Acute Hemorrhagic Leukoencephalitis
A Clinical and Electron-Microscopic Report of 2 Patients
Treated with Surgical Decompression

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Acute hemorrhagic leukoencephalitis has been regarded as a rare and uniformly fatal neurological catastrophe. Classically, the lesions are confined to cerebral white matter, frequently unilateral but occasionally involving both hemispheres, the brain stem or even the cerebellum. From the time of the original report of Hurst in 1941 until that of Kristiansen et al. in 1956 no cases were diagnosed prior to postmortem examination. In the latter report of 5 surgically explored patients, biopsies of the brain in the 2 who survived were compatible with this diagnosis although histologic sections were not shown. Nevertheless, it was emphasized that all cases of acute hemorrhagic leukoencephalitis need not necessarily terminate fatally, even though the specific diagnosis had until then not been made prior to death.

It is our purpose to report 2 additional cases of acute hemorrhagic leukoencephalitis, treated promptly by internal and external surgical decompression. In the second case the brain was processed for electron as well as light microscopy.

Case Presentations

Case 1. R.C., a 21-year-old white man, was admitted to the Neurology Service of St. Louis City Hospital at 11:00 p.m. on Oct. 6, 1960 with the chief complaints of severe headache of 36 hours' duration and increasing paralysis of the left side beginning 24 hours before admission. A review of his past history disclosed an episode of "flu" in February, 1960, during which he complained of frontal headache, malaise, cough and vomiting lasting for 1 week. Two months before admission he was treated at another hospital for bilateral otitis media. He was well again until 2 weeks before his present illness, when he had a productive cough followed 1 week later by chills and vomiting. One day before admission he complained of mild headache, left-sided pain in the chest, coughing, vomiting and slight left-sided weakness. On the morning of Oct. 6, 1960, he was awakened at 6:45 a.m. by a severe right-sided headache localized mainly to the retro-orbital and temporal areas. He went to work and remained there although the left-sided weakness, headache, nausea and vomiting became increasingly worse. He noticed "tingling" and "numbness" of the left side as well as slurred speech. He returned home at 5:30 p.m. but became weaker and less alert during the evening. Later that evening his physician sent him to the hospital.

Examination. He was a thin, uncooperative, obtunded man with normal vital signs. There was partial paralysis of the right 3rd nerve, left central facial weakness and a left hemiplegia. The optic disc margins were sharp but the veins were engorged. At lumbar puncture the pressure was 170 mm. of water and the fluid was clear with only 1 white blood cell. Cerebrospinal fluid protein was 74 mg. per cent, sugar 62 mg. per cent, and chloride 115 mEq. The count of white blood cells was 19,400, hemoglobin was 14.3 gm. and urinalysis gave normal findings.

Course. Within 2 hours he became even less responsive with a dilated, fixed right pupil. Suspecting an expanding mass in the right hemisphere, a right carotid arteriogram was done which showed marked displacement of the right anterior cerebral artery to the left. The lateral projections were of no help. He was transferred immediately to Barnes Hospital by which time he was barely responsive to loud commands. The eyes were divergent with complete paralysis of the right 3rd nerve and extremely engorged retinal veins. Ventriculography was performed immediately and demonstrated a pronounced shift of the entire right ventricle to the left side (Fig. 1). During this procedure he had a generalized seizure.

Operation. As there was no definite anterior-
posterior limit to the process, a large right frontoparietal temporal bone flap was reflected. The dura mater was extremely tense. The brain was needled in all directions in search of an abscess, hematoma, or cystic fluid, but only mushy white matter could be aspirated. Thirty per cent urea was given intravenously prior to opening the dura mater more widely but little relaxation was obtained. The meninges appeared slightly cloudy. The subcortical white matter in the frontal lobe was entered and found to be soft, edematous, and easily aspirated. A large amount was removed in search of a possible small tumor, but none was found. Though the brain swelling had been reduced it was still necessary to make a large temporal bone decompression in order to replace the bone flap.

Pathologic Report. The surgically excised cerebral tissue was fixed in formalin, embedded in paraffin, and stained with hematoxylin and eosin, phosphotungstic acid-hematoxylin and by Weil’s myelin stain. There was a distinct polymorphonuclear exudate present in the subarachnoid space, and about many of the larger intracerebral blood vessels. The cerebral cortex was normal except for the perivascular cellular infiltrate. In contrast to the cortex there was a distinct increased cellularity in the underlying white matter (Fig. 2). Numerous recent ball- or ring-hemorrhages (Fig. 3) were present, often around a central small blood vessel. Neutrophiles ringed some vessels. There was perivascular demyelination, especially in the regions of hemorrhage. Gitter cells were numerous. Edema was prominent.

Postoperative Course. He remained semicoma- tose for several days and a tracheostomy was necessary. Improvement was gradual and after 10 days he was able to talk rationally but still was severely hemiparetic on the left side. After several weeks of intensive physical rehabilitation he was able to walk and improved enough to go home, although weakness of the left arm and partial paralysis of the right 3rd nerve remained. A right carotid arteriogram performed 3 weeks after operation was considered normal (Fig. 4).

Subsequent Course. After discharge from the hospital he had several generalized seizures but since then has been seizure-free on Dilantin and phenobarbital. The neurological deficits have improved but he continues to have definite weakness of the left arm and hand, and a left homonymous hemianopsia. Function of the 3rd nerve is now normal. Extensive psychometric evaluation has shown remarkably little intellectual impairment. However, he has required psychiatric therapy because of periods of depression and anxiety but these, in large part, are caused by distressing and complicated family problems.

Case 2. C.R., a 12-year-old white girl, was admitted to St. Louis Children’s Hospital on Nov. 20, 1961 with a history of convulsions and coma of 12 hours’ duration. Although there was no definite history of an upper respiratory infection she had not felt entirely well for a week before admission. Two days before admission she remained in bed, slept most of the day, and the next day seemed subdued. At 3:00 a.m. on the day of admission she awakened her mother by crying out and complained of headache and abdominal pain. At 7:00 a.m. she was found on the floor convulsing. Thereafter she had numerous seizures, beginning on the left side and then becoming generalized. Between seizures she was semiresponsive and paralyzed on the left. At noon, she was treated with phenobarbital and then transferred by ambulance to this hospital, during which time the convulsions gradu-