picture and a diagnosis of hemolytic-uremic syndrome was made. Her course was further complicated by pulmonary edema, cardiac arhythmias, bradycardia and tension pneumothorax. She was maintained on the respirator. She became increasingly hypotensive and hypoxic during the last 12 hours of life. The entire illness lasted for 9 days. Past history was essentially non-contributory. She was hospitalized at 4 1/2 months for croup, thought to be due to milk allergy and formula consisted of soybeans. She had had 3 DPT shots and 3 doses of oral polio vaccine.

Visceral Pathology: Gross and histological changes consistent with hemolytic uremic syndrome, including acute renal tubular necrosis and glomerular capillary thrombosis.

CNS Pathology: 1. Acute hemorrhagic encephalopathy, especially involving cerebellum. 2. Mild gliosis of cerebral and cerebellar cortex (especially in marginal layer of cerebral cortex and Purkinje cell layer in cerebellum). 3. Acute striatal degeneration.

Stain submitted: Klüver stain for myelin.

Points for discussion:
1. Classification of striatal lesion
2. Selectivity of lesion for striatum
3. Relationship to hemolytic uremic syndrome
4. Comparison with 1961 Slide Session Case of chronic striatal degeneration in 25-month-old child