PAPILLOMA OF THE CHOROID PLEXUS
WITH REPORT OF AN UNUSUAL CASE

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Papilloma of the choroid plexus is a rare intracranial tumor. In a survey of the literature, less than 100 reported cases have been found and this particular tumor comprised but 0.5 per cent of Cushing's series of 2,000 verified intracranial new growths. The incidence of malignant characteristics in choroid plexus papilloma is rarer still. It seems of interest, therefore, to report the following case, which was originally diagnosed as an adenocarcinoma of choroid plexus origin, since the patient has a survival period to date of over ten years. Furthermore, a review of the pathologic material at the present time has not influenced a change in the original diagnosis.

According to Posey,1 papilloma of choroid plexus origin was first described by Guerard in 1892, and Van Wagenen2 states that the first surgical removal was attempted by Bielschowsky and Unger in 1902. This case, however, had a fatal outcome. Perthes in 1919 reported the first successful extirpation of a choroid plexus papilloma,1,3 but there seems to be some question regarding the pathologic histology in light of more recent classification. For this reason, the case reported by Sachs4 three years later with a survival period of six years (one of the longest ever reported to date) is generally recognized as the first successful attack on such a tumor. Davis and Cushing3 reported in 1925 the first 6 cases of Cushing's total of 12, and in 1930 Van Wagenen5 compiled what is still a classical review of the literature with investigation of all reported cases up to that time. He added 2 cases of his own—one surgical and one from autopsy material. Posey,6 in 1942, again reviewed the literature, and with the addition of a single case which came to autopsy, brought the recorded total at that time to 86.

The diagnosis of papilloma of the choroid plexus has been made from operative material relatively infrequently in the total reports of this lesion. Van Wagenen recorded only 11 operative cases with successful immediate outcome in 6, the survival period ranging from three months to six years. Posey states that at the time his paper was written, 22 cases had been diagnosed surgically, with complete recovery in but 5.

In his summary as to location, Van Wagenen states that 50 per cent of the growths have been in the fourth ventricle, 34.7 per cent in the lateral ventricles and 17.3 per cent in the third ventricle.

The age group is of some interest with the incidence seemingly greatest in early life. Posey reported that only 18 cases could be found occurring in persons over 40 years of age.

The question of malignancy in these papillomas has previously provoked discussion and, of course, in attempting to classify the earlier reported cases in the light of present classification, further difficulties arise. Davis and Cushing quoted Hart as stating that the literature contained records of only 2 examples of choroid plexus papillomas with carcinomatous changes. They were indefinite in their conclusions as to the carcinomatous tendencies, but pointed out that true papilloma of choroid plexus origin is usually a single lesion, whereas the previously reported cases accompanied by “seeding” of the tumor were more likely papillary ependymomas. As recently as 1942, Posey pointed out that since these tumors were observed in only a few persons of cancer age, it was not surprising that only 2 cases with histologic cancer were to be found in the literature. We have found no additional cases. Ewing,7 however, refers to these tumors as a group as “papillary adenocarcinomas of the choroid plexus.”

The following case is of interest, therefore, in that it concerns a papilloma of the choroid

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plexus occurring in the fourth ventricle in a 44-year-old man and reported originally as adenocarcinoma. Ten years subsequent to his first admission, having survived three successful extirpations of the recurrent tumor, the patient is alive and relatively well. On reviewing the pathologic material, there is a striking similarity in the three specimens, and on the basis of this material, the present diagnosis is still papillary carcinoma of the choroid plexus.

CASE REPORT

G.E.P., a 44-year-old salesman, was referred by Dr. J. S. Jamieson of Portland, Maine, and was admitted to the New England Deaconess Hospital on April 21, 1936, complaining of unsteadiness of gait of 9 months' duration.

The patient had been perfectly well until the previous June, when, following a long drive one evening, he had a sudden episode of unsteadiness and was forced to seek support to keep from falling. Similar episodes recurred about once a month and lasted for 2 or 3 minutes, being followed by unsteadiness in walking of about 10 minutes’ duration. There was no nausea nor vomiting at the time and no tinnitus. There had been partial deafness of the left ear for 33 years, with no recent change in this defect. For 6 months prior to admission he had been continually unsteady with improvement on lying down. Two months before admission he began to have episodes of vomiting about once a week. There was no true diplopia, but on turning the head suddenly, vision was temporarily blurred. He thought that his visual acuity had failed during the 6 months previous to admission. There was no headache and no suboccipital tenderness.

Neurologic Examination. The pupils were small but equal and reacted promptly to light and accommodation. There was a persistent induced nystagmus on lateral gaze, with the slow component to the left. Conjugate movements of the eyes were restricted on upward gaze, but elevation slightly above the horizontal was possible. The optic fundi showed bilateral papilledema of 3 D. with tortuous and engorged veins. There was slight diminution of hearing in the left ear. The patient showed marked swaying in Romberg’s position and the gait was staggering, with a tendency to fall to the right. He had slight incoordination bilaterally in performing the finger-to-nose test. The left corneal and palatal reflexes were decreased. General physical examination was negative and the remainder of the neurologic examination disclosed no additional findings.

1st Operation. In order to rule out the possibility of a tumor of the posterior 3rd ventricle which would require a supratentorial exposure, ventriculography was carried out under local anesthesia on April 24, 1936. The ventriculograms showed marked symmetrical dilatation of the lateral and 3rd ventricles, and, therefore, a posterior fossa exploration was immediately undertaken. The cisterna magna was large and the arachnoid was thickened and gray. The left cerebellar hemisphere was slightly larger than the right and the convolutions were slightly widened. Upon opening the cisterna, a large tumor was disclosed when the left tonsil was retracted. The tumor was of yellowish color and found to be extramedullary in location. It had compressed the medulla and upper cord to a marked degree, with displacement of these structures to the right. It was seen to extend far up under the left cerebellar hemisphere and downward to the floor of the fossa. The tumor was carefully mobilized and the fairly numerous entering vessels occluded as they were encountered. The 9th, 10th and 11th nerves were visualized and the 10th and 11th were carefully brushed aside. The 9th, however, was intimately adherent to the tumor and was sacrificed. The tumor was gradually freed from the surrounding structures and apparently completely removed with only moderate bleeding which was readily controlled.

Except for transitory respiratory difficulties at one or two stages during the operation, the patient withstood the entire procedure quite well.

Course. He made an essentially uncomplicated operative recovery and was discharged from the hospital 20 days following operation. At that time the papilledema was regressing and the disk margins were appearing. The nystagmus persisted and there was slight generalized unsteadiness but no definite paresis. He had likewise considerable hoarseness. In November 1936, the patient reported that he was working regularly and his only complaint was that his
voice was husky. He was heard from at intervals after that and seemed to be getting along perfectly well. In 1940 the pathologic material was reviewed, but the diagnosis was not changed from adenocarcinoma of choroid plexus origin.

2nd Admission. The patient returned to the New England Deaconess Hospital on Jan. 3, 1942. He had remained well until July 1941, at which time he began to have attacks of vertigo and a sensation of falling to the left. He complained also of diminution of hearing in the left ear, episodes of “inflammation” of the left eye and occasional numbness of the left side of the face. Neurologic examination showed that his voice was hoarse, and there was marked hearing loss on the left. He had a partial peripheral facial weakness on the left, with hypesthesia of the left face. The optic disks showed slight secondary atrophy, but no elevation. The gait was essentially normal with only slight unsteadiness in turning. There was a positive Romberg’s sign and slight ataxia in performing the finger-to-nose test.

The patient had previously received a course of radiation therapy with a total dose of 1800 r over two portals, but showed no appreciable improvement. Roentgenograms at this time disclosed erosion of the left petrous ridge medially. He developed a left hemihypesthesia and complained of numbness of the entire left side of his body.

2nd Operation. On Jan. 5, 1942, the old incision was reopened and no tumor found on the surface in the posterior fossa. On retracting the left cerebellar hemisphere from the cerebellum-pontine angle, many adhesions between dura and cerebellum were encountered, with pocketing of cerebrospinal fluid about the lower nerve group. Further retraction of the hemisphere revealed a cauliflower-like nodule of recurrent tumor about the size of the end of one’s little finger lying alongside the pons. By careful dissection and suction, the tumor was mobilized along the pons and dilated 4th ventricle opened. The posterior inferior cerebellar artery was preserved, but a large branch leading to the tumor was clipped. With mobilization accomplished medially, the lateral aspect of the growth was attacked and found to involve the 7th and 8th nerves. The former was sacrificed in an attempt to remove the tumor radically, but the 5th nerve was not involved. Another tiny implantation just below the major portion of the recurrent growth was removed and a complete gross extirpation apparently thus achieved, although it could not be determined definitely whether the tumor invaded the pons.

Course. The patient withstood the operation satisfactorily and made an uneventful recovery save for transient cystitis of the urinary bladder. There was marked hoarseness following operation in addition to a complete facial paralysis. He was discharged from the hospital 20 days following operation.

3rd Admission. April 20, 1946. Following the 2nd operation, the patient was relieved of symptoms of nausea and vomiting but had moderate unsteadiness of gait and a left facial paralysis. He was able to carry on with his work, however, and got along reasonably well until March 1946, when he began to have a recurrence of nausea and vomiting which grew progressively more severe so that he returned to the hospital for further surgery. His gait had become somewhat more unsteady during the 6 months preceding this admission and there was weight loss of 15 to 20 pounds, but no headache.

Examination showed no bulging of the suboccipital defect and the optic fundi were essentially normal. There was fine nystagmus to both right and left and slight hypotonia of the left arm. The left arm and leg showed definite ataxia with moderate dysmetria and asynergia. There was a slight left trigeminal hypesthesia and a complete left facial paralysis. The remainder of the cranial nerves were apparently intact. There was slight weakness of the left arm, but there were no other motor or sensory abnormalities. On April 24, reexploration was carried out.

3rd Operation. The left portion of the old cerebellar incision was reopened and a recurrent tumor was disclosed lying along the side of the greatly indented pons underneath the left posterior inferior cerebellar artery. The growth appeared to be well encapsulated and was quite tough in consistency. It extended forward to involve the greater portion of the dilated 4th ventricle. By gently retracting the tumor, it could be dissected away from the overlying attenuated cerebellum which was removed in the process. It was carefully dissected away from the roof of the 4th ventricle and from the pons beneath. The growth was approximately
4 cm. by 3 cm., and again a grossly total extirpation seemed to have been accomplished. Gel foam was utilized to cover the raw surfaces. In spite of the adherence of the growth to the floor of the 4th ventricle and the resultant operative attack, the patient withstood the operation without difficulty.

Course. Following this 3rd procedure, he again made an uneventful postoperative recovery and was discharged from the hospital on May 8, 14 days following operation. Two months later he reported that he was progressing satisfactorily, although bothered considerably by unsteadiness in walking. He wrote on Nov. 12, 1946, that he was getting along fairly well, being up and around the house, but complained of lack of balance as well as general weakness and flabby muscles. He also complained of marked fatigue.

PATHOLOGIC DESCRIPTION OF TISSUE REMOVED AT THE THREE OPERATIONS

Microscopically, the three specimens were essentially identical. There were numerous papillary stalks of loose, scanty, fibrous tissue in which vascular channels were prominent; about the periphery of the connective tissue there were tall, columnar cells arranged in a single or pseudostratified layer (Fig. 1). The latter cells had a slightly granular cytoplasm and a nucleus which contained moderate chromatin and which was elongated in the long axis of the cell, usually lying in the basilar half of the cytoplasm. Scattered throughout, but especially in the connective tissue beneath the columnar cells were occasional irregular, sometimes laminated and calcified, masses.

In some regions the papillary growths seemed to be lying in a cavity lined with a single layer of columnar cells, giving the appearance of a papillary adenocystoma, but this type of growth was not consistently present.
There were a few foci in which the cells had lost their papillary arrangement and single layering to grow in rather dense masses. Here, the cells showed more rounded and vesicular nuclei, with considerable variation in nuclear size and shape. Mitoses were rare and could be found only after prolonged search. While the stroma in general was very scanty, there were frequently regions where it appeared to be infiltrated by the more dense cell masses. No cells other than blood cells were present in either blood vessels or lymphatics.

SUMMARY

Papillomas of the choroid plexus showing malignant (adenocarcinomatous) characteristics are rare intracranial. Relatively few of these growths have been removed surgically, and fewer patients still have had a long, useful survival period. The present report is that of an adult male who had a tumor of this type. He has remained in relatively good general condition for over ten years since the initial removal of his malignant papilloma.

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REFERENCES


OSTEOPLASTIC FLAP METHOD IN THE TREATMENT OF SUBDURAL ABSCESS

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The chief points in the treatment of subdural abscess are: (1) removal of pus, and (2) sterilization of the subdural space.

The first objective—removal of pus—cannot usually be achieved by attempted drainage through one or more perforator openings in the skull or through a cranietomy. This is because the pus becomes walled off in separate compartments and free flow to the drainage tube is impossible. Insertion of drainage tubes far into the subdural space through perforator openings in the skull is dangerous because of the possibility of injury to the cerebral cortex with production of intracerebral abscess. Removal of pus through a cranietomy has two disadvantages: (1) exposure is insufficient if the abscess is large, and (2) an undesirable cranial defect is left if the patient survives.

Sterilization of the subdural space can be attained only by removal of infectious material and direct instillation of antibiotics. It is doubtful that sulfonamides, orally or intravenously, or penicillin, intramuscularly or intrathecally, reach the space in adequate concentration through the membrane that forms about the abscess. Penicillin instilled through a drainage