Abstract

A 51 year old man presented to the clinic with complaints of a missing portion of his visual field OS. Upon funduscopic examination the patient was diagnosed as having an amelanotic choroidal melanoma of the left eye.

History

A 51 year old Caucasian male presented to the ocular disease department of the Eye Care Center at the Southern California College of Optometry with a complaint of “a dark spot off center” from his central vision in the left eye. Associated symptoms also included an increase in floaters in his left eye over the past three months.

Ocular history was unremarkable. The patient reported that 10 months earlier he had a CT scan ran because of a suspicion of lymphoma, however he had not heard back with the results of the scan and assumed that it came out normal. His medical history was positive for hypertension, osteoarthritis, obesity and head trauma. He also reported having a heart attack two years earlier. A positive family history for cancer, diabetes, heart disease, and thyroid disease was documented. He was taking atenolol and a daily aspirin for hypertension.

Diagnostic Data

Best corrected visual acuities were 20/15 OD and 20/40 OS PHNI. HRR color vision testing was normal. Pupils were equal in size, reactive to light and near stimuli. No afferent pupillary defect was detected in either eye. Extraocular motility was full and smooth in both eyes. FDT visual field screening showed no defect OD, however a dense inferior nasal defect was present OS. Amsler grid testing was normal with no signs of
distortion or scotoma OD. In OS the patient noted inferior-nasal distortion and relative
darkening of the Amsler lines in this location.

Anterior segment slit lamp examination was normal OU; including lens, which
showed no signs of opacity, and corneas, which were clear in both eyes. The anterior
chamber was deep and quiet with no signs of cells or flare and angles were open on Von
Herick method OU. Goldmann applanation intraocular pressures were 26 mmHg OD and
OS.

Dilated 20D and 90D funduscopic evaluation of OD showed normal and
unremarkable vitreous, macula, retina, and retinal vasculature. Cup-to-disk ratio was
estimated at .30 x 30, rim tissue appeared pink and healthy. However the funduscopic
examination of OS revealed a large, depigmented, vascularized, elevated lesion
approximately 3 disk diameters in size (see Figures 1 & 2). Vascular deflection over the
lesion was noted. The lesion was located superior and temporal approximately 5 disc
diameters from the optic disc and two disc diameters above the macula. Inferior to the
lesion and superior to the macula was a liner subretinal hemorrhage running horizontally.
The optic disk appeared normal with a cup-to-disk ratio of .30x.30 and pink and healthy
rim tissue. The macula appeared flat and avascular with no signs of macular edema.

**Diagnosis**

Based on the findings the lesion in the left eye was diagnosed as an amelanotic
choroidal melanoma. Due to patient’s case history, the possibility of ocular lymphoma
was not ruled out.

**Treatment and Follow up**
Due to the potentially aggressive and lethal nature of this type of lesion the patient was immediately referred to a retinal specialist for confirmation of diagnosis and treatment. A few weeks following the referral a letter from the retinal specialist was sent to the Eye Care Center, which confirmed the initial diagnosis of an amelanotic choroidal melanoma. In the letter it was recommended that the patient receive immediate treatment for the melanoma due to the vision and life threatening nature of the lesion. The treatment recommended in the letter was plaque radiotherapy of the left eye.

Discussion

Choroidal malignant melanoma is the most frequently diagnosed primary intraocular tumor in adults (1). Most commonly the tumor will present in the 6th decade of life. It is rare, however, for the tumor to present after the age of 80, and there is a less than 5% incidence of the tumor in adults less than 30 years of age (2). The highest rate of presentation is in Caucasians with an incidence estimated at 5 cases per million each year (2). The most common presentation of a choroidal melanoma is as an elevated, pigmented, often oval or round shaped mass (3). Less commonly, the lesion may present like the one described in the case report, amelanotic (lacking pigment), and range from the aforementioned complete amelanotic variety to very densely pigmented black lesions (1,4).

Since the tumor is located in the choroid continued growth of the mass may lead to a rupture of Bruch's membrane allowing the tumor to progress into the retina and vitreous (5). Once Bruch's membrane has been compromised the tumor will often take on a mushroom shaped appearance on funduscopic examination (5). Other common clinical
presentations associated with malignant melanomas of the choroid include the accumulation of orange pigment (lipofuscin) in the retinal pigmented epithelium (rpe) (5). Choroidal folds, hemorrhaging, cataracts, uveitis, and secondary glaucoma can also be associated with choroidal melanoma at varying rates of presentation. Large tumors can also lead to secondary exudative retinal detachments (6).

Choroidal melanomas are often discovered in asymptomatic patients during a routine funduscopic examination (7). However symptoms such as visual field loss and a reduction in visual acuity can be reported similar to those described by the patient in the above case report. Diagnosis is often made clinically from the funduscopic appearance and associated symptoms (7). If histological diagnosis is required a transvitreal fine needle aspiration is the method most often used (1).

When making a differentiation between malignant choroidal melanoma versus choroidal nevi factors to take into consideration include thickness (>2mm), the presence of orange pigment clumps (lipofuscin), the absence of hard surface drusen, the presence of a serous retinal detachment over the lesion and margin of the tumor involving the optic disc (1,7,8,9). These criteria are more indicative of a potentially life threatening malignant choroidal melanoma. Patients presenting with 3 or more risk factors have a greater than 50% chance or tumor growth over the next five years (7, 10). Therefore proper examination of associated findings is very important in the initial diagnosis. However a close monitoring schedule should be considered even for harmless appearing nevi as their presence alone increase the risk of developing malignant melanoma in the future (3). Utilization of fundus photography can be very helpful in following nevi from year to year in order to monitor for changes in size, shape, and coloration. B-scan ultrasoundography
is another quick effective means of evaluating suspicious lesions for elevation and possible extraocular extension and should be used without hesitation (11).

It is estimated that 50% of all patients diagnosed with choroidal melanoma will die due to metastatic extension within 15 years of diagnosis, so early detection and treatment is of the utmost importance (11). Treatment modalities depend on several factors including the tumors size and location, the patients overall health status, and the patients age (1, 9). Enucleation is considered a last resort and is often not necessary as long as the tumor has not yet involved the optic disc or 1/3 of the ciliary body and angle (1). Plaque radiotherapy is often used as a first line of treatment. This treatment involves giving the tumor a high dose of radiotherapy from a close distance using a radioactive plaque placed over the tumor base (1). If successful the tumor will gradually flatten over a period of a few years leaving a pigmented mass with surrounding choroidal atrophy (11). While the lesion may have reseeded, a dilated funduscopic examination will be required every 6 months to one year for the rest of the patient’s life to ensure there is no reactivation (11). It will also be important to have regular screenings to monitor for metastases to other locations in the body including the brain and liver (1, 11).

Due to the potentially lethal end result of primary choroidal melanomas, it is highly important for the optometric clinician to be able to recognize presenting signs and symptoms of this condition. Though rare, the devastating outcome of misdiagnosis highlights the need to closely evaluate all suspicious looking choroidal lesions. Proper case history and funduscopic examination will ensure that nothing is missed, and if there is ever a doubt a second opinion is always a wise next step.
References


