Ocular Oncology Demystified

[a]Vikas Chadha
[b]Mandeep S. Sagoo

a. Scottish Ocular Oncology Service, Tennent Institute of Ophthalmology, Gartnavel General Hospital, Glasgow, UK.
b. Ocular Oncology Service, Moorfields Eye Hospital; Retinoblastoma Service, The Royal London Hospital; and NIHR Biomedical Research Centre for Ophthalmology, Moorfields Eye Hospital and UCL Institute of Ophthalmology, London, UK

Contact details:
Vikas Chadha: vchdh@aol.com
Mandeep S. Sagoo: m.sagoo@ucl.ac.uk

Editorial for Special Issue “Ocular Oncology”
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Ocular oncology occupies a unique position in ophthalmology. Not only are these conditions sight threatening, but they are also capable of eye loss, and many have the potential to shorten life. This issue, borne out of the symposium "Ocular Oncology Demystified" at the Royal College of Ophthalmologists, is dedicated to our late colleagues, Victoria Cohen, FRCOphth and John Hungerford, FRCS, FRCOphth.

The topics covered encompass the frequently encountered and the rare. The assessment of choroidal naevi, the commonest intraocular tumour seen in adults needs to be differentiated from small melanomas using imaging rather than tissue biopsy. DiSimone et al show that general ophthalmologists in the USA underutilise imaging modalities that are required in the TFSOM-DIM system for assessing such lesions.[1] The alternative MOLES system devised by Damato, does not require ultrasound and its rationale is reviewed for estimating the likelihood of malignancy in choroidal melanocytic tumours, allowing community practitioners to manage or refer such lesions appropriately.[2] These are important considerations in setting up virtual clinics[3] and for artificial intelligence algorithms that may in the future detect and refer to ocular oncologists only the lesions that are likely to need treatment.

Such is the utility of ocular imaging in choroidal tumours that a biopsy (trans-vitreal or trans-scleral) is seldom required for diagnosis, but more commonly performed for prognostic information in uveal melanomas.[4,5] Counselling patients regarding the benefits versus risks on an individual basis is important. On the horizon there are targeted treatments based on testing of uveal melanomas, and liquid biopsies may also make it easier to deliver personalized care.

Current treatments for uveal melanoma include plaque brachytherapy and proton beam radiotherapy. In the UK, the Clatterbridge cyclotron treated 1084 eyes with

uveal melanoma over a 10-year period, referred from the 4 national centres for ocular oncology.[6] Indications for proton beam over ruthenium plaque radiotherapy included poorly accessible small posterior tumours and large anterior tumours. Tumour control was up to 93%. In another study on ruthenium plaque radiotherapy for posterior melanomas, final visual acuity was ≥6/12 in 44%, with vision being dependent on development of radiation macular oedema.[7] The main therapy for the latter is by antiVEGF injections, given either prophylactically in high risk cases or once the vasculopathy develops.[8] Emotional distress after diagnosis of melanomas is related to physical factors, such as poor visual outcome, younger age at diagnosis, and psychological factors.[9] The latter includes the fear of systemic relapse, though there is widespread variability in the modality and frequency of imaging for distant (liver) relapse.[10]

Ocular surface tumours comprise benign and malignant lesions, but the caruncle is often overlooked. We are reminded that this tissue, which is derived from the lower eyelid rather than the conjunctiva, can manifest cancers such as lymphoma, basal cell carcinoma, squamous cell carcinoma, sebaceous carcinoma and melanoma.[11] Differentiating some types of conjunctival neoplasm is now possible with imaging, with the relishing prospect of the *in vivo* biopsy. Sripawadkul and colleagues found that using anterior segment optical coherence tomography, conjunctival papilloma, compared to ocular surface squamous neoplasia, had a thicker epithelial layer with an edge overhanging normal epithelium, corrugated epithelial surface, intrinsic spaces and posterior shadowing.[12,13] In contrast, conjunctival lymphoma, which is an extranodal mucosa-associated lymphoid tissue low grade cancer, requires a tissue diagnosis for histopathological, cytogenetic and molecular features.[14]

Oral nicotinamide is a form of Vitamin B3, available in food or in supplements. It has a beneficial effect on UV-induced immunosuppression of the skin, and

hence in some high-risk individuals there may be a role in the management of actinic keratoses and non-melanoma skin cancers.[15]

There is overlap between orbital surgery and ocular oncology. Lacrimal gland lesions are relatively common and Awotesu et al describe their experience of performing lacrimal gland biopsies in 248 patients during a 21-year period in a tertiary referral centre. Chronic inflammation (69%) lymphoma (15%), adenocarcinoma (4%) and pleomorphic adenoma (2%) were the histopathological diagnoses.[16] Other rare orbital tumours include solitary fibrous tumours which occur mainly in the middle aged. Though usually benign and locally infiltrative, rarer malignant forms can develop and metastasize.[17] Orbital tumours in the paediatric age group are uncommon, and Keren et al describe a series of paediatric orbital lymphomas.[18]

Paediatric ocular oncology is focused on retinoblastoma. Shields et al conducted a wide-ranging review with the latest ideas on genetics, global burden of disease, chemotherapy outcomes and psychological impact.[19] As globe salvage becomes a reality for many patients with intraocular retinoblastoma, attention needs to be paid to the visual outcomes of treatment.[20] Apart from retinoblastoma, medulloepithelioma is a very rare intraocular tumour of children, that can be teratoid or non-teratoid and benign or malignant. In high-risk cases after enucleation, features of malignancy prompted adjuvant chemotherapy to prevent metastasis.[21]

We have stood on the shoulders of giants to scan the horizons of our subject. There is much that has been learned in ocular oncology, but even more that needs to be explored and researched. Jeanon et al report a case of a medial orbital basal cell carcinoma that had globe sparing surgery to remove the tumour using

robotically-assisted wide local excision.[22] This may well pave the way for future directions in ocular cancer surgery.

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