Benign epithelioma of the iris

A clinico-pathological case report

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Neoplasms of the pigmented epithelium of the eye are rare, those of the iris epithelium being especially unusual. Reviews by Ashton (1964) and Duke-Elder and Perkins (1966) present benign epitheliomas of the iris as black masses arising from the epithelium and pushing forwards into the stroma. The present case is completely different in nature, and appears to be the first of its kind reported.

Case report

A farmer aged 50 years vaguely noted a change in the vision of his right eye for several months. In March, 1969, he attended an optometrist who found a lesion in the right iris. No abnormality had been noted by the same optometrist during an examination 5 years previously. The patient was sent to Melbourne for ophthalmological management.

Examination (March 24, 1969 : R.F.L.)

Both eyes were free of inflammation and the irides were of the blue type with scattered honey-coloured freckles.

The right eye showed a pale yellow spherical mass approximately 2 mm. in diameter within the iris stroma almost midway between the pupil margin and the root of the riris in the upper temporal sector (Fig. 1). Large curly new vessels coursed in the iris stroma from the periphery radially to,
around, and over the tumour extending to the pupil margin while, deep in the stroma and almost obscured by it, other vessels could be seen in the 12 o'clock position slightly away from the tumour.

The pupil margin was drawn slightly towards the tumour, there was no ectropion of the pigment epithelium and the pupil reactions were very little affected. The involved segment of the iris appeared to be lifted away from the lens which could be seen to have anterior opacities in this area. The visual acuity in each eye was 6/9.

Management

The tumour was causing lens opacities and as it did not involve the periphery of the iris, excision was advised rather than observation. The tumour presented such unusual features in its duration and appearance that no presumptive diagnosis could be given, but leiomyoma of the iris was thought to be probable. Some apprehension was felt for difficult haemorrhage from the numerous aberrant vessels.

The tumour was excised by sector iridectomy, after radial cuts on each side. No undue bleeding occurred at the time. On lifting the iris, a smooth, avascular, cream, globular tumour was seen to protrude from the pigment epithelium and a circular small crater in the anterior lens surface was seen at the site previously occupied by the tumour.

Three days postoperatively a moderate hyphaema showed as clotted blood over most of the iris, but this cleared in 5 days and progress was otherwise uneventful. In the sector iridectomy the lens showed irregular, anterior, radial opacities as well as the small crater noted above.

Result

One year postoperatively the visual acuity in the right eye was 6/6 with +0.5 D sph., +1.5 D cyl. axis 30°. A year later the vision was the same and the lens opacities were unchanged.

Pathology report (C.H.G.)

Microscopic appearances

Sections showed a lentil-shaped growth measuring 2 mm. in diameter by 1 mm. in thickness situated on the posterior surface of the iris midway between the extremities of the excised tissue. The growth was covered anteriorly by the iris and posteriorly by an incomplete layer of pigmented iris epithelium. The iris stroma contained large thin-walled supernumerary vessels devoid of the wide adventitial sheaths characteristic of normal iris vessels.

The tumour consisted of fusiform, cuboidal, and polygonal epithelial cells which appeared to have originated from the iris pigment epithelium round the perimeter of the growth (Fig. 2, opposite). A minority of the tumour cells contained melanin granules. In the centre of the growth the tumour cells were compressed into thin strands to form a network. The interstices of this net were filled with eosinophilic stroma in which small endothelial-lined channels containing red blood cells could be discerned. These vessels were probably branches of the large vessels in the iris.

The stroma varied in appearance, being faintly fibrillated and vacuolated in some areas while in others it was structureless and hyalinized. In the centre of the growth the stroma had undergone granular calcification and in the immediate vicinity of the granules there was a small amount of mucoid substance (Fig. 3, opposite). The appearance of this stroma and its relationship to the epithelial cells of the tumour suggested that it was a product of these cells and perhaps an excessive formation of basement membrane substance.
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Discussion

This benign epithelioma of the iris pigment layer is different from previously reported black tumours of the iris epithelium. Macroscopically it was pale yellow and microscopically the pigment cells were collected mainly around the periphery. In the centre of the tumour the pigment cells appeared to have taken on another function in forming an eosinophilic stroma with the excitation of a very vascular reaction.

Ashton (1964) showed the interrelationships between pigment epithelium and smooth muscle cells which both arise from the pigment layer of the neuroepithelium; and how tumours may arise containing cells showing not only the distinct features of these two cell types but also intermediate cells with features in common, thus making cell classification difficult.
The pigment layer of the neuroepithelium has other potentialities, namely, the formation of basement membrane. This is best seen with retinal pigment epithelium, whereas this function is not developed in iris epithelium. However, in tumour formation, some cells may manifest suppressed potentials and this benign epithelioma of the iris may have done so. The calcification may represent degeneration in an abnormally produced basement membrane substance.

Its appearances together with its unusual posteriorly directed growth are perhaps significant in indicating deeply ingrained features of the posterior neuroepithelium in contrast to the myoepithelial-pigment cell features of the leiomyomas which favour the anterior layer of the neuroepithelium.

Summary

A cream-coloured, rapidly growing vascularized tumour of the iris was excised and found on microscopy to be an unusual benign epithelioma of the iris. Propositions are submitted to suggest that this tumour has a different pathogenesis from the usual (but rare) pigmented benign epitheliomas of the iris and leiomyomas.

We wish to thank Dr. J. Paul Borger for referring this case and for his skilled assistance in management.

References