TUMORS OF THE SEPTUM PELLUCIDUM

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TUMORS of the septum pellucidum are so rarely recognized clinically
that it is possible to find published descriptions of only 31 cases.
Moreover, most of these reported tumors were encountered at autopsy
and in only 4 instances was successful removal by operation described. For
this reason, the material of a series of 5 such cases was reviewed. Four of
the patients were studied at the Illinois Neuropsychiatric Institute, Uni-
versity of Illinois, and 1 at the University of Chicago Clinics.

A review of these case reports indicates that the preoperative diagnosis
of tumors of the septum pellucidum is often quite difficult but may be
established if the possibility of such a condition is entertained. The impor-
tance of establishing the correct diagnosis is emphasized by the fact that 2
of the patients in this series were successfully operated upon and 1 has
survived 11 years in excellent health.

CLINICAL DATA


History. The patient was a 37-year-old man who had been in excellent health until 7 years
before admission. At that time, he noted the onset of periodic headache, particularly in the
vertex; these episodes occurred almost daily and usually began in the early evening. The
headaches lasted a few hours and were associated with subjective dimness of vision, impaired
hearing and a sensation of pounding in the ears. For about 2 months prior to admission, they
were further associated with numbness and weakness in the right leg. The past history in-
cluded episodes of upper abdominal pain 2 hours after eating relieved by ingestion of food,
and a minor head injury at the age of 7. One of 10 siblings was said to have had epilepsy.

Physical Examination. Pulse 88, temperature 99.2, respiration 22, and blood pressure
126/76. General physical findings were not abnormal. On neurological examination, the pa-
tient was found to be intelligent, cooperative, well oriented and well adjusted. There was
some blurring of the nasal portion of the optic nerve head but no measurable papilledema.
Visual acuity and fields were normal. Reflexes were normal and no other abnormalities were
noted.

Laboratory Findings. Blood and urine examinations and Wassermann and Kahn reactions
were negative. Spinal fluid examination was negative except for a pressure of 195 mm. of
water. The EEG showed temporoparietal asymmetry. Radiographs of the skull showed a
4 cm. irregular calcified area in the midline well above the sella turcica. On ventriculography,
this mass was seen to be situated between the bodies of the dilated lateral ventricles separating
the anterior horns (Fig. 1). A partial ventricular obstruction existed. Arteriograms showed
abnormal displacement of the anterior cerebral arteries but there was no increase in vascular
supply to the calcified mass.

Operation. The tumor was exposed through a right frontal transventricular approach by

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Dr. Eric Oldberg. It was a bluish-red, hard, encapsulated mass extending into the right lateral ventricle, to the floor of which it appeared to be attached. Complete piecemeal resection was accomplished after numerous vessels coursing into it were secured. During the resection, the major portion of the septum pellucidum was removed, thereby making a direct communication between the lateral ventricles.

Postoperative Course. The patient was somnolent, hypertonic, and incontinent although his general condition was quite satisfactory. There was slight elevation of temperature for 4 days, but thereafter pulse, temperature, blood pressure and respirations were normal. Two weeks after operation he was found to have papilledema of 1 D., slight right central facial weakness, and increased tonus of the extremities and neck, but reflexes, sensation, and motor power were normal. He was still somewhat confused and his memory was poor, but he responded slowly and rationally to questioning. At the time of his discharge from the hospital, he occasionally urinated involuntarily but was regaining control and improving in general quite rapidly.

Microscopic Examination of Tissue Removed at Operation. The tumor (Fig. 2a) was composed of small collections of cells separated or surrounded by large areas of acellular, fibrillar material. The cells contained round to oval nuclei of equal size, which were fairly heavily chromatinized and showed single, large nucleoli. The cytoplasm was scant and frequently not visible. A fairly prominent vascular stroma was present. There was some tendency for the cells to form rings around open areas. In other places, cells were almost completely lacking, being replaced by the fibrillar intercellular substance and by numerous calcareous deposits. Reticulin formation was limited to the rather prominent vascular stroma. The intercellular substance stained blue with phosphotungstic acid hematoxylin in the manner of neuroglial fibrillae.

Diagnosis. Subependymal astrocytoma of the septum pellucidum.


History. The patient was a 51-year-old policeman who had been well, except for a poorly defined recent loss of memory, until 4 months before admission. At that time, he began to complain of headache, dizziness, and a sense of unsteadiness while walking. Two months before admission, he was studied in another hospital following an episode of unconsciousness;
Fig. 2. Photomicrographs of the tumors reported in this series; a, b, and d (Cases 1, 2 and 4) are all subependymal astrocytomas; c (Case 3) is a cellular ependymoma. Hematoxylin-eosin stain.
there he was mentally retarded and was felt to be suffering from a purely mental disease. Subsequently, he experienced episodes of loss of consciousness and disorientation until admission.

**Physical Examination.** Pulse 84, temperature 36.8, respiration 11, and blood pressure 140/90. The patient was disoriented in all spheres although he retained contact with his surroundings to some degree. There were slight tremor of the tongue, suggestive blurring of the left optic disc, and a slow monotonous speech. No other general or neurological abnormalities were found.

**Laboratory Findings.** Blood count and urinalysis were normal. Wassermann and Kahn reactions were negative. Lumbar puncture: Pressure of 300 mm. of water, 6 cells per c. mm. of fluid, and slightly positive Pandy test; Wassermann reaction negative. Radiographs of the skull showed an area of faint calcification in the midline about 4 cm. above the sella turcica. There was a questionable posterior and downward displacement of the shadow of the calcified pineal body. The ventriculogram was essentially similar to that described in Case 1 except that the tumor was less heavily calcified and presented largely into the left lateral ventricle.

**Operation.** Through a left transventricular frontal approach by one of us (PCB), a shiny, gray, encapsulated tumor was exposed, which arose from the septum pellucidum and almost completely filled the anterior horn and body of the left lateral ventricle. It was removed piecemeal. The septum pellucidum was then incised around the mass and that portion of the tumor projecting into the right lateral ventricle was freed thereby and excised.

**Postoperative Course.** The patient was drowsy and semi-stuporous, responding only after persistent persuasion. There was a daily elevation of temperature as high as 103°F for 2 weeks. Occasional incontinence was noted. His mental status was characterized by memory defect and complete indifference, but his general and neurological status was otherwise quite normal. He continued to have episodes of loss of consciousness which were controlled later by moderate doses of phenobarbital. Because of his mental difficulties he was transferred to a mental hospital, where he remained for about 6 months.

**Subsequent Course.** In March 1937, the patient had some attacks "resembling convulsions" which were again controlled by phenobarbital. In April he complained of headache of 2 days' duration. Neurological examination showed no abnormality. Lumbar puncture was normal and a pneumoencephalogram showed no evidence of recurrence of the tumor. In June 1937, a psychological evaluation showed: (1) marked deficit of memory for recent events, (2) paucity of ideational and associational maturity, (3) disturbance in time sense, (4) general slowing-up of perceptual processes, (5) normal simple reaction time. He was not working but was able to carry out normal activities with reasonable ability.

In August 1938, he was readmitted to the hospital because of vomiting and the passage of tarry stools. He was found to have a bleeding peptic ulcer, which was controlled medically. Subsequently, it was learned that he had complained of upper abdominal pains associated with the ingestion of food for as long as 5 years before his cranietomy. Blood pressure at this time was 180/100.

The patient was next seen in 1941. He was not working but was self-sufficient with the aid of a police pension. He managed his own affairs and was independent in all his activities, which included a prolonged trip to Ireland alone.

He was not seen again until January 1947, when he reported that he was completely asymptomatic and was employed as a guard in a bank. He had never married and lived alone, being entirely independent in his activities. He stated that his memory occasionally "played tricks" on him but did not interfere with his work. He further volunteered that for a period of 6 months he could remember little or nothing concerning the events of his operation in 1936. He had had no "fainting spells" since 1938 and was still taking a small amount of phenobarbital regularly. He had had no further gastrointestinal symptoms except for a somewhat troublesome constipation which required regular catharsis. His neurological examination was essentially normal at that time.

**Microscopic Examination of Tissue Removed at Operation.** The tumor (Fig. 2b) was com-
posed of random collections of cells separated and surrounded by streams of fibrillar, acellular material. The cells contained round to oval nuclei showing fairly heavy accumulations of chromatin and one to two nucleoli. There was no particular arrangement of the cells aside from their accumulation into clumps. The intercellular material reacted to differential staining like glial fibrillae. Vascularization was quite prominent but the vessel walls did not appear to be particularly abnormal. Small round areas containing amorphous material and a few small flecks of calcium were scattered throughout the tumor. No connective-tissue stroma was seen other than that around the vessels.

Diagnosis. Subependymal astrocytoma of the septum pellucidum.


History. This 28-year-old housewife was perfectly well until 6 months before admission, when she became aware of visual difficulties. Her visual acuity became somewhat impaired and she complained of periods of “blankness” lasting a few minutes, during which she was unable to see. These difficulties increased until 2 months before admission, when she noticed double vision, persistent frontal headache, occasional vomiting, and a sense of aching in both arms. In addition, she complained of a feeling of numbness in her right leg, also intermittent, and a slight ataxia particularly when walking in poor light. There was also some impairment in her memory. The past history revealed 2 attacks of “St. Vitus dance” in childhood. One brother had epilepsy. Two brothers died, apparently of congenital heart disease. On the maternal side, 5 relatives died of tuberculosis and, on the paternal side, 4 relatives, including the father, died of cancer.

Physical Examination. Temperature 98, pulse 72, respiration 20, and blood pressure 110/80. The patient was well nourished but appeared chronically ill. General physical findings were not otherwise remarkable. On neurological examination, she was well oriented and cooperative but showed some memory deficiency. Visual acuity was sufficient only for light perception. A papilledema of 6 D. was present. There was a slight external strabismus but ocular movements were normal. The reflexes, motor power, and sensation were normal. The patient deviated slightly to the right in walking but the Romberg test was negative. Slight ataxia was present in the left extremities.

Laboratory Findings. Urinalysis, blood studies, and Wassermann reaction were negative. Radiographs of the skull showed no abnormality but a ventriculogram revealed a large filling defect in the region of the anterior portion of the 3rd ventricle on the left side.

Operation. During exposure of the tumor through the right lateral ventricle, such severe bleeding developed that operation had to be abandoned. The patient did not regain consciousness and died 24 hours after operation.

Necropsy. A soft, encapsulated tumor of grayish-red color was found arising from the septum pellucidum. The neoplasm compressed the corpus callosum dorsally and had extended 2 cm. below it ventrally. It extended 1.5 cm. to the right of the midline and 3 cm. to the left; the posterior portion of the tumor on both sides lay free in the widely dilated ventricles. The lateral and anterior portions of the tumor had extended to the region of the basal ganglia, completely filling the anterior portions of the lateral ventricles, particularly on the left side. The tumor was entirely free of the choroid plexus. Except for evidence of operative trauma and bronchopneumonia, the necropsy was not otherwise remarkable.

Microscopic Findings. The sections demonstrated a very cellular tumor (Fig. 2c). The cells were small, of uniform appearance, and were evenly dispersed throughout the tumor. The nuclei were oval and deeply stained, containing clumps of chromatin. The cytoplasm was faintly stained, poorly outlined and scant; no true processes were seen. The cells were close together but occasional small acellular areas were seen, particularly around vessels. Small, thin-walled vessels were numerous but no other stroma was present. The tumor was sharply separated from the surrounding brain but appeared to be seeding along the ependymal lining of the 3rd ventricle. No mitoses were seen.

Diagnosis. Cellular ependymoma of the septum pellucidum.

History. The patient was a 30-year-old housewife who was well until 2 years before admission. At that time, she began to suffer from episodes of headache and vomiting lasting 2 to 3 days. About 1 year later she became aware of increasing difficulty in her ability to move about and began to drag her right leg. It was also noted that she was unable to think clearly; she was confused at times and even hallucinated. She complained, occasionally, of blurred vision and diplopia during this period of time. For 2 months prior to admission, she was incontinent and found it difficult to sit up. Her past history was not contributory.

Examination. Pulse 86, respiration 20, temperature 98, and blood pressure 104/70. General physical examination was not remarkable. Neurologically, the patient was confused and responded very slowly to questioning. There was bilateral papilledema, but visual fields and extra-ocular movements were normal. There was a slight weakness of the left lower face and leg. The Babinski sign was present bilaterally but reflexes were otherwise equal and active. Cog-wheel rigidity was present in the left arm and leg. There were no other abnormal findings.

Laboratory Findings. Blood studies and urinalysis were normal. Radiographs of the skull showed some deformity of the sella turcica suggesting chronically increased intracranial pressure. At ventriculography a greatly dilated left lateral ventricle was evacuated and filled with air; it apparently did not communicate with the 3rd or right lateral ventricles. The left ventricle appeared to be shifted toward the left and a mass could be seen projecting into its anterior third.

Course. Following ventriculography the patient's condition remained unchanged except for short periods during which she would lapse suddenly into unconsciousness. She precipitously expired in one of these episodes before operation could be undertaken.

Neocopy. The general necropsy findings were not particularly abnormal. Grossly, the convolutions of the brain were flattened, and herniations into every possible crevice were present, so that the optic chiasm, 3rd, and 6th nerves were thinned like ribbons. On sagittal section a large grayish, well circumscribed tumor mass, 6 cm. in diameter, was disclosed which pushed the corpus callosum upward and compressed the 3rd ventricle downward so that it was completely occluded. The mass arose from the septum pellucidum and was secondarily attached to the corpus callosum, lateral wall of the dilated right lateral ventricle and lamina terminalis. Both foramina of Monro were obstructed by compression. The tumor mass was easily shelled away from its attachment and was found on section to be of gelatinous consistency, avascular, and full of small cystic areas.

Microscopic Examinations. The appearance of the tumor was identical with that described in Cases 1 and 2 (Fig. 2d). The general appearance was that of an astrocytoma but the grouping of the nuclei was unusual and of the type seen mainly in astrocytomas that develop in the subependymal region.

Diagnosis. Subependymal astrocytoma of the septum pellucidum.


History. The patient was a 30-year-old ordnance inspector who was perfectly well until 2 weeks before admission. At that time he was awakened suddenly by a severe left frontal headache which lasted 5 to 6 hours. The headache recurred 4 days later while the patient was asleep and this time was associated with nausea and vomiting. The severe symptoms subsided but a mild diffuse headache persisted until admission. He was admitted to another hospital where it was found that he had bilateral papilledema and a spinal fluid pressure of 420 mm. of water. His past history was non-contributory.

Examination. Temperature 98, pulse 84, respirations 20, and blood pressure 120/76. The patient was quite well oriented and did not appear ill. Neurological examination showed bilateral papilledema of 3 D., with numerous retinal hemorrhages. There was also slight weakness of movement in the left hand and wrist and in the left ankle.

Laboratory Findings. Blood studies were normal and blood Wassermann reaction was negative. Radiographs of the skull showed no abnormality.

Operative Procedures. Dilated ventricles which did not inter-communicate were demon-
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...trated at ventriculography. A bulging mass extending into both ventricles was clearly visualized. A right subtemporal decompression was done at this time. Subsequently, a left frontal transventricular craniotomy was made and a large blue-walled cystic tumor, 3 cm. in diameter, was exposed. The cystic portion of the tumor contained 10 cc. of xanthochromic fluid and the solid portion was about equal in size. It was quite vascular and removal was felt to be inadvisable. Both foramina of Monro were obstructed by compression. A biopsy was taken and a communication made between the ventricles.

**Postoperative Course.** The patient was well for 3 months. He was then readmitted to the hospital because of recurrent headache and vomiting for 3 days. On examination, he was found to react in a considerably more facetious and euphoric manner than when previously seen. His decompression was bulging but there was only slight elevation of both optic discs and visual acuity was normal. There was still slight weakness of left wrist and ankle. During his stay in the hospital the decompression became soft and he was discharged.

**Microscopic Examination of Tissue Removed at Operation.** Only a small amount of traumatized tissue was available. The predominant cells had the appearance of astrocytes and the tumor was considered to be an astrocytoma.

**Summary of clinical material.** It is evident from a perusal of these cases that the most frequent symptoms in these patients with tumors of the septum pellucidum was headache, which occurred in all cases and usually early. Visual disturbances were less common and, except in 1 case, transitory. Mental deterioration was frequent but in only 1 case severe enough to cause incontinence. Occasional unsteadiness of gait, weakness or numbness of limbs and epileptiform attacks completed the story.

A most striking thing concerning these patients was the frequent paucity of physical findings. The signs most commonly encountered on examination were varying mental confusion, papilledema, and only occasional ataxia, weakness or alteration of reflexes or sensation.

The pressure of the spinal fluid was elevated in each of the 3 cases in which lumbar puncture was done. In 2 cases, radiographs disclosed intracranial calcification; in another an alteration of the sella turcica. In all cases the ventriculograms conclusively indicated a tumor of the septum pellucidum.

**DISCUSSION**

The first attempted resection of a tumor of the septum pellucidum that we have found was reported by Puusepp in 1942. At that time cysts were considered to be the only operable lesions of the septum pellucidum and tumors of this region, even when recognized, were said to carry a very poor prognosis. Since then, 5 cases, including 2 reported in this series, have been described in which operation for such tumors has been successful.

It appears that tumors of the septum pellucidum are much more common than realized, also that neoplasms in this area are often benign and readily removable. It is to call attention to these favorable cases that the tumors reported in the literature are reviewed and evaluated with the 5 cases described in this series.

**Clinical Considerations.** Anatomically, the septum pellucidum lies between and behind the frontal lobes; it is bounded on two sides by the corpus callosum and on the third side by the fornix and third ventricle. In close
proximity to it across the lateral ventricles are the thalami, basal ganglia, and internal capsules, and the foramina of Monro are at the base of the septum pellucidum. Any space-occupying mass in this region, then, is in a very favorable location to interfere with the function of these structures on both sides.

An analysis of these cases indicates that the clinical manifestations of tumors of the septum pellucidum are the result of interference with adjacent areas of the brain, obstruction of the ventricular system, or a combination of these. As is the case with cerebral neoplasms elsewhere the additional element of associated developmental anomalies has been suggested by many observers where it has been difficult to explain symptoms on an anatomical basis. This consideration is not without foundation and will be discussed later; in all probability, however, it is here of minimal importance.

Mental abnormality, particularly related to memory, mood, and initiative, was frequently present in these patients. Such symptoms appeared to be largely the result of frontal lobe involvement whether by direct invasion, interference with association pathways, obstructive atrophy or surgical intervention. Tables 2 and 3 indicate that mental abnormalities were not present when the lesion was confined to the septum pellucidum or when only lateral extension into the region of the basal ganglia occurred. When, however, the corpus callosum was involved or when the tumor was sufficiently large to obstruct the ventricular system or invade the frontal lobes, mental abnormalities usually occurred. Obstruction alone did not produce mental changes in the 2 cases of Cardona. In 2 patients in the present series, mental symptoms were not present until after operative trauma to the frontal lobes; similar experiences were reported by Riskaer and by Caul et al., also after transventricular operations. Occasionally a case such as that of Globus was encountered, in which mental abnormalities were absent even though the tumor was very large.

Many observers felt that the mental abnormalities were a function of developmental defect rather than anatomical interference with lesions of the septum pellucidum. In the case of cysts, for example, Dandy suggested that such lesions denoted a general trend toward cerebral malformations and that these accounted for the mental disturbances in this condition. Bannwarth was of the opinion that only with unusually large lesions were the symptoms due to the cysts themselves. In practically all cases of tumor, however, the mental symptoms developed coincident with other indications of an enlarging mass, hence must be considered primarily of destructive origin rather than the result of disconnected cerebral anomaly. The evidence suggested, rather, a profound variation in the amount of disturbance required to interfere with the function of the frontal lobes in various individuals. Some patients were affected by relatively mild interference with the fronto-callosal pathways while others appeared normal even with large invading and obstructing neoplasms.

Relearning of function lost by resection of part of one frontal lobe after
removal of such a tumor was strikingly illustrated in Case 2 of the present series. This patient did not resume economic independence for 6 years after operation but subsequently became completely self-sufficient even though he was 57 years old and had had profound mental disturbances. It is, of course, conceivable that if he had not been supported by a police pension he might have become employed sooner.

Epileptiform manifestations were present in 2 cases (possibly 3) of this series and in 9 of the cases reviewed. Thus, in 11 out of 36 cases there was evidence of some type of seizure discharge; this percentage is low for all cases of brain tumor. The evidence for associated congenital defect as the etiological factor is a little stronger here than was the case with mental symptoms. Occasionally, as in the case of Bannwarth⁶ and of Riskaer,²⁸ the seizures antedated other symptoms of tumor for up to 17 years and, in still other instances, such as Cases 1 and 3 in the present series and Case 2 of Cardona,¹⁰ there was a family history of epilepsy in patients with tumors of the septum pellucidum. In other instances, the convulsions coincided with the onset of the other symptoms of tumor or even developed as a terminal manifestation. These facts suggest the possibility that the convulsions might have been the result of a congenital defect associated with the neoplasm rather than a symptom actually produced by the tumor. The frequent multiplicity of gross pathological lesions involving the septum pellucidum, which will be discussed later, lends credence to this concept. There is no question, however, that the enlarging tumors accentuated the convulsive tendency, if they were not entirely responsible.

Many of the symptoms presented by these patients were predominantly the result of obstruction of the foramina of Monro. Intermittent occlusion undoubtedly accounted for the periodic headache so commonly seen. Similarly explained were, probably, the evanescent visual³⁶ cataplectic-like³⁸ and auditory symptoms. Chronic obstruction was apparent in the majority of cases (25 to 32), as manifested by the presence of papilledema, dilated ventricles, or elevated spinal fluid pressure. Cranial nerve palsies, particularly of the first six nerves, were easily explained on this basis, as was indicated by the necropsy in Case 3 of this series; here, the 2nd, 3rd, and 6th nerves were compressed to ribbon thinness. The same case illustrated severe compression of the peduncles by herniation, which may account for the hemiparetic states occasionally seen.

The remaining symptoms were probably the result of invasion or compression of the region of the basal ganglia. Ataxia and hemidysesthesias resulted from interference with thalamic and subcortical radiations; masked facies, tremor and cogwheel rigidity from interruption of pallidal outflow; and localized weakness from compression of the internal capsule.

The onset and course of the disease were unpredictable. In many cases, symptoms had been present since childhood, while in others the onset of clinical manifestations was sufficiently severe to bring the patient under immediate attention. A course of months was the usual interval between
recognized onset and hospital admission. The relative malignity of the tumor did not seem to be an important factor in this course although the few patients that had long histories all had benign neoplasms. Benign neoplasms were encountered also in all patients responding satisfactorily to operative treatment.

Of passing interest were the 2 cases in this series in which symptoms of peptic ulcer were present. A similar case was described by Bailey. This problem has been reviewed by Vonderahe and attention called to the relatively high incidence of gastrointestinal ulceration in cases of tumor in the vicinity of the diencephalon.

Two symptoms in patients with tumors of the septum pellucidum previously recorded were not present in the 5 cases reported here. There was no instance of anosmia as discussed by Caul et al. of apraxia as mentioned by Bailey and others.

Radiographically, the tumors were occasionally visualized as circum-scribed areas of increased density in the midline well above the sella turcica. Calcification of the neoplasm was present in 5 of 32 cases (where sufficient data were included for evaluation) of which 4 were probably subependymal astrocytomas. From such radiographic evidence, other calcified tumors of the midline, such as gliomas of the corpus callosum or meningiomas of the falx, were considered frequently in the diagnosis. Ventriculography, however, dispelled all doubts in every case. The characteristic defect, as demonstrated in Fig. 1, can be produced only by a tumor or, if calcification is not present, possibly a cyst of the septum pellucidum. In either case, localization is absolute.

To summarize the clinical manifestations mentioned above, gliomas of the septum pellucidum occur predominantly in young adults; the course varies from a few months to years. The early symptoms are mainly those of episodes of headache, visual symptoms, or cataplectic-like weakness; mental abnormality; and possibly epilepsy. Later manifestations are those of progressively severe organic mental disease, symptoms of increased intracranial pressure, and localized motor or sensory disturbances.

The above clinical description is clearly not pathognomonic of tumors of the septum pellucidum. The symptom-complex can be reduplicated by cysts of the septum pellucidum, and closely simulated by tumors of the corpus callosum, of the third ventricle, of the lateral ventricles, of the falx and of the frontal lobe. Even hemorrhage or congenital deformity of the septum pellucidum may present somewhat similar manifestations. Careful clinical evaluation and radiographic studies will serve to indicate the correct location of the lesion but its exact nature can be determined only by operative intervention in most cases.

There has been some discussion as to the most suitable operative approach to tumors of the septum pellucidum. In most of the successfully operated cases, the approach has been via the transventricular route. In the 2 cases reported in this series, there appeared to be little choice between a
right and a left-sided exposure (both patients were right-handed). The alternative route has been by way of the sectioned corpus callosum; it has been employed particularly for cysts, although Gherzi used this route for tumors. Until further evidence is available, decision for operative approach must remain one of individual preference.

The results of operation are quite hopeful for these previously fatal neoplasms. To date there have been at least 5 cases in which the tumor was successfully resected. In 4 of these cases the tissue removed was diagnosed as, or strongly resembled, a subependymal astrocytoma. The result was excellent 11 years after operation in 1 patient. Three cases can be considered to show satisfactory results and 1 only fair. It must be noted, however, that all 4 of these latter cases were observed less than 2½ years after operation, which is too short a time for accurate evaluation, judging from our experience with Case 2.

Pathological Considerations. Examination of the pathological material available in these cases of neoplasm of the septum pellucidum indicates that a wide variety of tumor-types occur in this area. As the septum pellucidum doubtless has an origin similar to that of the cerebral hemisphere embryologically, it is not surprising to find essentially similar neoplasms in both places. The following kinds of neoplasms in the septum pellucidum are recorded: Astrocytoma, by Bailey, Cardona and by Riskaer; spongioblastoma unipolare by Marras; spongioneuroblastoma and cellular ependymoma by Globus; glioblastoma multiforme by Puusepp; astroblastoma by Gherzi; and oligodendroglioma by Caul et al.

The probable origin of the various tumors of the septum pellucidum is of fundamental interest in a discussion of this subject. It has been long recognized that the subependymal region was particularly important as the site of origin of many gliomas. Bailey and Cushing in 1926 called attention to such periventricular areas in this connection. Globus in 1942 described a series of 21 cases in which he was able to trace neoplasms to formations of relatively undifferentiated cells in the subependymal plate or “matrix.” This matrix was particularly prevalent around the thalamostriate groove, head of the caudate nucleus, and septum pellucidum. He considered that these undifferentiated cells were bipotential, that the resultant tumors frequently contained neuroblastic as well as spongioblastic elements and that they were predominantly malignant and invasive in nature. In his series, 5 of 21 tumors arose from the septum pellucidum.

In 1945 Scheinker described a type of neoplasm which appears to be identical with that present in Cases 1, 2 and 4 reported here. He thought that these tumors arose from the subependymal plate because of their resemblance to the tissue found in that location and because of the location of the tumors. His 7 tumors all arose in the floor of the fourth ventricle. He called these tumors subependymomas. However, this name is not suitable as it is but a localizing and not a descriptive term. As the present review shows, many different types of tumors may arise in the subependymal region. From
a study of our material, in Cases 1, 2 and 4, which appears to be identical with his, we conclude that these tumors are astrocytomas. As they differ somewhat in their appearance from astrocytomas to be found elsewhere we have designated them “subependymal astrocytomas.” Similar tumors arising in the septum pellucidum have been reported by other authors. Those reported by Ghersi (Case 1),17 by Riskaer (Case 1),28 and by Souques et al.31 appear to be subependymal astrocytomas; while those reported by

<table>
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<th>Operation or Necropsy</th>
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<td>Necropsy</td>
<td>Protoplasmic astrocytoma</td>
<td>Invaded corpus call. &amp; frontal lobes.</td>
</tr>
<tr>
<td>Urechia &amp; Kernbach33</td>
<td>1929</td>
<td>9</td>
<td>Necropsy</td>
<td>Astrocytoma</td>
<td>Confined to septum pell.</td>
</tr>
<tr>
<td>Barré &amp; Fontaine7</td>
<td>1930</td>
<td>10</td>
<td>Operation Not removed</td>
<td>Fibrillary glioma</td>
<td>Patient died 34 hrs. after op.</td>
</tr>
<tr>
<td>Marchand31</td>
<td>1930</td>
<td>11</td>
<td>Necropsy</td>
<td>Glioma</td>
<td>Compressed corpus call.</td>
</tr>
<tr>
<td>Marras22</td>
<td>1933</td>
<td>13</td>
<td>Necropsy</td>
<td>Spongioblastoma unipolar to astrocytoma</td>
<td>Large tumor invading surroundings.</td>
</tr>
<tr>
<td>Dandy14</td>
<td>1934</td>
<td>14</td>
<td>Operation Unsuccessful (case 9)</td>
<td>Embryonal tumor of ependyma</td>
<td>Confined to septum pell. &amp; left lat. vent. Died after op.</td>
</tr>
<tr>
<td>Reported by</td>
<td>Year</td>
<td>No.</td>
<td>Operation or Necropsy</td>
<td>Diagnosed</td>
<td>Notes</td>
</tr>
<tr>
<td>-------------</td>
<td>------</td>
<td>-----</td>
<td>-----------------------</td>
<td>-----------------------------</td>
<td>--------------------------------------------</td>
</tr>
<tr>
<td>Cardona</td>
<td>1936</td>
<td>15</td>
<td>Necropsy</td>
<td>Fibrillary astrocytoma</td>
<td>Compressed corpus call.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>16</td>
<td>Necropsy</td>
<td>Cystic astrocytoma</td>
<td>Invaded caudate nucleus to int. capsule.</td>
</tr>
<tr>
<td>Bannwarth</td>
<td>1939</td>
<td>17</td>
<td>Operation</td>
<td>Calcified glioma</td>
<td>Cond. satisfactory 6 wks. after op.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Not removed</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Puusepp</td>
<td>1942</td>
<td>18</td>
<td>Operation</td>
<td>Glioblastoma</td>
<td>Probably arose frontal lobe.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>19</td>
<td>Operation Removed</td>
<td>Astrocytoma</td>
<td>Invaded frontal lobe. Reported well 6 mo.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>20</td>
<td>Operation</td>
<td>Oligodendroglioma</td>
<td>Compressed both frontal lobes. Died 6th day.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Not removed</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>22</td>
<td>Necropsy (case 2)</td>
<td>Same</td>
<td>Invaded frontal lobe, corpus call. &amp; fornix.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>23</td>
<td>Necropsy (case 6)</td>
<td>Spongioblastoma mult.</td>
<td>Invaded left cerebrum &amp; basal ganglia.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>24</td>
<td>Necropsy (case 9)</td>
<td>Spongioneuroblastoma</td>
<td>Infiltrated corpus call. Multiple gliomas.</td>
</tr>
<tr>
<td>Ghersi</td>
<td>1943</td>
<td>26</td>
<td>Operation</td>
<td>Cystic glioma with calcification</td>
<td>Invaded corpus call. &amp; 3rd vent. Died 3rd day.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>27</td>
<td>Same</td>
<td>Astroblastoma</td>
<td>Died 1st day. Large invasive tumor.</td>
</tr>
<tr>
<td>Caul et al.</td>
<td>1948</td>
<td>28</td>
<td>Operation Removed</td>
<td>Glioblastoma with preponderance of oligodendroglia</td>
<td>Cond. satisfactory 2½ years after op.</td>
</tr>
<tr>
<td>Riskaer</td>
<td>1944</td>
<td>30</td>
<td>Operation Removed</td>
<td>Atypical encapsulated astrocytoma</td>
<td>Postop. torpor. Cond. good 21 mo.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>31</td>
<td>Operation</td>
<td>Edematous astrocytoma</td>
<td>Cond. unchanged 14 mo.</td>
</tr>
</tbody>
</table>

The reported tumors of the septum pellucidum are listed in order. The clinical and pathological descriptions were frequently poor, and in a few cases (e.g., Puusepp's) the tumors were considered by others to have invaded the septum pellucidum secondarily. One case was not included because sufficient data were not available, and 1 because of questionable localization.
Wolf, Caul et al., and by Riskaer (Case 2) were quite similar. It is probable that all of the tumors that have been successfully removed were subependymal astrocytomas.

In view of the above data there appears to be an interrelationship in the origin of these various quite dissimilar tumors and even possibly of congenital cystic abnormalities of the septum pellucidum. There is considerable evidence to indicate that all of these processes may diverge from embryological abnormalities in the anlage of the septum pellucidum. The persistence of zones of embryologically undifferentiated cells in the subependymal plate have been clearly indicated by Bailey and by Globus and the more malignant tumors have been attributed to these. The benign tumors show a stronger resemblance to congenital cystic abnormalities of the septum pellucidum than to such malignant tumors. Microscopically, subependymal astrocytomas reduplicate the appearance of the normal subependymal region and strikingly resemble the tissue shown by Meyer in his paper concerning cysts of the septum pellucidum. The cells in all such states are adult, differentiated astrocytes and masses composed of them resemble more heterotopia than neoplasia. In this connection, Bailey has previously suggested that fibrillary astrocytomas are perhaps not blastomata at all but of the nature of heterotopia. Bergstrand has made the same suggestion.

The coexistence of multiple abnormalities involving the septum pellucidum and cerebral hemispheres lends some support both to the common origin of these neoplastic and developmental states and to their interrelationship. Not only have multiple glial tumors been described in this connection, but also various admixtures of neoplastic and congenital deformi-
<table>
<thead>
<tr>
<th>Group</th>
<th>Tumor extent and involvement</th>
<th>Case of</th>
<th>Mental symptoms and epilepsy</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Confined to septum pellucidum</td>
<td>Urechia and Kernbach</td>
<td>Neither reported.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Bannwarth</td>
<td>Neither reported.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Globus</td>
<td>Neither reported persisting.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dandy⁴</td>
<td>Neither reported.</td>
</tr>
<tr>
<td>II</td>
<td>Involved septum pellucidum plus thalamus or basal ganglia</td>
<td>Saltykow</td>
<td>Neither reported.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cardona</td>
<td>No mental symptoms. Patient and mother had epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Riskaer</td>
<td>No mental symptoms. Long history of &quot;spasms&quot; of left side.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Bailey⁵</td>
<td>Neither reported.</td>
</tr>
<tr>
<td>III</td>
<td>Involved septum pellucidum plus corpus callosum. No obstruction c-s.f.</td>
<td>Marchand</td>
<td>Mental difficulties predominated. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Wolf</td>
<td>Theatrical behavior. Epilepsy.</td>
</tr>
<tr>
<td>IV</td>
<td>Invaded or compressed surrounding structures. Obstructed c-s.f.</td>
<td>Bailey⁴</td>
<td>Loss of memory and indifference. Myoclonic twitching.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Souques et al.</td>
<td>Confused and disoriented. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cardona</td>
<td>Lucid but somewhat restrained. Epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Puusepp</td>
<td>Apathy. Disturbance of thought and orientation. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Case 2</td>
<td>No interest in surroundings. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Case 3</td>
<td>Difficulty in thinking. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Case 8</td>
<td>Indifference. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Case 2</td>
<td>Responses vague. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Gherzi</td>
<td>Impaired memory. Emotional instability. No seizures.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Globus</td>
<td>Impaired memory. Seizures.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Case 2</td>
<td>Difficulty concentrating. No seizures.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Case 6</td>
<td>Loss of memory. Melancholia. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Case 9</td>
<td>Loss of memory. Depression. Epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Caul et al.</td>
<td>Irritability. No other mental symptoms. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Globus²</td>
<td>Psychically unstable. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Riskaer</td>
<td>Psychic torpidity. Convulsions.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Marras</td>
<td>&quot;Witzelsucht.&quot; Convulsions.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Moniz and Lima</td>
<td>Neither reported.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Berne</td>
<td>Uncooperative. Mentally dull. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Bailey⁵</td>
<td>Loss of initiative and lack of interest. No epilepsy.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dimitz &amp; Schilder</td>
<td></td>
</tr>
</tbody>
</table>

The symptoms of mental abnormality and of epilepsy are evaluated according to the size and degree of involvement of the tumors previously reported. Mental defects appear to correlate more closely with the extent of the tumor.

Multiple congenital abnormalities involving the septum pellucidum are common.¹²,²²

The pathological nature of these various tumors of the septum pellucidum and available indications concerning their origin serve to clarify the clinical manifestations resulting from their presence. The commonly en-
countered subependymal astrocytomas must be clearly differentiated from the more malignant tumors in this area and their radical removal undertaken. It is to be expected that such tumors will be more frequently recognized and intelligently managed as experience in this matter accumulates.

SUMMARY

1. The clinical and pathological material of 5 cases of tumor of the septum pellucidum is presented. Three of these tumors (possibly 4) were subependymal astrocytomas and 1 was a cellular ependymoma. Two tumors were resected; 1 patient is alive and well 11 years after operation and the other for a much shorter time.

2. The published data on 31 similar cases were examined. Only 3 (possibly 4) were successfully operated upon and these all within the last 5 years. Gliomas of various kinds were diagnosed microscopically in these cases; in 6 instances the published material strongly suggests that they were subependymal astrocytomas. The probable origin of these various tumors in the subependymal plate was discussed.

3. Tumors of the septum pellucidum predominantly affect young adults. The clinical manifestations of all space-occupying masses of the septum pellucidum are similar and are predominantly the result of local compression and obstruction.

4. The common symptoms associated with this condition are: Episodes characterised by headache, occasionally visual and aural disturbances, and less frequently cataplectic-like weakness; mental disturbances, particularly related to memory; occasionally convulsions; less commonly unsteadiness or weakness of movement or numbness.

5. The predominant findings associated with this condition are: Mental defects, frequently papilledema, less commonly pareses, paresthesias, ataxia and defects of movement or tonus.

6. Radiographic studies are frequently diagnostic. Calcification in the tumor is occasionally visible. The ventriculogram shows dilated, separated lateral ventricles with biconcave medial borders.

REFERENCES

TUMORS OF THE SEPTUM PELLUCIDUM