MEDULLO-EPITHELIOMA*

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MEDULLO-EPITHELIOMATA, rare tumours that arise from the ciliary body, may be divided into the embryonic, adult, and mixed types.

The first type, also termed "diktyoma" (Fuchs, 1908) arises from the non-pigmented epithelium of the ciliary body and tends to reproduce the structure of the embryonic retina. The tumour grows as a cellular many-layered membrane which is folded and fused together to form papillary projections, gland-like structures, and bizarre sheets of cells. Though they are generally composed of non-pigmented epithelium, occasional tumours, like that reported by Klien (1939), are composed predominantly of pigmented epithelium.

The adult type includes a heterogenous collection and the available material is insufficient to warrant a collective description characteristic of the whole group. They usually occur in eyes that have suffered severe ectogenous or endogenous inflammation in which ciliary epithelial hyperplasia has developed or in eyes that have been blind and atrophic since childhood (Reese, 1951). Histologically the tumours are characterized by membranes each composed of a single layer of cells. Sheets of cells with vacuoles or degeneration resulting in the formation of hyaline intercellular material may also be present. The pigment epithelium is much more prominent in the adult forms of the tumour and the presence of melanin in the cells is characteristic.

The mixed type combines the characteristics of the other two.

The embryonic medullo-epitheliomata occur in infants and young children. The average age is 5 years and the oldest patient reported so far was 28 years old (Soudakoff, 1936). The age distribution of the adult forms is wider, from 10 to 63 years.

Embryonic medullo-epitheliomata are extremely rare; not more than twenty authentic cases were assembled by Kesavachar and Junnarkar (1960), who published the only case so far reported from India. The following case is considered worthy of presentation because the patient was so much older than any reported previously.

Case Report

A 35-year-old Muslim male came for examination on October 3, 1959, with a 20-day history of gradual diminution of distant vision in the right eye. There were no other complaints and the eye had previously been normal.

Examination.—A severe glaucoma rapidly developed after the instillation of homatropine as a preliminary to refraction but this was relieved by Diamox. A dark nodule was seen at the base of the iris at the 5 o'clock position and there seemed to be a swelling behind which was pushing the iris forwards.

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Operation.—As the lesion was thought to be a foreign-body granuloma which was producing glaucoma by pressure on the filtration angle, surgical removal was attempted. On cutting through the iris, a large pale papillary tumour was seen arising from the ciliary body and pressing on the lens. Removal of the lens revealed that the tumour was too extensive to be excised completely and the attempt to do so was abandoned.

Result.—The immediate post-operative recovery was uneventful although the tumour could be seen as a white mass through the iris; 2 weeks later the tumour started to grow rapidly and after another week the eye was enucleated. During a 6-month follow-up the patient remained perfectly well. There was no evidence of recurrence and the other eye continued healthy with a visual acuity of 6/6.

Pathological Report.—Gross examination shows the eyeball to measure $24 \times 23 \times 23$ mm. Meridional section shows a pale nodular growth 4×3 mm. in size, arising from the ciliary body and extending behind the cornea. The lens is not present having been removed at the first operation.

Histological examination shows the growth to be composed of non-pigmented columnar cells arranged in a membrane consisting of two or three layers of cells. The membrane is much folded upon itself to form papillary and gland-like structures (Figure).

FIGURE.—Typical papillary and gland-like architecture with a single layer of pigmented epithelial cells at the periphery. Haematoxylin and eosin (low power).

The former have cores of delicate fibrous connective tissue and blood vessels. The latter show a fine limiting membrane and blunt protoplasmic processes proximally and small central clear spaces. Groups of tumour cells are also present in the connective tissue of the ciliary body and the iris. The pigment epithelium does not take part in the neoplastic

process, but there is a layer of these cells at the periphery of the growth and deposits of melanin in the surrounding connective tissue. A dense neutrophilic exudate is seen between the groups of tumour cells. No evidence of invasion of the sclera is detected.

Discussion

The microscopic appearance of this tumour is characteristic of the embryonic type of medullo-epithelioma and shows none of the features of the adult type; the tumour is composed of non-pigmented columnar cells arranged in a multi-layered convoluted membrane, there is no formation of ciliary epithelium, and none of the usual antecedents of the adult medullo-epithelioma are seen, such as previous pathology or a blind atrophic eye. A typical embryonic medullo-epithelioma (diktyoma) without areas suggesting a mixed pattern developed in the previously healthy eye of a 35-year-old man. This is much beyond the age of the oldest case previously reported which was seen in a 28-year-old patient (Soudakoff, 1936). The inflammatory exudate was the only feature that even remotely suggested the appearance of an adult medullo-epithelioma; it was largely neutrophilic and the onset was too acute to have preceded the neoplasm, so that the exudate was evidently caused by the earlier operation.

The fairly benign behaviour of the tumour resembles that generally described (Willis, 1953; Freidenwald, Wilder, Maumenee, Sanders, Keyes, Hogan, Owens, and Owens, 1952); an occasional case, such as that described by Malone (1955), behaves in a much more malignant manner.

The clinical and morphological features lend support to the opinion of Andersen (1948) that the embryonic and adult medullo-epitheliomata are different types of the same variety of neoplasm.

Summary

A typical embryonic medullo-epithelioma (diktyoma) occurred in the previously normal eye of a 35-year-old male.

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