Intraventricular craniopharyngioma

Report of two cases

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Craniopharyngiomas are usually considered to arise along the pituitary stalk and upper aspect of the pituitary gland. Two cases of craniopharyngioma confined to the third ventricle are reported.

KEY WORDS  craniopharyngioma  third ventricle  brain tumor

In 1953, Dobos, et al., reported an intrinsic tumor of the third ventricle that was probably a craniopharyngioma. He was unable to find any cases reported in the literature of other tumors like this, confined to the ventricles. Since then there have been no other cases reported except ours. We are reporting two cases of craniopharyngioma confined to the ventricular system.

Case Reports

Case 1

This 46-year-old man suffered headaches and malaise for 16 months before death. Three months after the onset, examination showed bilateral papilledema but no other neurological change. Lumbar cerebrospinal fluid (CSF) pressure then was 360 mm with 130 lymphocytes per cm, glucose 24 mg%, and protein 74 mg%. In the next month repeated CSF analyses revealed elevation in pressure, white cell count, and protein. During the next 12 months the patient became steadily worse; the papilledema became more marked, but cerebral arteriography and ventriculography were normal. He died following a series of grand mal seizures, with evidence of increased intracranial pressure.

Postmortem Examination. The brain weighed 1400 gm. There had been herniation of the cortex through the occipital burr holes, and the convolutions were flattened. The floor of the third ventricle showed a prominent bulge. There were adhesions where a subtemporal decompression had been performed.

FIG. 1. Case 1. Section of brain showing tumor in third ventricle with intact floor of ventricle.
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been made. The temporal cortex here showed cystic encephalomalacia.

When the brain was sectioned the third ventricle was found to be greatly dilated and filled with a firm granular tumor 4.5 cm long and 3.5 cm in diameter (Fig. 1). The tumor was pedunculated and attached to the floor of the third ventricle; it partially blocked both foramina of Munro and reached nearly to the aqueduct of Sylvius. Microscopic examination of the tumor revealed sheets of squamous cells with keratin and pearl formation, and intercellular bridges. The tumor was supported by an edematous cellular connective tissue infiltrated by round cells (Fig. 2).

The autopsy also revealed bilateral lobular pneumonia, several small pulmonary emboli with early infarctions, and portal cirrhosis.

Case 2

This 26-year-old man developed headache, lethargy, and episodes of syncope 4 years before death. Papilledema was present 6 months after onset, and a third ventricle craniopharyngioma was biopsied at this time. With steroids, x-ray treatment, and ventriculojugular shunting he was maintained for more than 3½ years. He died because of increased intracranial pressure not controlled by a ventriculojugular shunt.

Postmortem Examination. There was a bone defect in the right frontal area. Beneath this was a large cystic space filled with clear fluid where the frontal lobe had been partially removed. The cerebral defect communicated with the ventricular system. The ventriculojugular shunt was intact. There was marked flattening of the cerebral convolutions and early necrosis of the cortex in the distribution of the left posterior cerebral artery. The optic chiasm was pushed forward,

Fig. 2. Case 1. Histological architecture of tumor showing squamous appearance with keratin and peripheral palisading of cells. H & E, ×80.

Fig. 3. Case 2. View of base of brain showing intact floor of third ventricle.
and the tumor actually indented the sella. The floor of the third ventricle was intact (Fig. 3).

When the brain was sectioned a mass was found replacing the third ventricle and measuring 7.0 cm in length and 4.0 cm in diameter. The tumor was sharply circumscribed, firm, and gritty (Fig. 4). Centrally, the tumor had undergone necrosis and was partly cystic. Small calcifications were present. The left occipital lobe and anterior superior pons revealed extensive necrosis. Microscopically, the tumor was composed of sheets of squamous cells showing cystic degenerative changes and peripheral palisading of cells, characteristic of the so-called "adamantoid" pattern of craniopharyngioma (Fig. 5). Keratin production was common. Focal areas revealed calcification and early bone formation. The tumor was very tightly adherent to the ventricular wall and incorporated glial tissue in areas.

The autopsy also revealed bilateral lobular pneumonia.

Discussion

Developmental tumors in and around the sella have usually been divided into dermoids, epidermoids, and craniopharyngiomas. Russell, et al., state that this may not be entirely logical. The same reasoning would apply to tumors arising in the third ventricle, such as those reported here and by Dobos, et al. The colloid cyst of the third ventricle is no doubt a separate entity. 

Mosberg and Blackwood in a review of the collected cases of colloid cysts found reports of several cysts which were lined partly or wholly by squamous epithelium which produced keratin. This strongly suggests that some of these cases were epidermoid cysts and not colloid cysts. We see nothing to be gained by making a separate classification of epidermoids, solid or cystic, which arise in the third ventricle from those arising from Rathke's pouch remnants or the parapituitary, suprasellar, or other epidermoids. We do not believe, as did Dobos, et al., that there are any significant histological features which allow one to tell the intraventricular epidermoid from those in other locations. 

Craniopharyngiomas arise usually from epithelial cell rests along the pituitary stalk and upper aspect of the pituitary gland. Svien in listing seven major locations for 20 craniopharyngiomas did not include the ven-
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ticle. Matson and Crigler in a report of 74
cases stated that all of these tumors arose in
the region of the pituitary stalk. No instance
of a craniopharyngioma entirely within the
ventricle has been described.

Summary

We have reported two cases with the rare
occurrence of a craniopharyngioma entirely
within the third ventricle.

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