EPIDERMOID TUMOR (CHOLESTEATOMA) OF THE LATERAL CEREBRAL VENTRICLE

CASE REPORT

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Epidermoid tumors (cholesteatomas) are uncommon intracranial lesions and Cushing reported only 15 examples (including dermoid tumors) in his series of 2,023 verified brain tumors. He did not describe the location of each tumor.

Grant and Austin reviewed the literature in 1950 and reported their series of 22 epidermoids operated upon over a period of 20 years. Fifteen were intradural and were located as follows: 4 in the cerebellopontine angle; 4 near the pituitary; 2 in the third ventricle; 2 in the cerebral hemispheres; 2 in the cerebellar hemispheres; and 1 in the vermis. They cited Mahoney, who in 1936 collected 142 cases from the literature. Of these tumors, 112 were intradural and were located as follows: cerebellopontine angle, 53; parapituitary, 44; and fourth ventricle, 15.

Epidermoids comprise approximately 1/4 to 1 per cent of all intracranial tumors. Cruveilhier's classic description is well known. His term "tumeur perlée" describes the glistening mother-of-pearl appearance of the outer membrane.

Most authorities agree that the tumor arises from embryonic inclusions of the epidermal portion of the epiblast. They may occur anywhere along the base of the brain from the chiasm to the posterior cistern or may involve the cerebellopontine angle. The extradural lesions may appear in the cranial bones, under the temporal lobes or within the middle ear. The interior of the tumor is entirely avascular and resembles "cottage cheese" in color and consistency.

Bailey described the microscopic structure as follows:

"On the outside is a thin fibrous layer, within which are flattened epithelial cells which sometimes contain granules of keratohyaline. . . . The central portion is composed of what one might call the ghosts of epithelial cells; their internal structures have disappeared leaving only the shells. This portion resembles the appearance of a dead plant. Besides the skeletons and debris of dead cells one finds some fat and usually crystals of cholesterol."

CASE REPORT

Miss V.D., aged 37, was admitted to Temple University Hospital on Sept. 13, 1955, complaining of progressive loss of vision in the right eye for 3 years. One year before onset of visual impairment, the patient had had "terrific" right supraorbital headaches for 4 months, and none thereafter. Twenty years before admission, she had had generalized convulsions preceded by an epigastric aura associated with a "far away feeling." At that time, an air study done at another hospital was negative. The patient had taken Dilantin gr. 1 1/2 a day for many years. She had had no convulsions for the last 7 years.

Examination. The pupils were equal and reacted well to light. The external ocular movements were normal. The visual fields showed a superior temporal quadrant anopsia on the left with 6/6 vision and a small isle of temporal vision on the right with almost complete blindness in this eye. Fundi showed a pallor of the left optic nerve and atrophy of the right optic nerve. The cranial nerves were otherwise negative. The deep tendon reflexes were hy-
peractive. There was no Hoffmann or Babinski sign. There was no impairment of sensation, muscle power or coordination.

The laboratory findings, which included complete blood count, sedimentation rate, urinalysis and blood Wassermann, were normal. The electroencephalogram suggested impairment of the left hemisphere, possibly caused by a deep-seated lesion. Roentgenogram of the skull was negative. The spinal fluid contained 1 cell and 50 mg. of protein per 100 cc.; Wassermann and colloidal gold curve tests were negative.

An encephalogram disclosed an irregular cauliflower-like tumor, measuring approximately $7 \times 4.5 \times 4$ cm., occupying the right temporal lobe and ventral horn and actually visualized by the surrounding air (Fig. 1).

**Operation.** A right temporo-occipital craniotomy was done on Sept. 19, 1955. The dura mater was opened to permit selective herniation of the temporal lobe. A cannula was inserted into the anterior portion of the lobe and it encountered air and slightly turbid ventricular fluid. A horizontal incision was made through the middle temporal gyrus, and an epidermoid tumor was exposed. The white crab meat-like tissue filled the entire right temporal horn up to the body of the lateral ventricle. It was completely removed from the ependymal lining, but a capsule could not be found (Fig. 2). However, when the major portion was removed, a core of tumor was found in the tip of the temporal lobe extending mesialward across the midline of the cranium apparently into the chiasmal area and outside of the 3rd ventricle. This tissue was also removed and a capsule could be seen in the depth of this small finger-like extension. It was thought unwise to strip this capsule because if hemorrhage occurred, it could not be reached through the lateral exposure. However, a small portion of it was removed for biopsy and a silver clip was placed in the area for future identifi-
Fig. 3. Note silver clip in parapituitary area. This was probably site of origin of tumor before it broke into tip of temporal horn.

Fig. 4. Photomicrograph of epidermoid tumor. (Hematoxylin and eosin)
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There was no extension of the tumor to the foramen of Monro. The dura mater was closed and the bone flap was replaced.

Pathologic Report. Sections of the cyst wall show a lining of squamous epithelium with keratinous proliferation. Sections of the cyst content show keratinous debris. Diagnosis: Epidermoid inclusion cyst from right lateral ventricle of brain—cholesteatoma (Fig. 4).

Course. The patient made an uneventful recovery. Recent examination showed no change in her visual condition, but she is comfortable and has no complaints.

COMMENT

The clinical course shows clearly that this lesion arose in the prechiasmal parapituitary area on the right, outside of the ventricle, and produced progressive loss of vision in the right eye. The sudden relief of the terrific headaches which were present for 4 months was the result of rupture of the tumor into the tip of the right temporal horn, thus giving the lesion adequate space to expand. The grand mal attacks preceded by an epigastric aura and a "far away feeling" were probably uncinate in nature and caused by involvement of the mesial portion of the temporal lobe.

The classical appearance of the mass in the lateral ventricle as seen in the air studies has been reported as diagnostic of an epidermoid tumor by Hauser and Elkins,4 who cited similar observations by Krieg, Dyke and Davidoff, Weinberger, Peyton and Baker, and Childe and Young.

The air patterns have been described as irregular streaks, filigree lacework of communicating channels of gas, reticular mass of air, and sponge-like irregular collections of air. Weinberger and also Dyke believed this appearance is explained by the distribution of gas in the folds of the walls of the tumor and that no other known brain lesion produced a similar picture.4 Lindgren6 also stated that the epidermoid cholesteatoma is the only space-occupying lesion that gives a typical pneumoencephalographic appearance.

REFERENCES