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Editorial: Prognosis in ocular melanoma

The following note was received after we went to press with last month's issue. It is the editorial which should have appeared in that number and it would be a great pity to miss it, since it answers many of the questions asked in the stopgap editorial. It illustrates nicely the difference in interpretation of a difficult subject between the ordinary clinician and the specialist.

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There is no doubt that the survival prospects of individuals who develop uveal melanoma are better when this tumour arises in the iris than when it does so in the ciliary body or the choroid. The relatively benign behaviour of iris melanomas in comparison with the behaviour of those which develop at other sites within the uveal tract may be explained by special features of their clinical presentation and by their cytological characteristics.

The experience from posterior melanomas is that large tumour size is the single most important clinical indicator of a poor prognosis for survival,[1] though a large volume is also closely linked with epithelioid cell type.[2] Iris melanomas are externally visible, and this probably accounts for their tendency to be diagnosed at a younger age[3-5] and when they are smaller than melanomas elsewhere in the eye.[6] The better outlook associated with iris melanomas may depend substantially on their tendency to present at an earlier stage in their development.[7] The most important cytological indicator of a poor prognosis for posterior melanomas is the presence in the tumour of a high proportion of epithelioid cells.[8,9] Iris melanomas are notable for having few epithelioid cells and a preponderance of naevoid or spindle A cells of doubtful metastatic potential, so that the division between a benign and malignant tumour is less distinct than elsewhere in the uvea.[5] This feature of iris melanomas led one author to reclassify 87% of a group of iris melanocytic tumours, previously diagnosed malignant on histological criteria, as benign naevi.[10]

The accepted treatment of an iris melanoma which is deemed to have malignant potential is surgical excision. Sector iridectomy is recommended for localised tumours not involving the drainage angle and without evidence of dispersion of melanoma cells within the eye. Enucleation is reserved for diffuse iris melanomas and those with intraocular dissemination. Local surgical excision of an iris neoplasm is associated with ocular morbidity, and most melanocytic tumours in this location pose no threat to life or to the function of the eye. Nevertheless it has been shown that some 10% of iris melanomas which do contain epithelioid cells will metastasise even though they are small.[6] It follows that there is a need to distinguish clinically between the harmless tumours which require no treatment and the aggressive tumours which constitute a threat to life. Possible clinical indicators of malignancy are large tumour size and evidence of growth, though benign lesions may expand and invade adjacent intraocular structures.[6,10] Many iris lesions of worrying appearance have been present for a long time, and surprisingly few can be documented to grow over a five-year period of observation.[11] Fluorescein iris angiography may define the limits of tumour involvement but is of uncertain value in predicting malignancy.[6] In one study it was considered helpful in only 50% of cases.[12]

The trend away from immediate surgical treatment of iris melanomas and towards a policy of serial observations to record growth is probably a reasonable compromise, though it may miss some tumours which have epithelioid cells and which cannot be shown to increase in size. The experience from posterior melanomas is much greater than that from anteriorly situated tumours. It is by no means certain that any treatment influences the metastatic rate of posterior uveal melanomas, because it is likely that many lesions have already disseminated while very small.[13] This observation may also apply to iris melanomas, but, until it can be shown how big a tumour must be before it is too large for excision or destruction to be beneficial to survival, it seems wise to institute active therapy as soon as there is clinical evidence that any uveal tumour is malignant. For iris melanomas, as for those more posteriorly situated, it remains at issue whether eye conserving procedures offer as good survival prospects as enucleation of the eye.

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Editorial: Retinal vein occlusion and glaucoma

For a long time there has been a tradition among ophthalmologists that occlusion of the central retinal vein (CRVO) is more common in cases of primary glaucoma, even of a mild degree, than in the rest of the population. However, such cherished ideas do not always bear close examination.

Verhoeff[1] found CRVO in eight of 39 cases of glaucoma and 'endovasculitis at the level of the lamina cribrosa' in all the others. But these were cases of secondary glaucoma, and it was not until Foster Moore's important monograph in 1924[2] that primary glaucoma became implicated. It has to be remembered as well that many of Verhoeff's cases, even if they had originally been of primary glaucoma, were in any case terminal in that they came to enucleation, and most of us would accept central vein occlusion as a not unexpected finding in an eye with 'absolute glaucoma'.

Foster Moore's paper, which incidentally is extremely difficult to winkle out from most of the major ophthalmological libraries in London, is quite modest in its claims. He studied 31 cases of branch vein occlusion (BVO) and 31 of CRVO. His chief conclusions were that the principal cause of occlusion was arteriosclerosis associated with hypertension and this was more marked in BVO and CRVO. The second conclusion, which is the one that has received the most attention, was that there was a possible link between primary glaucoma of 'an insidious type' and CRVO. He based this conclusion on the fact that well marked cupping of the optic disc was seen in only one of 31 cases of BVO but 13 of 31 cases of CRVO. There is really not much more evidence than this, but from this fairly shaky start the tradition has originated.

Salzmann[3] in 1933, in another painstaking monograph including a wealth of beautiful clinical and histological drawings, thought that the glaucoma might lead to venous occlusion by causing collapse of the veins. This is quite distinct from the type of glaucoma which results from rubeosis iridis due to retinal ischaemia,[4] though it was pointed out in 1955[5] that in some cases primary glaucoma could precede the CRVO-neovascular glaucoma sequence. In the latter paper the author described a number of illustrative cases and followed much the same lines of reasoning as Foster Moore, and pointed out the existence of primary glaucoma in the other eye. In three of the cases pathological specimens became available, but all they showed were changes typical of neovascular glaucoma. It has to be admitted that the paper, which was written by the author of this