PEARLY TUMOR (EPIDERMOID CHOLESTEATOMA) OF THE BRAIN
CLINICOPATHOLOGIC STUDY OF TWO CASES

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(Received for publication May 8, 1958)

EPIDERMOID tumors comprise less than 1 per cent of intracranial growths yet they have characteristic clinical and pathological features. Among the latter is the gross appearance of the tumor which is such that a correct diagnosis may be made on sight. The smooth, glistening, pure white mother-of-pearl surface led Walter Dandy to refer to these as "the most beautiful tumors in the body," and the term "pearly tumor" identifies the lesion perfectly to one who has seen it in situ glistening beneath an arachnoidal lake. Intracranial epidermoids grow very slowly. They are believed to arise from fetal epiblastic cell rests and rarely become symptomatic during childhood. In a survey of 313 verified intracranial tumors in children, Ingraham and Matson did not find one case. They may attain considerable size before producing symptoms, particularly if their growth does not directly obstruct the cerebrospinal fluid pathways. Little reaction is provoked in adjacent brain tissue. Love and Kernohan found that "the only change in surrounding brain as the result of the tumor is one of local atrophy, attributable to pressure exerted by the expanding cyst." This type of brain tumor seems to be capable of growing to almost greater size than any other before producing symptoms. The case from the Hôtel Dieu, reported in Cruveilhier's pathology text in 1829 and for which he coined the term "tumeur perlée," was an incidental autopsy finding in a man who died of a blow on the head and was not suspected of having a brain tumor. Bailey, in commenting on a case of Thurman's, who described an epidermoid of the fourth ventricle in a 50-year-old mentally deranged woman, stated: "The most remarkable feature in this case is the advanced age this woman reached with what was undoubtedly a congenital tumor without apparently any severe pressure symptoms intervening. . . ." Horrax, in reporting the experience of the Brigham Clinic in the management of intracranial dermoid and epidermoid tumors, said of his first case: "So extremely insignificant was the evidence of organic intracranial lesion in this patient that an exploratory operation was undertaken with great misgivings." A large hour-glass, lobular epidermoid in the left temporal lobe was found which required two operations to remove. Holle, in a paper whose title translated from the German is A clinically unsuspected brain cholesteatoma
the size of a man’s fist, described a huge frontal tumor in a soldier who died on the day of his first symptom, and whose findings on neurological examination were normal. Rand and Reeves\textsuperscript{16} and Peyton and Baker\textsuperscript{15} have treated similar large intracranial epidermoid tumors without pressure symptoms.

Many authors have commented on the unusually high incidence of personality change and frank psychosis among patients harboring this type of brain tumor.\textsuperscript{14,17} Mahoney\textsuperscript{14} collected 112 cases of intracranial epidermoid tumor from the literature; 57 per cent of the patients were psychotic. Bailey attributed the mental change to slowly developing internal hydrocephalus when these tumors develop in the posterior fossa. Epidermoids of the brain are found most often in the subarachnoid cisterns somewhere near the midline of the base of the brain or in the region of the fourth ventricle. In Mahoney’s series, 53 were in the cerebellopontine angle, 15 in the fourth ventricle and 44 were parapituitary.

Pearly tumors are known to veterinarians because of the frequency with which they occur in the lateral ventricle of the horse. Only 8 intraventricular epidermoid tumors in man have been reported in the literature\textsuperscript{6,19} but Dyke and Davidoff,\textsuperscript{4} in 1937, and Weinberger,\textsuperscript{20} in 1938, reported a characteristic and pathognomonic encephalographic picture. Because of insinuation of the gas into the convolutions of the tumor, an irregular streaking and filigree lacework of air is seen in the tumor mass forming a sponge-like communicating collection of gas. This distribution of gas in the substance of the tumor is unique, and Lindgren\textsuperscript{12} stated that the epidermoid cholesteatoma is the only space-occupying brain lesion that gives a typical encephalographic picture.

Two cases are presented which illustrate some of these clinical features.

**CASE REPORTS**

**Case 1.** E. K., a 58-year-old Latvian woman, mother of a hospital surgical nurse, had ataxia, headache and visual disturbance 20 years prior to her present symptoms. A ventriculogram had been done in Riga, Latvia and the family was told that there was a deeply situated, inoperable brain tumor. Her symptoms, however, gradually subsided and, except for visual impairment and occasional headache, she raised two children and survived the German and Russian occupation of her country. A hysterectomy was done in 1948 and a subtotal gastric resection for a benign ulcer in 1953. No noteworthy neurological defect had been found during either of these hospitalizations and anesthesia was well tolerated. When seen on June 5, 1956, she complained of weakness and unsteady gait of 3 weeks’ duration and suboccipital headache, worse during movement.

**Neurological Examination.** The patient was an apathetic woman, with broad-based ataxic gait and a positive Romberg sign. There was bilateral secondary optic atrophy and the peripheral visual fields were full. Vertical and horizontal nystagmus was present, the neck was supple and the stretch reflexes were increased everywhere without pathological reflex change.

Roentgenograms of the skull showed parasagittal burr holes, a large midline pineal calcification and erosion of the posterior clinoids. Lumbar puncture revealed a cerebrospinal fluid pressure of 120; protein was 39 mg. per cent. A pneumoen-
cephalogram was attempted, but there was no passage of air beyond the cisterna magna. Ventriculography showed a dilated ventricular system containing 120 cc. of fluid (Figs. 1 and 2). Good filling of an enlarged 4th ventricle showed it to be displaced markedly towards the clivus without any deformity of the roof. The distance from dorsum sellae to floor of 4th ventricle measured 2.5 cm. (normal range, 3.1–4.3 cm.), suggesting a lesion displacing the entire 4th ventricle, pons and medulla downwards into the pontine cistern.

Operation. A suboccipital craniectomy was performed and, upon opening the dura mater over the cerebellum and cistern, a shimmering pearly mass was visible immediately in a deep subarachnoidal lake extending up from the cistern. The mass filled the superior portion of the cisterna magna and covered the posterior part of the 4th ventricle, inferior vermis and a portion of the lateral cerebellar hemispheres.

![Figs. 1 and 2. Case 1. Ventriculogram showing obstructive hydrocephalus and ventral displacement of 4th ventricle.](image)

The capsule was avascular and nodular, and flaked and peeled when manipulated. The central contents of the tumor had the consistency of dry cottage cheese and were easily removed with brain spoon and pituitary rongeur. The capsule of the ventral surface of the tumor was intimately adherent to the pia mater over the cerebellar hemispheres and could be removed only partially. Dissection was carried into a large, smooth-walled 4th ventricle. All tumor tissue, except a portion of capsule adherent to the pia mater, was removed.

Course. The patient has remained well for 19 months. There is no headache and the gait is improved. Vision is unchanged.

Histological Report. The capsular wall of the tumor is composed of thick bundles of acellular fibers (Bailey’s stratum durum), then a layer of stratified epithelium (stratum granulosum) containing deeply staining keratohyaline granules characteristic of epidermis (Fig. 3). There are no epidermoid derivatives such as glands or hair. The central portion of the tumor is composed of desquamated epithelial cells in various stages of cornification and degeneration. Here may be found masses of structureless, flattened “woody” cells, cholesterol crystals and fatty material. No liquefaction of the central tumor mass is found.
Case 2. D.L., a 30-year-old woman, had been described by her family as neglectful and paranoid. Married to a migrant farm hand, she had four children. There was a history of enuresis until age 10 and occasional petit and grand mal seizures throughout childhood. She had been seen at the clinics of the University of Michigan where an I.Q. of 82 had been determined and a diagnosis of compulsion neurosis with mental retardation was made in 1938.

Fig. 3. Case 1. Section of capsular wall demonstrating keratohyaline granules among epithelial cells.

The patient was admitted to the Peoria State Hospital for the first time in 1952 because of seizures. She was then in the 5th month of her 4th pregnancy. The convulsions were attributed to hysteria. The diagnosis of mental deficiency and hysterical seizures was made. The seizures stopped following delivery of a normal child, and she was discharged from the hospital.

She was not seen again until 1955 when seizures and "fainting spells" occurred again during pregnancy. An electroencephalogram at this time showed a grand mal-seizure pattern. She was delivered of a full term, normal boy on Aug. 17, 1955. The day following delivery, she had numerous and prolonged grand mal seizures. Ten days later, she became acutely ill with a high, intermittent fever ranging from 98° to 106°F. over the next 9 days. During this period she was examined by a neurological surgeon who found no evidence of increased intracranial pressure or meningismus. On Sept. 7, 1955, the temperature rose to 108°F. and the patient expired.

Autopsy showed a pulmonary infarction with recent thrombosis of a pulmonary artery and a large epidermoid tumor of the brain replacing most of the centrum
ovale of the frontal and parietal lobes (Fig. 4). The description of the brain is quoted directly from the pathological report by Dr. Dorothy Eshbaugh:

"The cerebral cortex and white matter are not unusual, except for slight edema of the white matter, manifested by numerous small spaces in some regions. The meninges at some levels show slight collagenous thickening, but are otherwise not unusual. The tumor has the appearance of an epidermoid cyst. The edge of the tumor mass where it adjoins the brain is composed of a layer of cells resembling cuboidal epithelium, irregularly arranged, and ranging from two to three to slightly more cells in thickness. In most regions they have round or oval vesicular nuclei, and they often appear slightly flattened, the long axis being parallel to the cyst wall. In other regions where there is a thicker cell layer, there is a single layer of somewhat flattened cells nearest to the cyst content resembling the granular layer of the epidermis, containing many dark blue-stained granules in the cytoplasm with the appearance of keratohyaline granules. The tumor mass itself, except for the above-described border, is composed of fibrillar, acellular, eosinophilic material, arranged in strands above and parallel to the epithelial surface, resembling desquamated cornified epithelium. Deeper in the tumor, they sometimes are arranged in circular formations. In the more central portion they are separated from each other by regular, oval spaces, and in some regions there is a small amount of hyaline eosinophilic material between the fibrils, apparently edema fluid containing tiny round vacuoles, possibly of fatty material. Between the cellular borders of the tumor
and the brain tissue is a narrow layer of collagenous fibers containing focal infiltrations of round cells. In some regions the collagenous layer is the width of several cells, and hyalinized. In one region the epithelial-like cells extend downward toward the brain, forming a small structure somewhat like a rete peg, having sharp lower border. There is no evidence of sweat-gland or sebaceous-gland formation. In most regions the border of cells adjacent to the brain is perfectly regular. However, there is no visible basement membrane. One section has the border of the tumor separated from the cortex by a small amount of loose connective tissue. This connective tissue contains a few blood vessels and is covered on the surface by a lining of cells. It has slight infiltrations of small round cells (Fig. 5).

The cause of death was attributed to the postpartum pulmonary infarction. However, invasion of the hypothalamus may have played a role in her terminal state. There was no evidence of increased intracranial pressure or focal neurological signs other than dementia and epilepsy.

**COMMENT**

In Case 1, the congenital epidermoid brain tumor caused acute symptoms at the age of 38, severe enough for ventriculography to be done. Symptoms subsided after the diagnostic procedure and did not occur again for 20 years despite the presence of a slowly expanding mass in the posterior fossa. Although the capsule is recognized as the only proliferating portion of the tumor and should be removed as completely as possible, the extremely slow
growth of the tumor indicates that this may not be important in the older patient, particularly when the capsule is adherent to surrounding neural structures and small pial vessels.

Case 2 again illustrates how large these tumors may grow without causing focal neurologic changes. This demented woman had a history of epilepsy and showed no sign of increased intracranial pressure even in her terminal illness. She appeared no different than the epileptic patient with mental deterioration seen in great numbers in any neurologic clinic.

NOMENCLATURE

Although the term cholesteatoma implies a mass harboring excessive quantities of cholesterol, it is generally used to designate the epidermoid tumors described in our 2 cases. Masses of reactive hyperplastic epidermis may invade the calvarium and meninges following chronic otitis media complicated by perforation of the tympanic membrane and have been designated as reactive cholesteatoma. Dermoid cysts and tumors of Rathke's pouch (craniopharyngiomas) frequently contain quantities of cholesterol but the term cholesteatoma is not used for such lesions. There are a variety of disorders in which cholesterol and its esters are prominent morbid anatomic features. Some authorities have used the term xanthomatosis to designate such states while others use the terms cholesterol lipoidosis, cholesterosis and xanthomatosis interchangeably. Reference has been made to the pearly tumors of the lateral ventricles of the horse which have been known to veterinarians for many years. Some of these may be xanthomas of the choroid plexuses. An excellent study and review of this subject was made by Wolf, Cowen and Graham. In the xanthomas of the choroid plexuses, the stroma and not the overlying choroidal epithelial cells are the elements harboring the cholesterol. These cells break down and extracellular accumulations of cholesterol and its esters develop. Primary tumors of the meninges rarely contain cholesterol in quantities great enough to be significant but one of us described such a meningioma. A review of other cholesterol lipoidoses can be found in a text of neuropathology.

SUMMARY

The occurrence of two epidermoid tumors of the brain is reported, one supratentorial and the other in the posterior fossa. A huge tumor in the frontal and parietal lobes was clinically unsuspected and caused grand mal and petit mal seizures with progressive mental deterioration over a 30-year period.

The posterior fossa tumor caused acute symptoms in the patient's third decade and a period of 20 years then elapsed during which the tumor was quiescent and caused no further neurologic disturbance. At the time of surgery, obstructive hydrocephalus and marked displacement of the hindbrain were found.

Several characteristics of intracranial epidermoid tumors are illustrated:
1. The pathognomonic appearance of the tumors in situ.
2. The slow growth and paucity of localizing focal signs or increase in intracranial pressure.
3. The characteristic pneumoencephalographic picture.
4. The frequency of mental change accompanying this type of tumor.

REFERENCES