Case Reports and Technical Notes
Reticulum Cell Sarcoma of the Septum Pellucidum

Case Report

DAVID E. CRAIG, CAPTAIN, MC,* TYSON C. COBB, CAPTAIN, MC, and HOWARD W. HUNTINGTON, CAPTAIN, MC
Pathology and Medicine Services, Brooke General Hospital, Fort Sam Houston, Texas

Tumors of the septum pellucidum are rare, a total of 73 cases having been reported in the literature through 1966. All but two of these have been gliomas, one a fibrosarcoma and one that resembled a sarcoma. 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 microfloculation 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-doidin 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.² 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

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**Fig. 1. Left.** Coronal section through basal ganglia, showing the wide septum pellucidum. **Right:** Coronal section at the level of the anterior commissure, showing tumor and thickened basal leptomeninges.
Reticulum Cell Sarcoma of the Septum Pellucidum

Fig. 2. Photomicrographs of tumor, showing proliferation of reticulum cells. *Left:* H. & E., × 225. *Right:* H. & E., × 475.

Fig. 3. Photomicrographs from area several centimeters away from grossly apparent tumor, showing tendency for cells to grow in the Virchow-Robin spaces. *Left:* H. & E., × 225. *Right:* H. & E., × 475.
similar neoplastic type were present in the basal leptomeninges; these apparently accounted for the thickening of the meninges.

**Discussion**

The reported tumors of the septum pellucidum which have been examined microscopically have been gliomas\(^5,6,9,8,9,12,14,15,18\) except for one fibrosarcoma.\(^9\) At least 18 of these gliomas were glioblastomas,\(^3,5,6,8,9,12,14,15\) some of which were multicentric in origin. There were eight oligodendroglomas,\(^3,5,9,12\) five ependymomas,\(^3,5,9\) and three neuroastrocytomas.\(^6,9\) All the rest were astrocytomas Grade I and II, or gliomas with no further designation. There has been no previous report of a reticulum cell sarcoma of the septum pellucidum.

The most common extension of tumors of the septum pellucidum has been into the corpus callosum (10 cases);\(^5,5,6,9,18\) this extension was also present in our case. Other gross involvements have included the frontal lobes in seven cases,\(^5,6\) third ventricle in five cases,\(^6,9\) the basal ganglia in three cases,\(^6,9\) and the thalamus in one case.\(^9\) There was extension into the hypothalamus in our case, but no gross involvement of the frontal lobes, basal ganglia, or thalamus. However, there was perivascular extension of the tumor throughout the entire brain, in a manner typical of reticulum cell sarcomas.\(^2\)

A common finding has been complete obstruction of one or both foramina of Monro with widely dilated lateral ventricles.\(^3,5,6,9\) In our case the tumor partially filled the lateral ventricles, but the angle formed by the caudate nuclei and the corpus callosum was sharp, indicating no hydrocephalus. The gyri were flattened and the sulci compressed, however, suggesting increased intracranial pressure; this was further confirmed by brain stem hemorrhages. The increased intracranial pressure in this case was felt to be due to brain swelling caused by the perivascular spread of the tumor, in contrast to the internal hydrocephalus in the cases previously reported.

The largest series of tumors of the reticuloendothelial system of the brain has been presented by Burstein, et al.\(^2\) In their series the majority of the tumors were solitary and localized in the cerebrum. Three cases had diffuse involvement, and four had multiple separate lesions. Of the 34 cases with a single neoplastic mass, 24 were in the cerebral cortex, eight in the cerebellum, one in the basal ganglia, and one in the internal capsule. Most of the other cases of the literature have been solitary and cerebral. Three of Russell's cases, however, had extracranial lesions.\(^17\)

The gross appearance of primary reticulum cell sarcomas does not differ from that in other organs of the body. The tissue is firm and homogeneous and varies from white to gray-pink. There are no areas of necrosis, hemorrhage, or cysts; these features were also lacking in our case. Evidence of increased intracranial pressure is usually present.

Reticuloendothelial cell sarcomas are very cellular, the cells of which are uniform in size and shape and measure 12 to 15 μ in diameter. The nuclei are oval and vesicular with fine, widely dispersed nuclear chromatin. The nucleoli are generally inconspicuous, although they may be prominent in anaplastic tumors,\(^10\) as noted in our case. Perivascular infiltration by tumor cells at some distance from the tumor mass is a constant finding. These tumor cells are often mixed with lymphocytes. Extension of the tumor cells to the leptomeninges occurred in our case, although tumor cells were not found in the Virchow-Robin spaces in sections of the spinal cord.

It has been emphasized that tumors of the septum pellucidum cause no specific syndrome.\(^9\) It is only when they eventually compress or obstruct the foramina of Monro that symptoms related to increased intracranial pressure appear. As previously stated, there was no evidence of obstruction or internal hydrocephalus in our case, but there was evidence of increased intracranial pressure.

Tumors of the septum pellucidum have been more frequent in young adults, although children aged 4 and 6 and adults aged 72, 62, and 59 have been reported. The most frequent symptoms have been mental deterioration, forgetfulness, change in personality, headaches, and visual and aural disturbances.\(^3,6,9\) Approximately one-third of the patients have had convulsions. Our pa-
tient had no history of convulsions or aural disturbances, but she had all the other symptoms. The most common signs have been papilledema, vomiting, mental aberrations, and ataxia. Cranial nerve palsies have not been uncommon and they usually involved the first six nerves. Our case had developed only a right third nerve paralysis and papilledema.

Arteriography was performed in our patient to exclude an aneurysm of the right internal carotid artery. Complementary air studies (ventriculography or pneumoencephalography) were not performed. Potts has emphasized that both angiography and air studies are often needed to determine the exact position of a mass lesion, its vascularity, its relationship to major vessels and the ventricles, and whether there is obstruction to the cerebrospinal fluid pathway. Although carotid arteriography alone is very useful in establishing the presence of a supratentorial tumor, air studies are essential to diagnose a tumor of the septum pellucidum; electroencephalography has been of no localizing value and was not done in our case.

The mononuclear cell pleocytosis of the spinal fluid in our case was interpreted as indicating a granulomatous leptomenigitis. In retrospect, many of these mononuclear cells may have been malignant reticulum cells. To our knowledge, there is no report of a reticulum cell sarcoma of the brain in the literature in which the diagnosis has been made by CSF cytology. Pleocytosis is common with intracranial neoplasms, with equal frequency in gliomas or metastatic carcinomas. In one study, malignant cells were present in up to 40% of the cases. Thus, it would seem worthwhile to obtain cytologic examination on the CSF if a brain tumor is even remotely suspected.

Once signs of increased intracranial pressure develop, tumors of the septum pellucidum assume a fulminant course. French and Bucy have reported five cases in which surgery was successful, and Chusid and de Gutiérrez-Mahoney have added three more in which surgery or X-ray or both were successful. Burststein, et al., have pointed out the rapidly fatal course of the reticuloendothelial cell sarcomas; these patients live only a few weeks after operation. Radiation therapy did extend their survival periods to an average of nearly 4 years after tissue diagnosis.

Summary

A review of the literature reveals 73 cases of tumors of the septum pellucidum, all but two of which have been gliomas. We have described a case which is apparently the first report of a reticulum cell sarcoma of the septum pellucidum. The diagnostic significance of arteriography, air studies, and examination of the cerebrospinal fluid for tumor cells has been discussed, and the value of the last two techniques emphasized.

References


