Epithelial (Epidermoid) Tumors of the Cranium
Their Common Nature and Pathogenesis*

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Epithelial (epidermoid) tumors of the cranium comprise a confusing group. Misused words and misnamed lesions abound: the adjectives epidermoid and dermoid have been transformed into nouns, adamantinomas never contain enamel, craniofibryngiomas arise from structures having more oral than pharyngeal potentialities, cholesteatomas are named for a chemical substance rather than cell or tissue of origin, a tumor may be solid and still be called epidermoid cyst, and the dermoid tumors which are simply variants of epidermoids are mistaken for teratomas. Custom seems to have justified certain usages, but sometimes at the expense of clear understanding.

The epithelial tumors under consideration occur in diverse locations such as the ear, scalp, calvarium, jaw and base of the brain. The diagnostic problems and the surgical approach to these regions are so different that they are usually considered as separate entities. The otologist views “cholesteatoma” of the ear as a pseudo-tumor, the result of long-standing infection. The neurosurgeon thinks of “cholesteatoma” at the base of the brain as etiologically distinct from the otic mass, and both are thought to be different from craniofibryngioma. We intend in this paper to show the unitary character of these epithelial tumors of the cranium, based on histological appearance and embryological origin. We contend that they arise by a common process of epithelial misplacement early or late in life, that they range in growth capacity from masses enlarging by accretion of normally formed cellular material to malignant neoplasms growing by cellular proliferation and properly designated as carcinomas, and that modern embryological theory accounts for most of these phenomena and the wide range of biological behavior.

Histology of Epithelium

The epithelia discussed here are derived from the primary germ layer, the ectoderm, and form the epidermis and its appendages such as hair and sebaceous glands, and the ectodermal components of mucous membranes and teeth. These epithelial membranes are attached to connective tissue. Thus skin is composed of an outer epithelial portion, the epidermis, and an inner dense connective tissue derived from mesoderm.

Epidermis is squamous epithelium composed of five layers. The deepest layer is the stratum germinativum (basal cell layer), attached to the connective tissue of the dermis, and formed by columnar epithelial cells normally germinating new cells which are pushed into the layer above. The stratum malpighii (stratum spinosum, prickle cell layer) is several layers thick, having a spiny appearance formed by the intercellular bridges. The stratum granulosum is thinner, the cells are flattened and the cytoplasm contains keratohyalin granules. In this layer, the cells of the epidermis undergo physiological death or necrobiosis. The 4th layer, the stratum lucidum, is a thin homogenous band, seen best in the thick skin of palms and soles, and is not present in thin skin. The outermost layer, the stratum corneum, has horny scales of keratin, the result of cell death by desiccation. Mucosal epithelium

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is similar to epidermis, but lacks a stratum granulosum and is not keratinized, although granules of keratohyalin may be found; hair and glands are absent. The tooth bud or enamel organ is characterized by an outer and inner layer of epithelium enclosing a central stellate reticulum. The inner layer is composed of ameloblasts, capable of forming enamel.

The epidermoid tumors under consideration arise from ectoderm and then differentiate into epithelium resembling epidermis, mucosa, or tooth bud. Epidermis and its derivatives are found in the cholesteatoma of brain and ear, and in epidermoid tumors or cysts of calvarium and jaws. Mucosal epithelium occurs in some cranio-pharyngiomas, and structures resembling tooth buds are seen in “adamantinomas” in the pituitary or the jaw.

Material and Selected Case Reports

Sixty-three specimens from 61 patients were studied. The origin of the tumors is shown in Fig. 1. Forty-one were in the ear, 11 were suprasellar or in 3rd ventricle, 5 were diploic or in the fontanelle, 3 in the pontine angle and 3 at the base of the brain. Three epidermoid cysts of the jaw were also examined.

Case 1. The lacerated scalp of a 28-year-old man was sutured primarily. The patient returned 1 year later complaining of a slowly enlarging mass at the site of suturing. The tumor was excised and was found to be an epidermoid cyst containing keratin (Fig. 2).

Comment. This case illustrates the surgical induction of an epidermoid tumor. The epidermis was approximated in such a way as to allow desquamated stratum corneum to accumulate in a dermal-lined cyst. Epidermoid cysts may thus be induced late in life (see Discussion).

Case 2. A full-term male baby was born spontaneously to a 24-year-old mother. The child lived only a few hours. The cranial vault was absent. The cerebral hemispheres lay at the base of the skull in a mass about 4 cm. in thickness. Epidermis was seen microscopically in the leptomeninges of the occipital region, extending deep into the sulci, and containing abundant linear strands of keratin. The diagnosis was epidermoid tumor in an anencephalic newborn infant.

Comment. This epidermoid tumor is clearly an intrauterine malformation of development, comparable to reported cases of cranio-pharyngioma and adamantinoma present at birth.

Case 3. A 41-year-old woman had progressive loss of vision for 2 years. The left eye was blind and the nerve atrophic; papilledema was not