Solid Tumour Section

Mini Review

Head and neck: Posterior uveal melanoma

Karen Sisley

Department of Ophthalmology and Orthoptics, Royal Hallamshire Hospital, Sheffield, S10 2JF, UK (KS)

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Identity

Note
Melanomas derived from the pigmented uveal tract of the eye, which comprises the iris, ciliary body and the choroid.

Classification

Note
Choroid melanomas are the commonest (60-70%), and iris melanomas are particulary rare (5-8%).

Clinics and pathology

Embryonic origin
Neuroectodermal cell lineage.

Epidemiology
Annual incidence in the USA of 5-7 million population annually, similarities to cutaneous melanoma as the incidence is higher amongst fair skinned pale eyed individuals; median age for presentation 55 years, and a slightly higher incidence for males; the aetiology is unclear, but possibly sun exposure is a risk factor.

Clinics
Obscured vision symptoms of a retinal detachment, cataract formation, and occasionally painful eye.

Pathology
Classified into three categories depending on cell type; spindle cell melanoma has the best prognosis, epithelioid is most likely to spread, whilst mixed cell melanomas have an intermediate behaviour; (of note, the size and position of the tumour also affects the prognosis of individual melanomas); uveal melanomas can invade locally within the eye, and form deposits in other organs, but most commonly the liver.

Treatment
Local resection, enucleation, photocoagulation, external beam irradiation, brachytherapy, and laser therapy.

Evolution
Metastasis occurs, mainly to the liver, with approximately half of patients treated with enucleation dying within ten to fifteen years; the highest rates of metastasis occur in the first five years, but have been recorded over forty years after the primary tumour was detected.

Prognosis
Spindle cell tumours, and those less than 10 mm in diameter have the best outcome; ciliary body melanomas and those where there is scleral invasion have a worse prognosis.

Cytogenetics

Cytogenetics Morphological
Most analysis performed on ciliary body and choroid melanomas; usually relatively simple chromosome alterations, often with diploid karyotypes; the commonest alterations are those of chromosomes 1, 3, 6, 8 and the Y chromosome; monosomy 3 and isochromosome 8q occur in association in ciliary body melanomas, and are both independently predictive of poor prognosis, with the increase in the copy number of 8q also indicative of the disease free interval; chromosome 1 changes have been linked with large aggressive tumours and abnormalities of chromosome 6 are suggested to indicate a better prognosis.

Cytogenetics Molecular
Studies are not as comprehensive as cytogenetic studies; comparative genomic hybridization has mainly confirmed cytogenetic data, but also identified less
frequent alterations including those of chromosome 9p; no specific genes implicated, p16, p53, BRCA 2 have been investigated but appear to be involved in only a small percentage of tumours; studies suggest that genes on chromosome 3 play a role, and several regions have been implicated; no association between MYC and poor prognosis has been made, and flow cytometric analysis suggests that DNA index is not a reliable predictor of prognosis.

References


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