Case Reports and Technical Notes

Reticulum Cell Sarcoma of the Septum Pellucidum

Case Report

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Tumors of the septum pellucidum are rare, a total of 73 cases having been reported in the literature through 1966.1,2,5,6,8,9,12,14,15,18 All but two of these have been gliomas, one a fibrosarcoma9 and one that resembled a sarcoma.19 No sarcomas of the reticuloendothelial cell system have been reported arising at this site.

The purpose of this paper is to report a case of reticulum cell sarcoma of the septum pellucidum and to emphasize the importance of air studies (pneumoencephalography or ventriculography) and electroencephalography in cases of possible intracranial tumor. The value of cerebrospinal fluid cytology in cases of suspected brain tumor is also discussed.

Case History

This 59-year-old white woman had been in good health until 2 months prior to admission when, following a fall, she experienced headaches in the right retro-orbital area. Three weeks before admission, she had become forgetful, easily confused, and complained of dizziness, and 1 week later developed nausea, vomiting, and ptosis of the right eye. On July 5, 1966, she was admitted to the neurosurgical service with the clinical impression of an unruptured cerebral aneurysm.

In 1932, she had been treated for syphilis with "shots in the arm." In 1958, the serologic test for syphilis had been negative. In December, 1963, she had a reactive cardiolipin complement fixation with no titer and was treated with benzathine penicillin, 1.2 million units weekly for 4 weeks.

Examination. The patient appeared in no acute distress; her temperature was normal. A grade 3/6 midsystolic, ejection murmur was heard at the apex and along the left sternal border. She was extremely confused and disoriented as to time and place. She did know her name and could follow simple commands. The cranial nerves were intact except for a right third nerve paresis. Sensory responses were generally depressed but symmetrical. Coordination was normal. Deep tendon reflexes were hypoactive bilaterally. No Hoffman or Babinski reflex was elicited. The Romberg sign was questionable, and her gait was very unsteady.

Initial laboratory values included a white blood count of 7200/mm³ with a normal differential count, hematocrit 42 volumes %, and hemoglobin 14 gm/100 ml. Cardiolipin microflocculation was reactive without titer, and the cardiolipin complement fixation was nonreactive. Electrolytes, urea nitrogen, fasting blood sugar, and urinalysis tests were normal. An intermediate purified protein derivative test (PPD) was negative, and cocci-diodin and histoplasmin skin tests were both indurated to 8 mm at 48 hours. Chest films were normal. Skull radiographs showed a non-specific thickening of the skull.

Initially, the patient complained of right-sided headache. A right carotid arteriogram was interpreted as normal. Shortly after this, she began to deteriorate, with increasing paralysis of the right third cranial nerve and depression of her mental status. No other neurological signs became evident except for a stiff neck. The cerebrospinal fluid (CSF) contained 428 cells/mm³ all were lymphocytes. Cardiolipin complement fixation was nonreactive at 0.1 and 0.25 cc concentrations, and a colloidal gold curve was 0122210000. A concentrated smear for acid-fast bacilli and an India Ink stain for
Cryptococcus were negative. It was felt that the cells and the basilar signs were suggestive of tuberculous meningitis. Therefore, the patient was transferred to the Infectious Disease Service.

At the time of transfer, the patient was semicomatose, responding only to simple commands such as moving her head, eyes, or extremities. She could recall her name but was not oriented to time or place. Repeat neurological examination was unchanged. She was immediately started on triple antituberculous therapy. A repeat spinal tap had an opening pressure of 130 mm of water and a closing pressure of 60. There were 41 polymorphonuclear leukocytes and 41 lymphocytes. Cerebrospinal fluid sugar was 20 mg%, protein 280 mg%, and a chloride was 114 mEq/liter. Because of a rapidly deteriorating course, chloramphenicol, sulfadiazine, and high doses of penicillin were started. The patient was comatose the following day, and another spinal tap had an opening pressure of 210 mm of water and a closing pressure of 150. The CSF contained 50 cells/mm\(^3\); 36% polymorphonuclear leukocytes, and 64% lymphocytes. The protein was 240 mg% with a markedly increased globulin fraction; sugar was 12 mg% and chloride was 112 mEq/liter.

Eight days after admission, the patient developed Cheyne-Stokes respiration and several hours later died. Subsequently, cultures for acid-fast bacilli, fungi, and pyogenic bacteria of all spinal fluids were negative.

**Autopsy.** The significant pathological findings were limited to the brain. The basal leptomeninges, especially about the oculomotor nerves, were prominently thickened. The floor of the third ventricle was bulging, suggesting a mass lesion or hydrocephalus. The convolutions were moderately flattened, indicating some degree of cerebral swelling. Otherwise the external surface of the brain was normal. On coronal sectioning of the brain a markedly thickened septum pellucidum (Fig. 1 left) was evident. Actually, the septum had been replaced by a soft and slightly friable, ivory-colored tumor that extended into the corpus callosum and into the hypothalamus (Fig. 1 right). The remainder of the sectioned cerebrum was normal. Hemorrhages were present in the tegmentum and upper midbase of the pons.

**Histological Examination.** The tumor was composed of a proliferation of neoplastic reticulum cells (Fig. 2). The tumor extended much farther than was grossly apparent. The tumor cells tended to aggregate about blood vessels and to grow into and fill up the Virchow-Robin spaces (Fig. 3). Some of these spaces were filled with tumor cells several centimeters away from the grossly apparent tumor. Reticulum cells of a