PEARLY TUMOR (EPIDERMOID CHOLESTEATOMA) OF THE BRAIN

CLINICOPATHOLOGIC STUDY OF TWO CASES

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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 pneumoenc-