TUMORS OF THE EYE AND ADNEXA

Algernon B. Reese, M. D.

ARMED FORCES INSTITUTE OF PATHOLOGY

ATLAS OF TUMOR PATHOLOGY

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TUMORS OF THE EYE AND ADNEXA

by

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Algernon B. Reese.

TUMORS OF THE EYE AND ADNEXA

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NEUROECTODERMAL TUMORS—Continued

PLATE I

MALIGNANT MELANOMA

A. Malignant melanoma of the limbus. On gross section, the tumor was 3 mm. thick and appeared to invade the cornea slightly at the limbus. Histologic section showed the tumor arising in the limbal region but extending into the conjunctiva, cornea, sclera, and anterior chamber angle. This right eye was from a 23-year-old white man who first noticed a reddish brown spot in the area of the tumor two and one-half years before enucleation. Eight months before enucleation the spot became larger and growth was progressive. (From the files of the Armed Forces Institute of Pathology.) A. F. I. P. Acc. No. 609918.

B. Malignant melanoma of the choroid showing the mushroom shape and the detachment of the overlying retina. This horizontal slice of the right eye was from a 30-year-old white woman who was six months pregnant at the time of enucleation. She had had symptoms of photophobia and loss of vision of the lateral field for three months and "arclike" flashes for several days. (Contributed to the Armed Forces Institute of Pathology by Dr. H. William Gray, Washington, D. C.) A. F. I. P. Acc. No. 627592.

C. Malignant melanoma of the choroid with complete detachment of the retina and extension of the tumor through the posterior emissary adjacent to the optic nerve. The patient was a 54-year-old white man who had been blind for five months before enucleation of the right eye. Although no autopsy was performed, the patient died a year and a half later with clinical evidence of generalized metastases. (Contributed to the Armed Forces Institute of Pathology by Drs. B. H. Neiman and G. H. Woodruff, St. Joseph's Hospital, Joliet, Ill.) A. F. I. P. Acc. No. 506871.

RETINOBLASTOMA

D. Retinoblastoma showing dissemination into the adjacent vitreous. Throughout the tumor mass chalky calcium deposits can be seen. The specimen was obtained from a 20-month-old girl, whose left eye had shown gradual enlargement for several months before it was enucleated. (Contributed to the Armed Forces Institute of Pathology by Dr. W. Clayton Anderson, Winchester, Va.) A. F. I. P. Acc. No. 608318.

TUMORS OF THE EYE AND ADNEXA

EPITHELIAL TUMORS OF THE EYE

EPITHELIAL TUMORS OF THE EYELID, CONJUNCTIVA, AND CORNEA

Basal cell and squamous cell carcinomas of the eyelid do not differ structurally from such tumors seen elsewhere. The basal cell type is by far the more common (85 percent), and the lower lid is much more frequently involved than the upper.

Adenoid epithelial tumors are also encountered, arising from the hair follicle (trichoepithelioma or adenoid cystic epithelioma), from the sweat glands (syringoma), and from the meibomian glands. Benign calcified epitheliomas may occur in the eyebrow, the eyelid, or the orbit (Ashton). The meibomian gland is a modified type of sebaceous gland occurring in the tarsi of the upper and lower lids, and the epithelial tumor arising from it may manifest itself as a rapidly growing carcinoma.

When the conjunctiva is affected by carcinoma, the squamous cell type is at least 10 times more common than the basal cell type. Although its site of predilection is the limbus, with extension to the cornea and the adjacent bulbar conjunctiva, its primary site may also be the bulbar or palpebral conjunctiva. According to some, a papilloma may occasionally be the forerunner of an epithelioma. Leukoplakia, although seen much less commonly on the conjunctiva than in the mucous membrane of the mouth, does occur occasionally.

Intraepithelial epithelioma (Bowen's disease) occurs on the cornea, conjunctiva, and skin of the lids.

EPITHELIAL TUMORS OF UVEAL TRACT

Tumors arising from the ocular tissues which stem embryologically from the optic vesicle are all neurogenous in nature, although all of them do not have their counterparts in the nervous system elsewhere. In figure 1, the optic vesicle is divided into three sections representing the anlagen of the epithelial elements of the choroid, the ciliary body, and the iris. "F" is the anlage of the retina proper, and tumors of this layer are discussed in the section on retinoblastoma. "C" is the anlage of the dilator and sphincter muscles of the iris, and tumors arising in it are discussed in the section on leiomyoma. It is my purpose in this section to discuss the tumors whose parent tissues have their anlagen in "B" and "E" (the pigment and nonpigment layers of the ciliary body), in "D" (the pigment layer of the iris), and in "A" (the pigment epithelium of the retina).

TUMORS OF THE EPITHELIUM OF THE CILIARY BODY

Tumors arising from the ciliary epithelium are among the rarest of ocular neoplasms. Excellent descriptions of them have been furnished by Fuchs and by Nordmann, but even a thorough study of all available cases yields insufficient material to provide criteria for a sharp demarcation of groups.

In this discussion it is entirely logical to consider together the neoplasms arising from the nonpigment and pigment epithelial layers of the ciliary body, since both layers seem always to be implicated in one degree or another. Although normally the one epithelial layer is pigmented and the other is not, even the nonpigment layer, continuous as it is with the pigment epithelium of the iris, is potentially a melanin-producing tissue; the ability to fabricate melanin could not be a constant trait of the one layer and never manifest itself in the other. As a matter of fact, in otherwise normal ciliary epithelium the nonpigment layer may be pigmented (fig. 2) and thus may or may not contain melanin in its cells.

The following types of true neoplasms of the ciliary epithelium occur: (1) benign epithelial tumor (benign epithelioma, adenoma); (2) carcinoma (malignant epithelioma); and (3) medulloepithelioma, consisting of (a) an embryonal type (diktyoma), (b) an adult type, and (c) a mixed type.

Benign Epithelial Tumor of the Ciliary Epithelium

SYNONYMS AND RELATED TERMS: Adenoma; benign epithelioma.

These tumors usually produce no symptoms and are found only accidentally in the microscopic examination of eyes removed for other causes (fig. 3). They are therefore usually seen in the eyes of patients of advanced age. Fuchs believes them to be congenital, while Wadsworth feels they are acquired by an ingrowth of the nonpigment layer. They are usually round to oval in shape, with a maximum diameter of no more than 1 mm. They may be imbedded in a ciliary process, in some instances in the neighborhood of its apex, and will thicken the process and cause it to protrude further into the posterior chamber than the other processes.

Structurally the growth is an invagination of both the nonpigment and pigment epithelial layers into the substance of the ciliary process; this results in an ampulla-shaped cavity, with the hilum or orifice of the ampulla facing the surface of the ciliary process. The inner layer of this ampulla, i. e., the nonpigment epithelial layer, grows into strands and folds of epithelium which are arranged in scalloped formations, particularly around the periphery of the cavity. In cross sections these folds appear as bands or membranes with a double row of cells, or, in pure cross sections, as rosettes. All these membranous outgrowths of the nonpigment epithelial layer that fill the cavity of the invagination form an epithelial network. The interstices of this network, however, instead of being filled by the usual connective tissue characteristic of adeno-

mas, are occupied by a homogeneous eosin-staining material which is no doubt the product of the epithelium, although its precise nature is unknown. With thionine and mucicarmine it does not give the staining reaction characteristic for mucus. The fact that this invaginated space opens on the surface of the ciliary processes has suggested to some observers that the homogeneous substance represents vitreous. This is improbable, however, in view of the absence of a direct connection between the mouth of the ampulla and the vitreous cavity.

The pigment epithelial layer encloses the tumor but does not participate in the tumor growth. The cells may even lose some or all of their pigment, apparently as a result of the pressure exerted on the cells by the expanding tumor. This pressure may cause the cells not only to lose their pigment but even to disappear, no doubt from atrophy, so that dehiscences may be found in the pigmented capsule of the tumor. If the atrophy of this pigmented layer is extensive, the layer may be missing, and the impression may be given that the proliferating nonpigment epithelium has broken through and extended into the tissue of the ciliary body. Such an appearance, however, is probably due, not to an invasive power of these small and benign tumors, but to the disappearance of the encapsulating pigment layer through atrophy. There is never any tendency for connective tissue to encapsulate these tumors. At the site of the mouth of the ampulla-shaped invagination on the surface of the ciliary body, both epithelial layers are missing, of course, and this area is covered by the proliferated tissue. At the site of this hilus the interstices of the epithelial network are in direct connection with the posterior chamber (figs. 5, 6).

If these tumors are located near the base of the iris where the nonpigment epithelium changes into the pigment epithelium of the iris, the nonpigment epithelium composing the tumor may be pigmented.

The homogeneous substance occupying the retiform spaces of the epithelium can constitute an appreciable portion of the tumor so that there appears to be extensive cystic degeneration. Apparently the expanding homogeneous substance can cause atrophy of the imbedded epithelial tissue, leaving only individual nuclei or sparsely scattered nuclei. Such an arrangement resembles that of an actual cyst (fig. 4).

Toward the central part of the tumor the epithelial strands become thinner and have fewer nuclei with less uniform polarity. In the majority of cases vacuoles are found in the epithelial cells, usually in the basal portion. Numerous confluent vacuoles in the base of a cell may separate the rows of nuclei widely and even form cystoid spaces. These spaces are inside the epithelial folds and membranes and are not to be confused with the previously described spaces between the folds filled with the homogeneous substance.

This tumor cannot be considered an adenoma in the usual sense because connective tissue stroma is not a part of its structure. Both this tumor and a true

OPTIC VESICLE

Figure 1.* Diagram of secondary optic vesicle, showing: A, anlage of pigment epithelium of retina; B, anlage of pigment epithelium of ciliary body; C, anlage of dilator and sphincter muscles of iris; D, anlage of pigment epithelium of iris (retinal epithelium of iris); E, anlage of nonpigment epithelium of ciliary body; F, anlage of retina. A. F. I. P. Acc. No. 219935–1.

PIGMENTATION OF CILIARY EPITHELIUM

Figure 2.* Section showing pigmentation of the nonpigment layer of the ciliary body in an otherwise normal-appearing epithelium. A. F. I. P. Acc. No. 219935–2.

BENIGN EPITHELIAL TUMOR OF CILIARY BODY

Figure 3.* Benign epithelial tumor of the ciliary processes. The eye was removed because of malignant melanoma of the choroid. The hilus is seen at "A." A. F. I. P. Acc. No. 219935–3.

Figure 4.* A cystic benign epithelial tumor of the ciliary body. The lesion was not detected clinically. The eye was removed because of malignant melanoma of the conjunctiva. A. F. I. P. Acc. No. 219935–6.

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*From Reese, A. B. Tumors of the Eye. New York: Paul B. Hoeber, Inc., 1951. Figures 31, 32, 33, and 35 are our figures 1, 2, 3, and 4. Figure 1 also appears in less detail as figure 3 in Reese, A. B. Pigmented tumors. Am. J. Ophth., 30: 537–565, 1947. Figure 3 is also figure 8-b in Wadsworth, J. A. C. Epithelial tumors of the ciliary body. Am. J. Ophth., 32: 1487–1501, 1949.

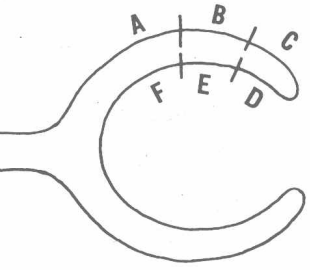

Fig. 1

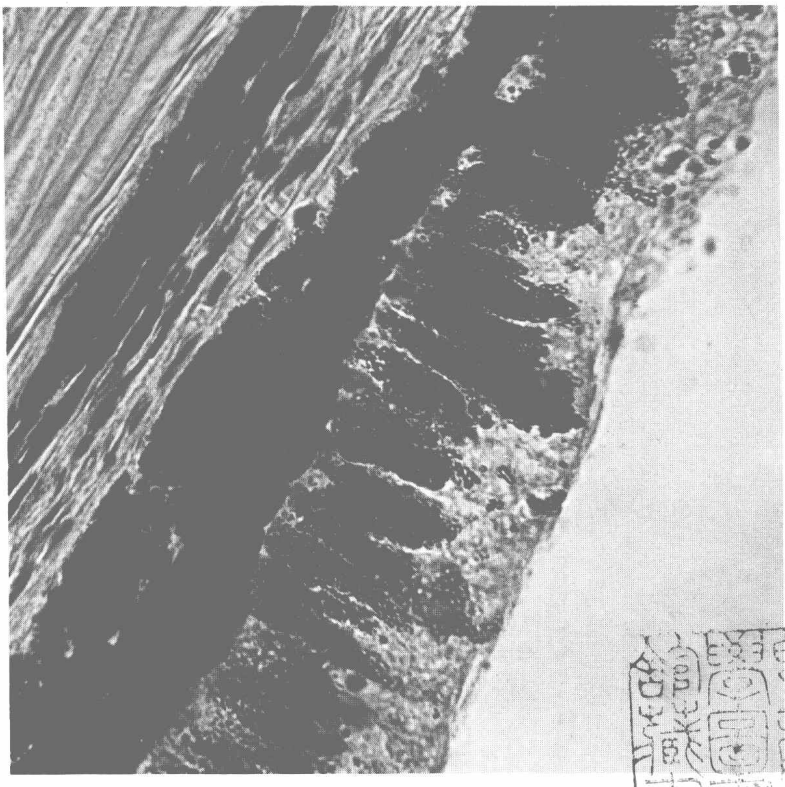

Fig. 2

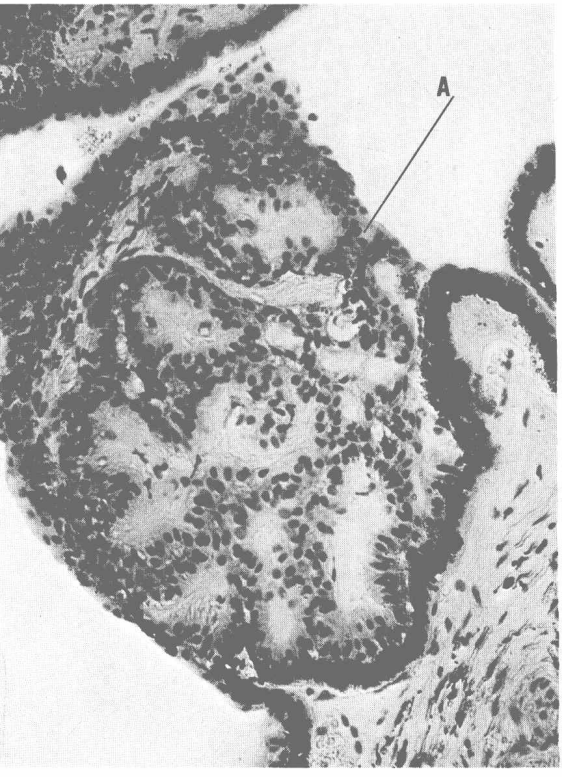

Fig. 3

Fig. 4