HYDROCEPHALUS FROM OVERPRODUCTION OF CEREBROSPINAL FLUID

(AND EXPERIENCES WITH OTHER PAPILLOMAS OF THE CHOROID PLEXUS)*

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Papillomas of the choroid plexus are rare tumors. There were but 12 (0.6 per cent) in Cushing's series of 2023 intracranial tumors. This has been the approximate percentage in other large clinics. We are presenting our series of 7 of these tumors because one case seems to prove beyond a doubt the commonly held view that hydrocephalus can on occasion be caused by an overproduction of cerebrospinal fluid.

In the excellent monograph on the pathology of hydrocephalus by Dorothy Russell the three generally accepted ways in which hydrocephalus can be produced are clearly stated: "first, through over-secretion from the plexuses; secondly, through the interposition of an obstruction at some point in the cerebro-spinal pathway; or, thirdly from impairment of absorption."

It is Doctor Russell's belief that though this first method of the production of hydrocephalus may exist, there is not as yet a well enough documented case to give absolute proof that hydrocephalus can result from the overproduction of cerebrospinal fluid alone.

CASE REPORTS

Case 1. R.W., a 20-year-old housewife, entered the hospital in July 1946 because of headaches of 5 years' duration. They had been particularly severe during and after a pregnancy which had terminated normally 3 weeks before.

Examination. There was papilledema of 4 D. with hemorrhages. Visual fields showed concentric contraction. Otherwise, the neurological findings were normal. X-rays of the skull were entirely normal.

Ventriculography revealed marked symmetrical dilatation of the ventricular system, with air in the 4th ventricle and in the cisterna magna (Fig. 1). There seemed to be a small amount of air in the cisterna interpeduncularis. There was a mass the size of a walnut in the region of the glomus of the choroid plexus in the left lateral ventricle (Fig. 2).

Clinical Diagnosis. The case was discussed with Dr. Max M. Peet, whose opinion was that this was a papilloma of the choroid plexus with an overproduction of cerebrospinal fluid. The operator (E.A.K.), however, wished to rule out a lesion of the posterior fossa which might be producing a ball-valve action. It was decided, there-

fore, that the posterior fossa should be explored first and, should this be normal, the mass would then be removed from the left lateral ventricle.

Operation, Aug. 16, 1946. The cerebellar approach was made through a curved transverse incision. The occipital bone was of usual thickness. The cisterna magna was exceedingly large; its arachnoid appeared normal. The tonsils of the cerebellum were in normal anatomical position. The 4th ventricle was entered and CSF could be seen coming from the aqueduct of Sylvius. The cerebellar hemispheres appeared normal and were not needled. The wound was closed in the usual manner. It was now believed certain that the significant lesion lay in the left lateral ventricle. The patient was immediately placed on her side and an osteoplastic flap was turned down, centering over the left parietal eminence. A convolution was selected which was thought to be just posterior to the postcentral gyrus and above the supramarginal gyrus. An opening was made here into the lateral ventricle. A tumor about the size of a walnut, yellowish, quite smooth, and firm in consistency was seen arising from the choroid plexus itself. The entire tumor was removed, but some choroid plexus was left in the temporal horn.

Microscopic Diagnosis. Papilloma of the choroid plexus (Fig. 3). The choroid plexus, containing psammoma bodies, from which the tumor had arisen was evident histologically (Fig. 3B).

Course. There was a marked receptive aphasia for several weeks but this soon disappeared. The papilledema
cleared and there were no residual neurological signs. The patient is entirely well 5 years postoperatively except for occasional headaches which she states are nothing "compared to the ones before operation." She has since had another child, who is now 3 years old. The patient is pregnant again at the present time.

Discussion. In Case 1 a symmetrical dilatation of the entire ventricular system was evident. Air was present in the cisterna magna in large quantity and some appeared to be in the cisterna interpeduncularis as well. This shows that an obstructive hydrocephalus was not present, a fact also borne out by exploration of the 4th ventricle.

It is believed that nothing was done to open the communicating channels as no radical search was made beneath or around the cerebellar hemispheres, a procedure which could possibly have aided the cerebrospinal fluid in reaching the surface of the cerebral hemispheres. Again, nothing was done to improve the absorbing mechanism and the adding of blood to the cerebrospinal fluid might conceivably have diminished its absorption.

Since all symptoms of increased intracranial pressure have been absent for 5 years following excision of a papilloma of the choroid plexus of a lateral ventricle, and nothing further was accomplished by the operative procedures, it must be assumed that the hydrocephalus was caused in the first
place by an overproduction of cerebrospinal fluid. The tumor undoubtedly functioned as a tremendously hypertrophied choroid plexus.

Case 2. D.J., a 2-year-old girl, entered the hospital June 2, 1950 with a 2-week history of ataxia, dragging of the left leg, lethargy, headaches, nausea, vomiting, and generalized tremors of the extremities. A left convergent strabismus developed 2 days before admission.

Examination. She was a restless, irritable child who was unable to sit or stand by herself. The head was definitely hydrocephalic and emitted a cracked-pot sound on percussion. There was bilateral papilledema. A left 6th nerve palsy was present. There was marked ataxia and an intention tremor of the left upper extremity with athetoid movements. X-rays of the skull showed evidence of increased intracranial pressure. Ventriculograms showed a marked displacement of the ventricular system to the left with a widely dilated left lateral ventricle.

1st Operation. On June 6, 1950, a right temporal craniectomy, 3 cm. in diameter, revealed a large vascular subcortical tumor involving the temporal lobe. Because of the extent of the lesion and marked operative hemorrhage, only a biopsy was done. It was the operator’s opinion that the tumor was a glioma.

Pathological Diagnosis. Papilloma of the choroid plexus.

2nd Operation. One month later, because of the benign microscopic appearance of the tumor, a second operation was performed with further removal of the growth. Again because of severe hemorrhage, the operation was abandoned after considerable tumor had been removed.

Course. The patient did well and was discharged home 2 weeks after operation. She returned to the hospital 2 months later. There was improvement in her walking. A left hemiparesis was present. Babinski’s sign was bilaterally positive. She was started on irradiation therapy in the right and left lateral mid-skull fields, receiving 100 r. to two fields per day for a total of 2000 r. per field. Her condition seemed to deteriorate throughout her stay in the hospital. The last x-ray treatment was given on Nov. 16, 1950.

She was brought back to the hospital on Dec. 10, 1950, apparently moribund. A ventriculostomy tube was placed in the left lateral ventricle for 5 days. Ventriculograms showed little change from previous examination. She was discharged on Jan. 5, 1951, just able to sit up.

Two months later the patient returned to the hospital remarkably improved to the extent that she could stand and walk with support. Though the decompression was bulging, it was thought that the tumor might have been sufficiently reduced in size by x-ray therapy to make removal feasible. Accordingly, ventriculography was again performed. In comparing the ventriculograms with those taken just after irradiation therapy, the actual tumor outline could still be seen and had not changed in any way.

The patient returned home and 8 days later began showing signs of increasing intracranial pressure. She re-entered the hospital moribund and died April 10, 1951.

Autopsy. A massive infiltrative papilloma of the right temporo-occipital parietal area projected into the lateral ventricle, but remained entirely subependymal, as far as could be determined (Fig. 4). The tumor seemed to be separated from the choroid plexus of the right lateral ventricle by ependyma so that it could not be determined for certain from where this typical papilloma arose. Both foramina of
Monro were patent and the 4th ventricle was dilated. There was no evidence of obstruction of the ventricular system.

Microscopically the papilloma showed no definite evidence of x-ray change in the sections examined; in fact, it appeared generally more cellular than the specimens taken at biopsy before x-ray treatment was instituted (Fig. 5).

Discussion. Van Wagenen⁶ reported a papilloma of the choroid plexus of the lateral ventricle in a girl of 3 months. The neoplasm was histologically similar to that in our Case 2, though smaller and noninfiltrative in character. After a biopsy of the lesion was done, x-ray therapy was given. At a second operation the tumor appeared to be smaller

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Fig. 4. Case 2. Coronal section showing that the tumor was apparently entirely subependymal. Choroid plexus is seen but whether the neoplasm arose from its undersurface could not be determined with certainty.

Fig. 5. Case 2. (A) Biopsy of tumor showing typical picture of a choroid plexus papilloma. (B) Tumor at autopsy. Whether this is postirradiation effect is questionable.
and less vascular and was believed to have been totally removed. The child died 2 years later, but the cause of death is unknown.

In our patient, following decompression and two partial excisions of the tumor, there was little change in condition. X-ray therapy was given and 8 days following the last treatment the child was continuing to go downhill. Then a little over 2 months after the last x-ray treatment, she started to improve. The reason for this is difficult to explain since the decompression had been present for 9 months and the lesion as shown by ventriculograms did not seem to have changed in size.

It might be postulated that at this late date an oversecretion of cerebrospinal fluid had been reduced by x-ray. This theory, however, is scarcely tenable since a communication between the tumor and the ventricular system was not obvious. The marked hydrocephalus found here in the absence of ventricular obstruction may have been due to obstruction in the communicating channels or the absorptive mechanism, though this was not demonstrable.

Case 3. A.S., a 5-month-old male infant, was admitted because of progressive enlargement of the head. The child was thought to be normal in all other respects.

Examination was completely negative except for moderate hydrocephalus. There was no bulging of the fontanelles. A ventricular tap was performed and the cortex was judged to be about 1 cm. in thickness. Methylene blue was instilled and recovered by the lumbar route. A ventriculogram disclosed evidence of a tumor of the 3rd ventricle.

Operation. On July 23, 1948 a right temporoparietal osteoplastic craniotomy was performed. Exploration through the right lateral ventricle revealed the tip of a solid tumor lying in the posterior part of the 3rd ventricle. Total removal was impossible, so that a biopsy was taken and the incision closed.

Pathological Diagnosis. Well differentiated papilloma of the choroid plexus.

Postoperative Course. The patient never fully rallied, having frequent bouts of hyperthermia. His course continued downhill and respirations ceased on the 21st postoperative day. Permission for autopsy was denied.

Case 4. R.W., a 19-month-old boy, had been well until September 1950, when he struck his head on a cement sidewalk. Three days later he began to vomit and this was said to be projectile in type. One week later the gait was noted to be slightly ataxic, and he then entered the University Hospital.

Examination. There were no signs of increased intracranial pressure. The optic disks were sharply outlined. On lumbar puncture the pressure was 150 mm. of water, but the total protein was 140 mg. per cent.

Course. He soon became symptom free. There were no other abnormal neurological findings, and he was discharged.

Six weeks later staggering occurred, and he was taken to another hospital for study. A generalized weakness and lack of coordination were noted. Lumbar puncture revealed normal pressure and a protein of 246 mg. per cent. Ventriculography on Nov. 24, 1950 revealed suggestive evidence of a tumor of the 4th ventricle. A medulloblastoma was suspected and x-ray therapy advised.

Readmission. The patient was returned to University Hospital on Dec. 2, 1950.
Hydrocephalus from Overproduction of CSF

He was extremely ill and could no longer be aroused. A catheter was placed in the right lateral ventricle. Ventriculography showed definite evidence now of a tumor of the 4th ventricle.

Operation. Following 2 days of ventriculostomy drainage there was no improvement in the child's precarious condition, and a suboccipital craniectomy was performed. A large reddish-gray vascular tumor was removed from the 4th ventricle, except for a small amount that infiltrated the obex and extended into each lateral recess.

Pathological Diagnosis. Well differentiated papilloma of the choroid plexus.

Course. Convalescence was surprisingly good, and 1 year postoperatively the child seems normal in all ways. Because of the adult histological character of the growth, x-ray therapy was not given.

Discussion. Case 4 shows that a papilloma of the choroid plexus of the 4th ventricle in a child cannot be differentiated clinically from a medulloblastoma. A cerebellar astrocytoma that shows no localization by symptomatology or ventriculography can also mimic a medulloblastoma if its onset is comparatively acute. We are therefore opposed to x-ray therapy, before a lesion has been verified histologically, on the grounds that it is probably a medulloblastoma.

Case 5. V.A.K., a 40-year-old male, entered the hospital complaining of severe generalized headaches for the past 3 years. Three months before admission, he had had blurring of vision, diplopia, nausea, and vomiting.

Examination showed bilateral papilledema and a 6th nerve palsy on the right. There was no facial weakness. Diminution of auditory acuity was noted on the right. There were no signs of cerebellar disturbance. X-rays of the skull were normal. Ventriculography revealed evidence of an obstructive lesion in the posterior fossa.

Operation. A suboccipital craniectomy was performed, disclosing a reddish tumor in the 4th ventricle lying just cephalad to the obex. The tumor was removed in a piece-meal fashion until the aqueduct of Sylvius was exposed, and there was a free flow of cerebrospinal fluid through the aqueduct. The neoplasm was the size of a walnut and had infiltrated the medulla.

Pathological Diagnosis. Papilloma of the choroid plexus.

Postoperatively the patient never gained consciousness and expired 2 days later. Postmortem examination was not permitted.

Case 6. O.K., a 36-year-old male, was admitted on Nov. 23, 1933 with the history of deafness in the right ear for the past 5 years, and dysarthria and dysphagia for the past 6 months. Three months before admission there had been blurring of vision and ataxia.

Examination. There was bilateral papilledema. There was no sensory disturbance in the distribution of the right 5th nerve although the corneal sensation was a trifle less acute on the right as compared to the left. An almost complete peripheral right facial palsy was present. There was deviation of the soft palate to the left on phonation, and complete paralysis of the right vocal cord. X-rays of the skull showed irregular destruction of the right petrous ridge, and clouding of the mastoid cells on the right.

A biopsy of the lesion through the right middle ear was reported "an atypical papilliferous growth with the papillae and intervening spaces lined by a single layer
of cuboidal or columnar epithelium. The papillae are vascular and in some areas are indistinguishable from the choroid plexus."

Operation. A suboccipital craniectomy was performed, with disclosure of an extensive tumor lying in the right cerebellopontine angle. It was composed in large part of small cysts containing fluid varying in color from yellowish to dark green. This mass invaded the hemisphere of the cerebellum and appeared to pass through the incisura of the tentorium. Because of hemorrhage, it was possible to perform only a partial removal of the tumor.

Pathological Diagnosis. Papilloma of the choroid plexus.

Course. The patient made an uneventful recovery and was discharged on the 18th postoperative day. He never again was able to work and died 2 years later outside the hospital.

Discussion. An almost identical lesion in a man aged 26 was reported by Berlin. The tumor had been considered to be a papilloma of the choroid plexus, but was finally diagnosed as a ceruminous adenoma involving the cerebellopontine angle. This case is unique and is mentioned only so that it can be considered in the differential diagnosis.

Case 7. J.K., a 47-year-old male, entered the hospital because of frontal headaches, of 2 months' duration, which radiated to the occiput. He had had two generalized convulsions and staggering during this period.

Examination. Significant findings were ataxia with falling to the right and bilateral papilledema. X-rays of the skull were normal. Ventriculograms indicated a right-sided posterior fossa lesion.

1st Operation. On April 8, 1936, a suboccipital craniectomy was performed. A grayish-black tumor, larger than a golf ball, was seen on the surface of the right cerebellar hemisphere. It was thought to be a meningioma though it had no dural attachment, and the entire tumor was excised grossly.

Pathological Diagnosis. Papillomatous adenocarcinoma of the choroid plexus.

Course. Convalescence was uneventful, and the patient was discharged 10 days postoperatively. He soon resumed his work in a factory.

One year later he returned to the hospital because of a recurrence of his symptoms of 2 weeks' duration. He was extremely ill and was immediately taken to the operating room.

2nd Operation. The old incision was reopened. A large cyst filled with dark brownish fluid, as well as an olive-sized blackish granular tumor on the inferior lateral surface of the tentorium on the right were removed.

Pathological Diagnosis. Recurrence of papillomatous adenocarcinoma of the choroid plexus.

Course. The patient withstood the operation remarkably well and his convalescence was surprisingly smooth. He was discharged on the 14th postoperative day. He again resumed his work in a factory for at least 9 months. He died of recurrence 16 months postoperative. Autopsy was not performed.

Discussion. This was the only case in our series where a papilloma of the choroid plexus was frankly malignant.

SUMMARY

The conception that hydrocephalus can be produced by an overproduction of cerebrospinal fluid is proven by Case 1.
HYDROCEPHALUS FROM OVERPRODUCTION OF CSF

Six cases of benign papilloma and 1 of adenocarcinoma of the choroid plexus are presented. Two arose from the lateral ventricle, 1 from the 3rd ventricle, 2 from the 4th ventricle, 1 in the cerebellopontine angle, while the adenocarcinoma seemed to arise from the cerebellum itself. These neoplasms are reported as occurring most commonly in the lateral ventricle during the first decade of life, and in the 4th ventricle during the 2nd, 3rd and 4th decades.

Surgical experience with this tumor started with Bielschowsky and Unger in 1902. Since that time less than 100 choroid plexus papillomas have been reported, the majority having been disclosed at autopsy. Of the patients with papillomas that were removed surgically, about one half survived the immediate postoperative period.

In our series, only 1 patient has a survival period of 5 years and is assumed to be cured. One patient is alive 1 year postoperative and has no symptoms of recurrence. Another lived 9 months postoperatively and has recently died. Two patients survived for 2 years and have died, most likely from recurrences of their neoplasm. Two died in the immediate postoperative period.

Weinstein stated that these tumors are usually demarcated if not encapsulated, and this has been our experience. In spite of their benign character, however, their location in recesses of the 4th ventricle and their adherence to the brain stem usually preclude total removal.

Deep roentgen therapy has been used so infrequently that its effectiveness cannot be determined adequately. One would hardly expect, however, such well differentiated neoplasms to respond to irradiation therapy. It is possible that any beneficial effect of x-ray may be attributed to diminishing the overproduction of cerebrospinal fluid.

REFERENCES
5. Russell, D. S. Personal communication.