LEIOMYOMA OF THE IRIS

The clinical and pathological diagnosis of tumours of the iris presents many difficulties. Because of their rarity it falls to the lot of few observers to have more than a passing acquaintance with them. Fear of their possible malignancy warps the judgment of the surgeon, and neither the clinician nor the pathologist approaches the problem with an unbiased mind.

In 1943 Kahler, Wallace, Irvine and Irvine reported "the seventh case of leiomyoma of the iris." Following the lead of Verhoeff, several reported myomata of the iris and ciliary body were rejected because of insufficient histological proof. On the other hand, it is not unlikely that many cases have been wrongly labelled leuco-sarcoma (unpigmented melanoma) because their real nature was not apparent with the usual haematoxylin and eosin stain. Van Gieson's stain and Mallory's connective-tissue stain show that the tumour cells take the stain characteristic of...
muscle. With Mallory's phosphotungstic acid haematoxylin the fibrillar nature of the tumour is apparent. Dense fibrils appear to take their origin from each end of the cell and course for long distances through the tumour without anastomosing. They bear no resemblance to the delicate anastomotic network of processes from the cells of a melanoma.

Clinically they may appear as grey, vascular tumours sessile or pedunculated. They may give rise to hyphaema, or as in the following case to cataract. They may be slow-growing or show periods of activity. Their diagnosis is of more than academic interest as they are benign and can often be completely removed by an iridectomy. For this reason every unpigmented or lightly pigmented tumour of the iris should be treated conservatively or subjected to biopsy before deciding on the fate of the eye. In the following case a clinical diagnosis of unpigmented melanoma was made with some confidence, and the eye removed. The histological findings were those of a leiomyoma.

On 6/1/47 Mrs. A. McH., aged 65 years, complained that the sight in the right eye had been failing for the previous twelve months. The vision in the right eye was 6/36, and could not be improved. The vision in the left was 6/12 and improved to 6/6. There was a sessile, unpigmented grey tumour occupying nearly the whole of the outer lower quadrant of the right iris. The outer edge was hidden by the limbus. It was very vascular and surrounded by a gelatinous fringe spreading over the surface of the surrounding iris. At the margin of the pupil there was a little ectropion of the pigment epithelium. The edge of the pupil appeared to be lifted forward, bringing the posterior surface of the iris into view and revealing a radial band of atrophy of the pigment epithelium. The lens was opaque behind the tumour, and the opacity was spreading across the pupil, obscuring the details of the fundus. The tension was normal, and remained so after the pupil had been dilated with homatropine and cocaine. The pupil dilated almost fully, and there was little evidence of distortion where the tumour abutted on the pupil. The most probable diagnosis appeared to be an unpigmented melanoma (leuco-sarcoma). The absence of rigidity of the iris, and the atrophy of the pigment epithelium introduced an element of doubt. The vascularity of the tumour, and the lens opacity which appeared, to be caused by it were thought to be in favour of the diagnosis of malignancy. Iridectomy was not considered because it was probable that the tumour had already infiltrated the ciliary body, so that removal would not be complete.

The eye was removed, fixed in Zenker's fluid, embedded in celloidin, and sections stained in haematoxylin and eosin, van Gieson, Mallory's connective tissue stain, Mallory's phosphotungstic acid haematoxylin and Wilder's reticulin stain. The sections showed a tumour (Fig. 1) extending from the pupil to the base of the iris and beginning to invade the ciliary body. It appeared to occupy the interstices of the iris, compressing the normal tissues rather than infiltrating them. The posterior surface of the iris was curved backwards, dimpling the lens where it had caused proliferation of the sub-capsular epithelium (Fig. 2), and destruction of the fibres beneath. The pigment epithelium
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FIG. 1
Mallory's connective-tissue stain. The more darkly staining area is the compressed iris tissue.

FIG. 2

FIG. 3
Haem. and eosin. section through the gelatinous fringe. Tumour cells growing along the anterior surface of the iris, and a large new vessel.

FIG. 4
Haem. and eosin. Base of the iris. Infiltration with round cells. Swollen tumour fibrils in the lower right-hand corner above the pigment epithelium.

of the iris had disappeared at the posterior limit of the curve. Anteriorly the tumour had compressed the normal iris tissue. There was no evidence that it was actually infiltrating this structure, but it had escaped through the openings of the crypts. Sections cut at the edge of the tumour showed a thin layer of tumour-cells spreading over the surface of the iris (Fig. 3), and accounting for the gelatinous fringe seen clinically. At the base of the iris there was evidence of a recent haemorrhage being absorbed in one of the crypts. Where the tumour was invading the ciliary body there was a collection of small round cells with some oedema (Fig. 4). The tumour was composed of bundles and
whorls of spindle-shaped cells ending in long fibrils (Figs. 5 and 6). The nuclei were rod-like and arranged in the so-called palisade formation. In some areas where the bundles were cut transversely (Fig. 6), many nuclei were to be seen in the one section. In others, the nuclei were very few in number, and the bundle composed mainly of fibrils. The chromatin of the large cylindrical nuclei was scattered in fine granules with one or two more dense aggregations. No mitotic figures were seen. Very little protoplasm could be seen surrounding the nuclei except in those cells lying near the dilator muscle. Here each end of the cell tapered off rapidly to a coarse fibril which could often be followed across several high-power fields. There was no evidence of any anastomosis with neighbouring fibrils. Its thickness did not always remain constant, and it would often present a marked fusiform dilatation at some distance from the nucleus. In the main mass of the tumour the amount of the protoplasm surrounding the nucleus was greatly reduced, and the fibrils appeared to split to surround it. The swelling of the fibrils was seen everywhere, even in the isolated bundles of cells creeping round the base of the iris to invade the ciliary body (Fig. 3). In cross-section it appeared as a circular homogeneous mass (Fig. 6). The cell body, fibril and dilatation stained a uniform pink with eosin, yellow with van Gieson, red with Mallory's connective-tissue stain, and a deep violet with the phosphotungstic acid haematoxylin, in contrast in the latter case with the red tinge of the violet of the connective tissue of the iris. Mallory's connective-tissue stain showed that the connective tissue in the section belonged to the compressed iris. There was little evidence of new formation of connective tissue between the tumour cells, and no reticulin. There were several large blood vessels in the tumour, and many fully formed capillaries with connective-tissue walls. None was embryonic in type. The pigment content of the tumour was small and confined to the compressed iris tissue, where chromatophores were present (Fig. 7). Pigment-laden cells, probably phagocytes, were present in large numbers in the spaces of the pectinate ligament in both angles. In the general picture seen with Mallory's connective stain, and with that of Wilder, it was obvious that the tumour had compressed the normal tissue, and if it had not found a way out through the crypts of the iris and through the loose tissue of the base of the iris, it would have become encapsulated thereby (Figs. 1 and 7). The compression of the superficial tissues of the iris was so marked that, when stained with haematoxylin and eosin, the cells of the anterior border layer formed a distinct line in the depth of the tumour right across the section (Fig. 6). The tumour had all the characteristics of a leiomyoma.
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FIG. 5
Mallory's phosphotungstic acid. Posterior section of the iris. The pig-
ment epithelium is at the lower edge.

FIG. 6
Haem. and eosin. The anterior border layer runs from the lower left to
the upper right-hand corner. The anterior surface is above this diagonal.
A transverse section of a bundle of fibrils in the lower right-hand corner
shows many fibrils of varying thicknesses and few nuclei. Compara-
tively more nuclei are present in the upper left-hand bundle.
FIG. 7
Wilder's reticulin stain. Ectropion of pigment epithelium with tumour cells growing over it and also splitting it. Reticulin is apparent only in the compressed iris tissue. The tumour has reached the surface through the orifice of a crypt.

FIG. 8
There was some atrophy of the pigment epithelium of the iris posterior to the sphincter muscle on the side of the pupil opposite the tumour (Fig. 8). This was associated with proliferation of the cells of the dilator muscle in that region. There was an abnormal appearance of chromatophores in the area, as if they were migrating forwards into the sphincter muscle. There were other round pigment-laden cells, probably phagocytes, in the vicinity, some in the space between the sphincter and the dilator, others on the posterior surface of the iris. This may have been an early nodule of tumour of the dilator muscle distinct from the main mass.

**COMMENT**

It is impossible to say from which muscle the tumour arose. Some sections appeared to show the cells of the dilator muscle elongating, gradually losing their pigment content, and merging into a general stroma of the tumour. Four factors call for special comment:—

1. The presence of haemorrhage. Van Duyse comments on this with the remark that intra-ocular haemorrhage in the presence of tumour does not necessarily indicate malignancy.

2. The presence of inflammatory exudate where the tumour was invading the ciliary body. In addition, an occasional ghost cell was seen along the posterior surface of the iris.

3. The presence of proliferation of the subcapsular epithelium of the lens due directly to the pressure of the tumour.

4. The presence of fusiform dilatations along the fibrils of the tumour. These receive no mention in the reports of Verhoeff, Frost, Ellett and Kahler et al. Van Duyse refers to a hyaline degeneration of the cell protoplasm seen in sections stained with Van Gieson and most obvious in transverse sections. He gives no reason for his assertion that hyaline degeneration was present. He also comments on the contrast between the yellow colour of the tumour fibrils which assimilate the picric acid of the van Gieson and the red of the connective tissue nearby, stained with fuchsin.

**SUMMARY**

A further case of leiomyoma of the iris is described. The clinical history and appearance vary greatly. Those of van Duyse, Verhoeff and Ellett had a history of twenty years' duration, that of Kahler et al. two, and of Frost only one. At
the time of operation the patient of van Duyse was twenty-eight, of Verhoeff thirty-three, of Frost forty-six, of Ellett forty-seven, and of Kahler et al. forty-six. In each case the tumour was on the lower part of the iris, one in the lower nasal quadrant, one in the middle, and four (including the above) in the lower temporal quadrant. In the case reported by Verhoeff the tumour was suspended from the anterior surface of the iris by a narrow pedicle. The remainder presented as sessile tumours which appeared to originate from the depths of the iris. All were vascular, and several were prone to recurrent haemorrhage. van Duyse records that his patient had nine hyphaemas before iridectomy was performed. The only record of cataract is in the present case.

The site of the tumour calls for comment. Although the number of cases is too small to allow sure conclusions, it is remarkable that all were situated in the lower half of the iris. This would suggest that the tumours had arisen near the foetal fissure, and that their origin may be traced to a defect arising at the time of its closure. Verhoeff states that in his case the tumour was "nowhere connected with the iris muscles or pigment epithelium" and that it seemed likely "that it originated from stroma cells of the embryonic uvea, possibly from misplaced cells which ordinarily would have taken part in the formation of the ciliary muscle." The situation of the tumour in all cases reported adds some weight to his suggestion.

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REFERENCES