Primary Choroid Plexus Papilloma of the Cerebellopontine Angle

Case Report

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While it is not uncommon for a choroid plexus papilloma arising in the fourth ventricle to extend into the cerebellopontine angle, primary papilloma in this region is very rare. In a detailed review of the literature, Morello and Migliavaca found only nine such cases and added two cases of their own. Since then, to our knowledge, there has been no further report on primary choroid plexus papilloma in the cerebellopontine angle. This is a report of a primary cerebellopontine angle choroid plexus papilloma which was found at postmortem.

Case Report

This 34-year-old man was first seen in the neurological service on August 27, 1964, for left-sided facial weakness of 6 months' duration. He also complained of left-sided tinnitus, and diminished hearing. At that time, neurological examination revealed a lower motor neurone left facial weakness, left nerve deafness, and a nonfunctioning labyrinth on the left side. The patient refused admission.

Examination. On December 8, 1966, the patient was admitted with difficulty in speech and swallowing. He was drowsy but cooperative, and was oriented in time and space. There was marked dysarthria. The disc margin on the right side was blurred, and the left fundus could not be seen because of severe kerato-conjunctivitis. There was bilateral lateral rectus palsy, left fifth cranial nerve palsy (both sensory and motor), left lower motor neurone facial weakness, left nerve deafness, weakness of palatal movement on the left side, and profuse discharge from the left ear.

Left-sided cerebellar and right-sided and cortico-spinal tract signs were also present. The routine laboratory examination was normal. Lumbar puncture revealed a pressure of 180 mm of water, 130 mg% of protein, and 45 mg% of sugar with no cells. Plain x-ray skull films did not reveal any abnormality. A right brachial angiogram was normal.

Operation. A left suboccipital craniectomy was performed, and a partly cystic fairly vascular granulomatous extradural mass measuring $2.5 \times 1.5$ cm was encountered. The dura appeared to be thickened. Palpa-

![Fig. 1. Photomicrograph shows papillary processes with fibrovascular connective tissue core, covered by a single layer of cuboidal epithelium. Some hemorrhage into the stroma is seen in the center. H. & E., × 60.](image-url)
tion of cerebellum through the dura did not reveal any abnormality. The abnormal tissue was biopsied, but the dura was not opened as it was thought that associated arachnoiditis might have been the cause of the clinical signs. Postoperatively the patient steadily deteriorated and died 1 month after the operation. At microscopic examination, the tissue removed was described as “hyper-trophic arachnoid granulation tissue” (Fig. 1).

Postmortem Examination. A complete postmortem was done. The findings were confined to the central nervous system. On removing the brain the petrous part of the left temporal bone appeared quite prominent and the internal auditory meatus measured 7 mm in diameter. The superior surface of the brain appeared essentially normal. There was a dark irregular soft tumor measuring 4 × 3 × 2 cm in the left cerebellopontine angle extending into the petrous bone (Fig. 2). The brain stem was shifted to the right. Sections through the brain stem revealed a distorted fourth ventricle with no tumor in it. On the lateral aspect of the pons, there was a small area of hemorrhage (Fig. 3).

Microscopic examination of the tumor removed from the petrous bone was similar to the main tumor mass and consistent with a choroid plexus papilloma (Fig. 4). Histopathological examination of sections from midbrain, pons, medulla, and cerebellum did not reveal any abnormality.

Discussion

The cerebellopontine angle syndrome can be produced by a papilloma which is either an intraventricular extension or a primary tumor originating from the tuft of choroid in the lateral recess of the fourth ventricle. It is reasonable to expect early signs of intracranial hypertension in papillomas of intraventricular origin and signs due to involvement of various cranial nerves, vestibular nuclei, and cerebellar peduncles in those of extraventricular origin. The latter variety is rare.

Neither Bohm and Strang,¹ nor Wilkins and Rutledge,⁵ had a case of choroid plexus papilloma originating in the cerebellopontine angle in their series of 25 and 19 cases respectively. The 11 cases reported up to 1964 have been summarized by Morello and Migliavaca.⁶ In the majority of cases hearing loss was the main complaint. X-ray examination of internal auditory meatus was not done in most of them. In our case, lower
motor neuron facial weakness was the main symptom, although a hearing deficit was clearly demonstrated during clinical examination. The involvement of facial and statoacoustic nerves were further confirmed by the presence of tumor in the petrous bone and enlargement of internal auditory meatus seen at autopsy. Features quite similar to this case were also observed at operation by Morello and Migliavaca\(^4\) in one of their cases.

The primary origin of the tumor in the cerebellopontine angle is strongly suggested in our case because of the early cranial nerve involvement, late onset of intracranial hypertension, presence of tumor in the cerebellopontine angle (Fig. 2), and demonstration of a normal fourth ventricle except for distortion. Greene\(^2\) believes that the choroid plexus papillomas of non-ventricular origin possibly arise from the ectopic choroid tissue. In the cerebellopontine angle, these papillomas no doubt arise from the choroid tissue which normally projects outside the ventricle through the foramina of Luschka.

The value of angiography in the preoperative diagnosis of choroid plexus papillomas, particularly those in the lateral ventricle, has been recognized.\(^1\) In our case, although right brachial angiography was done, it did not reveal any abnormality. This may be because of the situation of the tumor on the opposite side and the strict laminar flow of blood in the basilar artery. The presence of a nodular mass of granulation tissue seen extradurally at operation in our case totally misled us to consider the whole pathology to be secondary to an inflammatory process. A retrospective review of the material removed at operation was quite diagnostic of choroid plexus papilloma (Fig. 1). We are not sure whether this represented seeding from the primary tumor or an extradural extension.

Among the 11 cases reported so far only four patients have survived.\(^4\) It is obvious from the review of literature, as well as details of the case presented here, that a choroid plexus papilloma of the cerebellopontine angle cannot be distinguished from the commoner clinical entity of acoustic neurinoma or neurological sequelae secondary to chronic middle ear disease, because the former can produce all the symptoms and signs of either, including bone destruction.

**Summary**

A case of choroid plexus papilloma originating in the cerebellopontine angle has been presented. We have stressed the similarity of signs and symptoms with those of an acoustic neurinoma, and the possibility of diagnostic confusion in countries where the incidence of inflammatory intracranial lesions is high.

**References**