Case Study

Monocular Malignant Melanoma

Case History

A 52 year old Caucasian female presented with a number of naevi on the skin and a right ciliary body malignant melanoma twelve years ago and had an iridocyclectomy for this. (Figure 1.)

The patient has had previous excellent vision of 6/6, normal pressures and no evidence of ocular inflammation. However, early in 2017 in a follow up to the oncology clinic the patient now presented with secondary uveitis and glaucoma, a raised intraocular pressure of 44mm Hg and a reduced vision of 3/60 in the right eye.

Epidemiology/Background:

“Although melanoma of the eye is rare, it is one of the most common types of eye cancer in adults.

“About 3% of all melanomas are ocular origin; of these, 85% are uveal, and the majority are choroidal. Uveal melanomas are rare, with standardised incidence rates ranging from approximately 2 to 8 cases per 1 million people in the United States and Europe. Mean age at diagnosis is approximately 60 years, with the incidence rate increasing to age 70. Risk factors for development of uveal melanoma include history of choroidal nevi, ipsilateral nevus of Ota*, and atypical mole syndrome. Weak correlations have suggested sunlight exposure as a possible causative agent. Welders and metal workers have also been reported as having a slightly higher incidence of uveal melanoma, possibly due to ultraviolet light exposure.” [5]

“Ciliary body melanoma occurs relatively often in the Caucasian population and has relatively low incidence in Asian or black populations. In addition, the incidence of the disease is similar for both genders and for both the left and right eyes. It is commonly observed in one eye and rarely observed in both eyes” [6].

“Ciliary body melanoma can develop as a circumscribed or annular type. The circumscribed type is often brown and can invade the anterior chamber, causing elevated intraocular pressure. In contrast the annular type grows around the ciliary body in a ring-type fashion. Ciliary body melanoma may be initially misdiagnosed as open-angle glaucoma because of the difficulty in visualising a distinct mass.” [1]

*Ipsilateral nevus of Ota also known as oculodermal melanocytosis. This is hyperpigmentation which occurs on the face usually blue or grey in colour. First noted by Dr. M.T. Ota of Japan in 1939.
Clinical Findings and Diagnosis

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In 2005 a trans-scleral biopsy was performed which confirmed a malignant melanoma of the ciliary body which invaded the ciliary body and iris root. The tumour was most probably of posterior uveal type (rather than of iris origin). [16]

Upon examination in early 2017 of the high intraocular pressure, the doctor decided to prescribe the patient Acetazolamide (Diamox) and Brimonidine (Alphagan) twice a day for a week before review.

At the same visit it was agreed that the patient should have liver screening to check if there was any progress with previous distant benign cysts. This ultrasound scan showed the patient to have extra liver cysts and which have gradually increased in size and quantity from previous visits, but they were not metastasis. It has been strongly suspected the cysts are focal nodular hyperplasia.

“The liver is the primary site of distant metastasis for melanomas, followed by the skin and lung; brain metastasis rarely occurs. ”[1]

The patient was then discussed at the Multi-Disciplinary Team Meeting to see what further action was required.

It was decided upon further review and investigation that the Diamox and Alphagan did not reduce the patients’ intraocular pressure far enough and therefore the patient needed surgical intervention via a trabeculectomy to reduce the raised intraocular pressure in the right eye.

The raised intraocular pressure was due to the obstruction of melanoma pigment in the trabecular meshwork, this caused a disruption to the flow of aqueous humour.

A trabeculotomy was performed in early 2017.

Unfortunately, complications arose: inflammation and the use of steroids caused a rapidly dense progressive cataract to form and due to the disruption in the aqueous humour flow, extra pigmented melanoma deposits formed on the inner surface of the cornea. [17]

This reduced the patients’ vision down to 3/60. (Figure 2.)

It was also noted that the zonules normally should be white, but the melanoma has turned them brown. (Figures 3, 4.)
Treatment

Under examination it was agreed that the patient urgently needed phacoemulsification and a vitrectomy due to the vitreous coming forward and it was also damaged from the iridioectomy. A capsule tension ring was used to stabilise the intraocular lens as the zonular dehiscence area from the iridioectomy was damaged. In turn the zonules would break and become weak if not used. The patient was then prescribed Levofloxacin (Oftaquix), Dexamethasone (Maxidex) and Ketorolac Tromethamine (Acular). [17]

Prognosis/Outcome

Biopsies of the iris root, anterior capsule, anterior vitreous and ‘phaco wash’ were taken during the cataract surgery and were sent to histopathology.

A three week follow up appointment was arranged where it was confirmed to the patient that there was a reoccurrence of the melanoma in the root of the iris root, and the angle of the ciliary body at 2 o’clock. [17] (Figures 5,6)

Unfortunately, the diffused pigmented cells spread into the posterior and anterior chamber. (Figures 7,8)

It has been thought that a trabeculotomy opening can spread the melanoma pigment further and in turn increase the likelihood for metastasis to occur.

The patient is now awaiting either a Ruthenium Plaque Radiotherapy which stops the tumour cells dividing and starves the tumour by destroying its blood supply, this will eventually cause a reduction in the size of the tumour.

Proton Beam Therapy is the second option which uses beams of protons (sub-atomic particles) to achieve the same cell-killing effect. A "particle accelerator" is used to speed up the protons. These accelerated protons are then beamed into cancerous cells, killing them. [9,10]

If there is reoccurrence in the future, enucleation of the right eye has been discussed.
Figure 1. Diffused light showing pigment around iris and coloboma from iridioipectomy

Figure 2. Melanoma deposits on inner surface of cornea

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Figure 3. Retrolumination of iridiocyclctomy and pigmented zonules.

Figure 4. Broad beam of pigmented zonules

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Figure 5. [S.Connell] - Ultrasound Biomicroscopy of melanoma obstructing trabecular meshwork [7]

Figure 6. [P.J.Toomey] - Slit lamp gonioscopy of melanoma obstructing trabecular meshwork. [8]

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Figure 7. OPTOS high resolution colour and autofluorescence of macular and optic disc showing pigmented cells which have invaded the anterior chamber.

Figure 8. OCT horizontal line showing pigmented cells deposited on inner limiting membrane.

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References

E-books

Case reports

Books

Journals

Images

Leaflets
Websites


Interviews
