Subependymomas of the Septum Pellucidum*

Report of Two Cases

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In 1944, Globus and Kuhlenbeck stressed the importance of the subependymal cell plate in the histogenesis of primary neuroectodermal tumors. Since then, 53 tumors, composed mostly of ectodermal elements derived from the subependymal glia, have been reported.1-4,6,8,11-13 These have been called subependymoma,13 subependymal astrocytoma,4 subependymal glomerate astrocytoma,9,6,12 and subependymal mixed glioma.3 More than 75% of these tumors arose in the fourth ventricle or its recesses; only five arose from the septum pellucidum.4,11,12

Identification of subependymomas arising from the septum pellucidum is a prerequisite for their appropriate surgical management; two such cases form the basis of this report.

Case Reports

Case 1. A 5-month history of personality changes and memory loss prompted admission of this 32-year-old woman. She had become progressively more irresponsible and complained of occasional generalized headaches. Amenorrhea and athetoid movements of the left hand were noted 3 months before admission. One month before admission she developed an ataxic gait.

Examination. On March 19, 1967, examination revealed dementia, a wide-based gait, and minor athetoid movements in the left hand and arm. A right central facial weakness was present, and deep tendon reflexes were symmetrically hyperactive. The Babinski response was plantar bilaterally, and a snout reflex was present. A brain scan (Hg197) demonstrated a large area of increased uptake in the midline anteriorly. Carotid angiography revealed symmetrical ventricular dilatation without other changes. The ventriculogram (Fig. 1 left) showed a large bilateral intraventricular mass centered at the level of the foramen of Monro. The ventricular fluid protein was 10 mg%.

Operation. On March 24, 1967, the intraventricular tumor mass was exposed through a left transcortical incision. The tumor arose from the base of the septum pellucidum on the left side, depressing the floor of the lateral ventricle. It was easily separated from the surrounding brain. Posteriorly the intraventricular portion extended as far as the trigone of the left lateral ventricle. The tumor mass was 6 × 8 × 5 cm, lobulated, white, fibrous, and rubbery hard. A portion of the tumor embedded in the basal ganglia was not removed. A ventriculopleural shunt was performed.

Postoperative course. Recovery was uneventful except for a temporary increase in dementia. Subcutaneous induration about the ventriculo-pleural shunt necessitated readmission 4 months later. A spinal air injection demonstrated no obstruction of the ventricular system. Spinal fluid protein content was 20 mg%. When last seen 1 year after operation, the patient was able to carry out her house work, had only mild dementia, and did not require a spinal fluid shunt.

Case 2. A 51-year-old woman was admitted to the University of Kentucky Medical Center on June 1, 1967, complaining of unsteadiness of gait for 9 months. At first her illness was thought to be psychiatric.

Examination. The patient had a slightly ataxic, hesitant gait, and a positive snout reflex. Mild cogwheel rigidity was present in both wrists, and the deep tendon reflexes were symmetrically hyperactive with transient quadriceps clonus bilaterally. Psychological tests were administered because of a suspicion of an organic dementia. The Wechsler Adult Intelligence Score was 73. The discrepancy between the verbal score of 84 and the performance score of 62 in a pre-
vously well-functioning woman supported the suspicion of an organic brain lesion. The awake electroencephalogram showed a diffusely fast bifrontal dysrhythmia with paroxysmal sharp waves; the sleep record was normal. Radiographs of the skull showed no abnormality. The cerebrospinal fluid examination was entirely normal. A brain scan (Hg197) showed an area of increased uptake in the region of the third ventricle, and carotid angiograms suggested moderate ventricular dilatation. A pneumoencephalogram showed poor filling of the ventricles with generalized dilatation; a questionable mass protruding into the right frontal horn prompted further study. A ventriculogram (Fig. 1 right) showed the tumor protruding into both lateral ventricles. The firm, fibrous, lobulated mass was $6 \times 6 \times 5$ cm and arose from a sessile base on the septum pellucidum which it had displaced into the left frontal horn.

**Operation.** The tumor occupied the frontal horn of the right lateral ventricle, and was totally removed along with the involved septum pellucidum.

**Postoperative course.** The day after the operation the patient became less responsive and demonstrated decorticate posturing; 1 hour later she was unresponsive with decerebrate rigidity. She improved immediately after ventricular puncture and spinal fluid drainage. Rapid improvement ensued after the insertion of a ventriculo-pleural shunt. The patient was discharged ambulatory 2 weeks after operation. Six months later she shows no ataxia and her dementia was improved significantly.

**Discussion**

The subependymoma was first recognized as a histological entity by Scheinker, who presented a series of brain tumors composed predominantly of ectodermal elements derived from the subependymal glia. The characteristic pathological features of these tumors are listed below:

1. The predominant cell is a mature fibrillary astrocyte
2. There is a great preponderance of glial fibers over cellular elements
3. The tumor is lobulated and well demarcated from surrounding tissue thus revealing an expansive type of growth, rather than in infiltrative one
4. The expansion of the tumor is usually directed toward the ventricular space
5. Mitotic figures, giant cells, and areas of necrosis are absent

Both of our tumors were densely fibrillary with a paucity of cellular elements, predominantly astrocytes with a scattering of ependymal cells (Fig. 2).

French and Bucy, reporting on tumors of the septum pellucidum, described five tumors arising from this structure, at least three of which were histologically identical.