Medulloepithelioma involving the iris

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Pigmented lesions involving the iris are rare in childhood and most commonly take the form of naevi arising from stromal melanocytes. Tumours of the pigment epithelium in children are exceedingly uncommon, Ashton (1964) describing a single hamartomatous lesion in a review of iris tumours seen at the Institute of Ophthalmology in London, and Morris and Henkind (1968) failing to record any instance of true neoplasia at this age. In this report we describe the clinical and pathological findings in a child with a tumour apparently arising from the pigment epithelium of the iris, in the absence of discernible ciliary body involvement, which histologically resembled a benign medulloepithelioma.

Clinical findings

A 3-year-old white girl was referred because of poor vision of 3 months' duration and an abnormal appearance of the right eye. Ophthalmic examination showed a pigmented nodular mass projecting into the anterior chamber with occlusion of the lower half of the pupil and appearing to arise from the inferior part of the posterior surface of the iris. Contact between the tumour and the lens was associated with a diffuse cataract, while anteriorly the mass was adherent to the corneal endothelium. Through an inferior *ab externo* incision the mass was removed and a sector iridectomy performed. A little pigment has remained on the corneal endothelium with some persistent overlying oedema. No local recurrence of the tumour has been seen after 10 months and consideration will be given to lens aspiration in the future.

Pathology

The resected specimen consisted of two rather fragmented and twisted pieces of tissue measuring approximately $6 \times 3 \times 3$ mm and $5 \times 4 \times 4$ mm. Microscopy of paraffin-embedded sections stained with haematoxylin and eosin showed infiltration and replacement of the iris stroma by large heavily pigmented cells which, although apparently randomly distributed in some places, in others showed a cord-like and even tubular arrangement (Fig. 1). Bleached sections showed that the proliferating cells were mainly cuboidal or polygonal with small compact nuclei and voluminous cytoplasm. Areas were also present in which the cells were only lightly pigmented and in such areas there was an orderly alignment along convoluted basement membrane-like structures (Fig. 2).

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The outer surface of the epithelial proliferations was covered by an alcianophilic mucinous material and in places where a tubular arrangement was seen the membrane-lined surface of the epithelium faced the lumen. Between the epithelial proliferations some areas contained cells with elongated nuclei and long fibrillary processes suggestive of glial differentiation (Fig. 3).

Comment

From a histological standpoint the differential diagnosis includes four possibilities: hyperplasia of the pigmented epithelium, adenoma of the pigment epithelium, medulloepithelioma, and magnocellular naevus (melanocytoma). The last of these can be fairly easily discounted since, although many of the heavily pigmented cells possessed the typical small nuclei with abundant cytoplasm, this diagnosis would not account for the areas showing a trabecular or tubular structure. Adenomas are essentially adult tumours arising from fully differentiated epithelium and would be most unusual in a child of 3 years: furthermore, while such tumours may similarly form a mucinous material, the secretion is usually to be found within the 'gland' lumina rather than on the outer aspects of the tubular structures. Hyperplasia is properly regarded as a cellular proliferation propagated by extrinsic factors such as trauma or inflammation and, consequently, in the absence of any evidence of underlying pathology there would seem little reason to suggest this diagnosis.

Evidence in favour of medulloepithelioma lies chiefly in the finding of cells aligned along a basement membrane-like structure with mucinous material on their opposite surface. This type of arrangement can be considered to recapitulate the embryonic secondary optic vesicle, with the mucin being analogous to developing vitreous and the membrane representing the outer limiting membrane of the retina. The presence of elements resembling glial tissue can also be construed as evidence in support of this diagnosis. Andersen (1962) has previously referred to the possibility that medulloepithelioma may arise from the iris, but he commented also that the limitations imposed by a small specimen of tissue make confident diagnosis difficult: correspondingly, origin from the anterior part of the ciliary body with secondary involvement of the iris cannot be excluded with certainty in the present case, even though the

FIG. 1 *Histological section showing part of the resected iris in which the stroma is infiltrated by pigmented epithelium arranged in cords and tubules. Haematoxylin and eosin. $\times 185$*

FIG. 2 *Part of the tumour showed spaces lined by a single layer of lightly-pigmented epithelium. The intervening stroma was largely composed of mucinous material. Alcian blue and neutral fast red. $\times 455$*

FIG. 3 Section showing glial elements within the tumour. Haematoxylin and eosin. $\times 455$

lesion appeared to be confined to the iris at the time of surgery. Nevertheless, although this type of tumour typically arises from the non-pigmented epithelium of the ciliary body, there is no theoretical barrier to its also involving the iris, especially as the pigment epithelium of the iris is continuous with the non-pigmented epithelium of the ciliary body. Zimmerman (1971) considered that the predilection of medulloepithelioma for the ciliary body can be ascribed to the relative delay in the maturation of the medullary epithelium in this region, rather than to any intrinsic difference, and added that identical tumours had been observed in the retina and even the optic nerve head (Reese, 1957). More recently, Green, Iliff, and Trotter (1974) and Mullaney (1974) have described malignant teratoid medulloepitheliomas involving the optic nerve head, in the latter case, apparently arising from the remnants of a primary hyperplastic vitreous.

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There is little reason to doubt the benignity of the present lesion in view of its regularity and inconspicuous mitotic activity, so that local excision would seem to have been the correct treatment.

Since writing this paper and 14 months after local resection the tumour has recurred. Enucleation has been performed and, contrary to the initial finding, there is now unmistakable evidence of ciliary body involvement.

Summary

The case is described of a benign medulloepithelioma in a child which originally was apparently confined to the iris.

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