Acinar choroid plexus adenoma

Case report

Richard L. Davis, M.D., and Gerald E. Fox, M.D.

Cajal Laboratory of Neuropathology, Los Angeles County-University of Southern California Medical Center, and Department of Pathology, University of Southern California School of Medicine, Los Angeles, California; Neuropathology Branch, Armed Forces Institute of Pathology, Washington, D.C., and St. John's Hospital, Springfield, Illinois

Mucin-producing cystic tumors of the choroid plexus are rare and most are papillomas or carcinomas. An acinar choroid plexus adenoma of the right lateral ventricle is described. Symptoms and signs included headache, vomiting, papilledema, and a gait disturbance. The neoplasm was located with ventriculography and totally removed, but the patient died of complications of surgery. The relationship of this tumor to others arising in the choroid plexus is discussed.

Key Words: brain tumor · choroid plexus · acinic adenoma · lateral ventricle

Tumors of choroid plexus epithelium are rare lesions. They constituted 0.5% to 0.6% of Zulch's cases and 2% of the gliomas in Russell and Rubinstein's series. All of these cases fall into the group of choroid plexus papilloma and carcinoma. Even rarer are cystic lesions, apparently deriving from choroid plexus epithelium, with a lining of obviously secretory (mucinous) cells.

We have had the opportunity to study a peculiar tumor, apparently unique, which seemingly arose from the choroid plexus of the trigone of the right lateral ventricle. This tumor was composed of acini of what appeared to be secretory epithelial cells; it also contained secretory goblet cells in the adjacent choroid plexus epithelium.

Case Report

The patient was a 22-year-old man whose complaint was severe morning headache for the previous month associated with vomiting. He stated that he had a "pounding feeling" in the ears on both sides and that the headaches were generalized, not localized to one side. He had noted slight blurring of vision, but no double vision. Balance had become poor in the last week.

Examination. No masses or bruit were detected in the cranium. There was acute severe bilateral papilledema with hemorrhages and exudates. The visual fields were full to confrontation testing. No nystagmus was present, and the pupils were normal. Perception of facial sensation was normal. There was a slight left peripheral facial weakness with a delayed blink of the left eyelid. Hearing was normal as were the tympanic membranes. Pharyngeal movements and sensation were normal. The tongue was in the midline. There was no neck stiffness. Examination of the extremities indicated no ataxia on finger-to-nose testing. A definite unsteadiness on walking, particularly on turning, was
present. Muscular strength was normal as was perception of all sensory modalities. The reflexes were normal and equal bilaterally. The Hoffman’s sign was negative and the plantar reflexes normal. It was felt that the patient had a rapidly growing posterior fossa tumor with marked increase in intracranial pressure. Ventriculography showed a filling defect in the posterior portion of the right lateral ventricle with displacement of the posterior portion of the third ventricle.

Operation. The patient was operated upon on the fourth hospital day. At craniotomy an encapsulated tumor was found firmly attached to the wall of the ventricle. The tumor was removed intact.

Histological Examination. The gross surgical specimen measured $7 \times 4 \times 3$ cm and appeared to be well encapsulated with a nodular external surface. On cut section it was pink-gray in color, with a soft gelatinous colloid consistency, and had numerous small cystic areas $1$ mm in diameter. Microscopically, the tumor was composed of lobes of polygonal to cuboidal to columnar cells forming small acini. The macrocysts contained degenerating cells and mucinous material as did some of the acini. The cytoplasm of the tumor cells had fine eosinophilic granules; in the more compact areas the granules were more numerous and the cytoplasm more eosinophilic. The nuclei were round to oval and vesicular. No mitotic figures were present, no necrosis was seen, and there was no vascular proliferation (Fig. 1).

In the adjacent choroid plexus there was a transition from normal choroid plexus epithelium to larger columnar cells with a finely granular, slightly eosinophilic cytoplasmic “globe.” These cells appeared identical to those forming the tumor proper (Fig. 2).

Special stains showed the cytoplasm of the tumor cells and the material in the acini and microcysts to be mucicarminephilic, PAS-positive and diastase-resistant, and AMP-positive and hyaluronidase-resistant. There was a fine pink granularity in these same areas with the PTAH stain, and a pale blue-gray appearance with a Masson trichrome stain. Reticulin fibers were present only around vessels and in the interlobar collagenous septae.

Postoperative Course. During the postoperative period two additional craniotomies

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Fig. 1. Left: Low-power photomicrograph showing tumor and adjacent choroid plexus. H. & E., $\times 7.5$. (AFIP Neg. 62-3410). Right: Photomicrograph showing tumor with prominent acinar structures and secretory cells. H. & E., $\times 260$. (AFIP Neg. 62-3412).
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were performed in an attempt to control postoperative bleeding, but in spite of these, the patient died on the third postoperative day. The autopsy, aside from evidence of intracerebral hemorrhage with cerebral edema and tonsilar herniation, showed no significant changes. No tumor, either gross or microscopic, was found.

Discussion

This lesion is a most unusual tumor of the choroid plexus. Professor Dorothy S. Russell and Dr. Lucien J. Rubinstein were kind enough to review sections of the tumor and could not recall seeing a similar lesion.\(^{8,4}\) Dr. Rubinstein, in fact, suggested the name “acinar choroid plexus adenoma.” A search of the literature and review of the AFIP files indicate that this lesion is unique.

The resemblance of the tumor to salivary gland was immediately apparent, but there was no evidence of a salivary gland tumor in this young man, either clinically or at autopsy, and by all criteria, the tumor was not histologically malignant. It is interesting that special stains of human mixed salivary gland showed the mucous portions to have the same staining characteristics as those of the tumor.

Although it is not absolutely established, it would seem that this tumor arose from choroid plexus epithelium. Very rarely one sees what appears to be a “goblet” cell in normal choroid plexus or ependyma. Their rarity has, however, thus far frustrated our efforts at performing special stains. The presence in this case of cells identical to those of the tumor in the adjacent choroid plexus certainly suggests origin from the latter structure.

The case reported by Hoenig, et al.,\(^1\) in which a multiloculated cystic tumor apparently arose from the choroid plexus of the fourth ventricle, also supports the hypothesis that neoplastic mucin-secreting cells may originate in choroid plexus epithelium. In their report the cysts contained “clear mucoid material” and were lined by columnar cells with vacuolated cytoplasm. The cytoplasmic vacuoles and cyst material were PAS-positive and mucicarmineophilic. They also noted continuity between normal choroid plexus epithelium and tall columnar cells identical to those lining the cysts. Tham, et al.,\(^7\) have reported a choroid plexus carcinoma of the fourth ventricle that showed transitions from normal choroid plexus epithelium to tall columnar mucous containing cells.

The relationship of this tumor to papillomas of the choroid plexus, colloid cysts, and choroid plexus cysts can only be speculated upon. The reader is referred to the excellent studies by Kappers\(^2\) and Shuangshoti, et al.,\(^6\) on the embryogenesis and development of the latter two lesions.

Summary

An acinar adenoma was found in the lateral ventricle of a young adult male. This tumor, a type not previously reported, apparently arose from choroid plexus epithelium. Although the lesion is histologically benign, the patient died of complications of surgery, and, therefore, the natural course of the tumor remains unknown.

References

1. Hoenig, E. M., Ghatak, N. R., Hirano, A., and Zimmerman, H. M. Multiloculated cystic tumor of the choroid plexus of the fourth ven-

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*Fig. 2. Photomicrograph illustrating adjacent choroid plexus with transition to goblet cells identical to those in the tumor. H. & E., \(\times 260\) (AFIP Neg. 62-3411).*


4. RUSSELL, D. S. Personal communication, April 7, 1964.


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Address reprint requests to: Armed Forces Institute of Pathology, Washington, D.C. 20305.