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, papilloma of choroid plexus origin was first described by Guerard in 1832, and Van Wagenen 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, but there seems to be some question regarding the pathologic histology in light of more recent classification. For this reason, the case reported by Sachs 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 Cushing reported in 1925 the first 6 cases of Cushing’s total of 12, and in 1930 Van Wagenen 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 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, 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 35 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 sub-occipital 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 papillledema 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