CASE 4

Submitted by: Leopold Liss, M.D. and Sezai Gursel, M.D. Ohio State University, Columbus, Ohio.

Ref. No. 3503

A 16-year-old boy had been mentally retarded for at least the last five years. He was well until a few days before admission, when he had sudden headaches while at school, and upon his return home had a stiff neck. In the evening he passed out and possibly struck his head. Subsequently, he was hospitalized. The spinal tap was described as bloody. He had severe frontal headaches and vomitted intermittently. A diagnosis of meningitis was made. His temperature was 102.8 F. There was left facial weakness, a left knee jerk of 3+, a right knee jerk of 2+, questionable Babinski reflexes bilaterally, and a fine nystagmus. Cerebellar testing was normal.

Spinal fluid pressure was 110 mm., and spinal blood sugar was consistently elevated (over 100 mg.%). Spinal fluid protein varied from 600-2,000 mg.%. Red blood cells (800-30,000) were consistently present. White blood cells varied from 55-300, with 80-100% of the cells identified as lymphocytes. Electroencephalogram was normal. X-ray of cervical, dorsal and lumbosacral vertebrae showed diffuse widening of the spinal canal. Complete myelogram showed wide interpedicular distance in the bone without obvious deformity of the spinal cord. The patient was treated with penicillin and discharged with a diagnosis of possible meningoencephalitis.

Eight months later, he was readmitted for evaluation of his mental retardation. Physical findings were essentially the same. Spinal fluid was xanthochromic, with 220 RBC's. CSF protein was 2,400 mg.%. A pneumoencephalogram showed moderate ventricular dilatation and a large pineal recess. There was no evidence of mass lesion.

Subsequently the patient became unresponsive, decerebrate, with dilated pupils and apneia. A ventricular tap revealed pressure over the top of the manometer at 500. A ventricular jugular shunt was performed, but he remained unresponsive and decerebrate, and died 12 days after his second admission. The final clinical diagnosis was that of undiagnosed disease of the central nervous system with hydrocephalus.

At autopsy, the brain was edematous. The meninges were thick and milky, especially over the basal aspect of the cerebellum, and suggestive of diffuse meningeal infiltration by neoplasm. Dissection of the brain disclosed in the frontal portion of the lateral ventricles grey, pinkish, friable tissue lining the ventricular system. Similar material was seen to surround the 4th ventricle. No masses were found in the brain or spinal cord.

Sections are submitted from representative areas of the central neuraxis.

The case is submitted as a diagnostic problem and for discussion of the basic pathological process.