DIKTYOMA*

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The purpose of this paper is to report a case of diktyoma of the ciliary body and to give brief review of the main features of these rare embryonic tumours.

Epithelial tumours of the ciliary body are rare. They have presented problems of classification (Nordmann, 1941; Wadsworth, 1949; Reese, 1956) but the simple scheme proposed by Fuchs (1908) is quite adequate. He divided them into benign and malignant types. The malignant tumours he further divided into two groups according to whether they were composed of adult or of embryonic ciliary epithelium. The first group of malignant tumours may best be called carcinomata. They occur chiefly in adults and nearly always there has been some previous inflammatory or degenerative lesion of the eye. They have a relatively simple and uniform histological structure, being composed of interlacing bands, one or several cells wide, of cuboidal or columnar cells which closely resemble the non-pigmented layer of epithelial cells in the adult ciliary body.

It is with the second group of malignant tumours of the ciliary body that this paper is concerned. Early case reports were those of Badal and Lagrange (1892) and Emanuel (1900), but it was Verhoeff (1904) who first clearly indicated the nature of these tumours. He described a malignant tumour, arising from the unpigmented epithelium of the pars ciliaris retinae, which exhibited the structure of embryonic retina in various stages of development. He also recognized the close relationship between these tumours and retinoblastomata, both being tumours of embryonic retinal elements.

These embryonic tumours are rare. Andersen (1948), in a review of the literature, found 22 case reports and added one of his own. Two cases in this review however, those of Greeves (1911) and Hine (1920), are undoubtedly carcinomata and not embryonic tumours. Other cases have been reported by Alling (1904), Beattie and Dickson (1943), Kuhlenbeck and Haymaker (Case 11, 1946), Fralick and Wilder (1949), Gasteiger (1949), Gareis (1955), Malone (1955), and Newell (1956).

Nomenclature.—Although this is a rare tumour it has acquired a formidable list of titles. With our present knowledge some of the earlier names (carcinoma, malignant epithelioma, glioma, glioneuroma, terato-neuroma) are clearly inappropriate. Medullo-epithelioma is another name still applied to this group by several authors: it has also been used for the tumours of adult ciliary epithelium. The term was introduced by Bailey and Cushing (1926) for certain tumours of the brain which were said to arise from, and to resemble, the medullary epithelium of the primitive neural tube. However, the concept of medullo-epithelioma as a type of cerebral tumour has not been

* Received for publication July 12, 1958.

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generally accepted; the term has fallen into disuse for these cerebral tumours and its continued use for embryonic ciliary tumours is archaic. The use of "medullo-epithelioma" for tumours of adult ciliary epithelium is singularly inapt.

In the light of the current view of histogenesis of this tumour (from embryonic retina) the term "retinoblastoma of the ciliary body" would be ideal. This usage would correspond to that for embryonic tumours of the kidney (nephroblastoma) and of the liver (hepatoblastoma). However, there might be confusion with retinoblastomata of the retina and it is perhaps better to retain the term diktyoma. This name was introduced by Fuchs (1908) to indicate the netlike appearance of the tumour; it is not an ideal name but it is distinctive and it has been used by several authors.

Case Report

In September, 1955, a 2-year-old girl was brought to hospital by her parents who had noticed that she habitually screwed up the left eye during the previous month. There was a doubtful history of injury by a garden fork at the onset.

Examination under anaesthesia showed a cataract with secondary glaucoma, but no evidence of a perforating wound. A needling and curette evacuation was performed by Mr. L. H. Savin in an attempt to reduce the tension. The globe gradually increased in size, and in December, 1957, the eye was enucleated for cosmetic reasons.

Pathology.—Examination of the excised eye showed it to be much enlarged, measuring 28 × 25 × 24 mm., with an anterior staphyloma. After fixation and freezing the eye was bisected. The coats of the eye were found to be in situ. The anterior chamber was shallow with very extensive peripheral anterior synechiae and the optic disc was deeply cupped. Some lens matter and capsule remained.

It was difficult with the naked eye to identify any tumour: one could just make out an annular white ridge in the anterior part of the ciliary body.

Sections showed a grossly distended eye with well-developed cupping of the optic disc. A tumour (Fig. 1) growing in an annular manner replaced much of the ciliary body.

![Fig. 1.—Anterior part of eye showing tumour involving ciliary body and iris, obliteration of filtration angle, and staphyloma. Haematoxylin and eosin. ×16.](image-url)
The iris was atrophic in places; it was pushed forwards and was extensively adherent to the posterior surface of the cornea with complete obliteration of the filtration angle. The tumour had everywhere extended forwards from the ciliary body into the iris, though to a varying extent in different sites. In one small area the tumour extended posteriorly through the pars plana of the ciliary body almost as far as the ora serrata. From this region a fine filament of growth protruded into the vitreous. There was no retinal involvement.

In one area (Fig. 2) growth was limited to the anterior part of the ciliary body and the immediately adjacent part of the root of the iris. Elsewhere there was more extensive involvement of both ciliary body and iris. In the region shown on the right hand half of Fig. 1, growth had almost completely destroyed the ciliary body (Fig. 3) and extended along the iris almost to its pupillary margin.

**Fig. 2.**—More solid area of tumour involving anterior part of ciliary body and adjacent iris. The iris is atrophic and adherent to the cornea. Haematoxylin and eosin. × 50.

**Fig. 3.**—Ciliary body completely replaced by tumour. Numerous cysts are present and a retinolike area is seen in the uppermost part of the tumour. Haematoxylin and eosin. × 80.
In places the whole thickness of the iris was destroyed and replaced by growth which then lay in contact with the cornea (Fig. 4): in several small areas growth extended into the cornea for a short distance. The pupillary zone of the iris, though not infiltrated by growth, had its posterior surface clothed by tumour tissue (Fig. 5).

![Fig. 4.—Tumour replacing iris and covering posterior surface of cornea. Haematoxylin and eosin. × 80.](image1)

![Fig. 5.—Tumour lining posterior surface of iris. The tumour here is cystic and has a retina-like structure. Haematoxylin and eosin. × 80.](image2)

The structure of the tumour itself varied considerably from place to place. There were quite large areas of predominantly solid structure which were highly cellular. They were composed of closely-packed round or polyhedral cells with rather indefinite cell margins, hyperchromatic nuclei, and quite frequent mitotic figures. In this region rosettes were present (Fig. 6, opposite) identical with those of a retinoblastoma. Even in these more solid areas some cystic spaces were present (Fig. 2).
Much of the tumour had a netlike appearance which was due to the presence of numerous small cysts (Figs 3–5). Many of these cysts were lined by a single layer of columnar cells closely resembling the unpigmented epithelium of the ciliary body with which they were in continuity in several places. Other cyst walls were composed of several layers of cells and the lining cells were identical with those abutting on the cavities of rosettes.

Finally, some areas closely resembled developing or adult retina in structure (Figs 3 and 5). In these regions a clear-cut external limiting membrane covered a layer of closely-packed cells with oval nuclei: deep to this was a zone of fine fibrils in which cells with small deeply staining rounded nuclei were scattered.

Melanin pigment was found only in an occasional tumour cell and then only in areas where the normal pigmented epithelium of the iris or ciliary body had been destroyed. These tumour cells were presumably phagocytic. The tumour had a very scanty stroma and only occasional blood vessels were seen. A few minute necrotic foci were present. No areas of cartilage or calcification were seen.

**Discussion**

The present case is a typical one and brings the total number recorded up to thirty. Although information is not complete in all cases it is now possible to outline the main attributes of this tumour. Its history and histogenesis and the problem of terminology have already been mentioned.

Little is known about the aetiology. Cases have been reported from Europe, North America, South America, and Asia. There is no evidence pointing to any hereditary factor. The incidence is equal in the two sexes (male 12, female 13). The onset of symptoms is almost always in infancy or early childhood: undoubtedly, in
many instances, tumours are present at birth. Some abnormality of the eye was noted at or soon after birth in six cases, before the age of one year in five cases, between one and 5 years in thirteen cases, and "in infancy" in a further two cases. The remaining four cases presented later but they had advanced tumours: a child aged 6 with a large tumour invading the retina (Klien, 1939); two children aged 8 at onset, one with a blind eye (Bock, 1929) and the other with a staphyloma and scleral invasion (Schepkalowa, 1938); and the case reported by Soudakoff (1936) of a Chinese aged 28 who had had a perforated eye for one year and whose eye was almost completely replaced by tumour tissue.

These tumours are single and unilateral. Growth arises in the unpigmented epithelium of the ciliary body: theoretically, a tumour could arise in the iris but none has been reported. As the tumour enlarges, it involves the rest of the ciliary body, often spreading in an annular fashion, and projects into the cavity of the eye. It shows a great tendency to clothe surfaces and extends over the iris and over the lens and the zonule. Owing to the site of the tumour obliteration of the filtration angle occurs early, producing glaucoma and loss of vision and frequently staphyloma formation.

Many of the structural characteristics of this tumour are illustrated in the present case. Features of any stage of the developing retina can be produced. Typically there is a predominant netlike or polycystic structure, with the cysts lined by a single or multilayered epithelium. Rosette formation is common and the more solid areas closely resemble the structure of a retinoblastoma. Areas of fibrillar structure are common. There is a scanty connective tissue stroma. Blood vessels are not prominent, but the tumour is slow growing and necrotic foci are small and infrequent. Islets of cartilage were noted in six cases: this is not really a surprising finding as cartilage is common in embryonic tumours elsewhere in the body.

These tumours are undoubtedly malignant and there may be invasion of the iris, or less often of the retina, and then extension into the sclera and cornea. Growth of the tumour is very slow however. In eleven of the reported cases there was no invasion of sclera or cornea and the average age of these patients was 3·8 years. Nine patients showed invasion of sclera or cornea and their mean age was 5·6 years. In five cases growth had extended right through the sclera; this spread took place anteriorly and the orbit was not invaded: the mean age in this group was 15·4 years.

Furthermore, several cases have been observed over a long period of time before excision of the eye. Rubino (1941) reported the case of a girl whose eye was excised at the age of 19 when an extrabulbar mass was present. A "white pupil" had been noted at the age of 3 years and although a tumour had been diagnosed when the child was 10 years old the parents had refused operation at that time. In the case reported by Andersen (1948), a tumour had been suspected since birth; there was a slow and steady increase in size of the tumour until the eye was excised when the child was 16 years old.
Growth had then invaded the cornea but there was no extension outside the eye. The case reported by Redslob (1923) had buphthalmos at birth but the eye was not excised until the child was 11 years old, by which time the growth had perforated the sclera and produced an extrabulbar mass. Bock (1929) reported a case in a girl aged 16 who had had a blind eye since the age of 8: the excised eye showed episcleral extension of growth.

In only two cases was the orbit invaded, and in both instances metastases also occurred. In the first case (Schepkalowa, 1941) a child aged 9 months had an eye enucleated: one year later there was a recurrence in the orbit and lymph nodal metastases of similar structure. In the other case (Malone, 1955) the tumour extended into the orbit at the first operation when the child was 3 years old. Death occurred 3 years later with an orbital mass extending into the skull and brain and microscopic metastases in the lung.

The clinical mode of presentation was enlargement of the eye in seven cases, a visible tumour in seven, opacity of the eye in three cases, a "defective eye" noted at birth in three, pain and blindness in two instances each, and perforation of the eye in one case. An exotic manifestation of this tumour was beautifully depicted by Spicer and Greeves (1914-15). In their case cysts had become detached from the main tumour mass and could be clearly seen floating freely in the anterior chamber.

Owing to the early age at onset of the tumour and to its situation, the chances of preserving sight in an affected eye are almost zero. Indeed, many patients are blind when first seen. However, cure of the tumour is obtained in the vast majority of cases by excision of the eye. Apart from the two fatal cases already mentioned, no others showed even local recurrence of growth with periods of observation ranging up to 4 years.

Finally it is worth noting the main ways in which a diktyoma differs from a retinoblastoma. The site of origin is different. No cases have been reported of bilateral or multiple diktyomata and there is no evidence of any hereditary factor. The diktyoma has a more variable histological appearance and one typical feature is the presence of areas of netlike or polycystic structure. The diktyoma grows more slowly and has a much better prognosis.

Summary

A case of diktyoma of the ciliary body is reported. The main features of cases recorded in the literature are analysed.

We wish to thank Prof. H. A. Magnus and Mr. L. H. Savin for their advice and criticism, and Mr. G. Harwood for the photomicrographs.

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