Early Detection of Uveal Melanoma

Monitoring nevi in annual examinations can help find and treat tumors sooner.

BY KOMAL B. DESAI, BA, MD

outine eye examinations are an important regimen for patients, not only because they help evaluate visual acuity, but also because they can help detect eye disease and other adverse health conditions. A check for glaucoma is a typical part of the routine eye examination. Less common is a routine check for uveal melanoma.

The incidence of uveal melanoma, the most common primary intraocular cancer in adults, is relatively low, as it is for other forms of eye cancer. These tumors can be difficult to spot and monitor due to their slow growth. The fact that they grow slowly might suggest that the need to check for uveal melanoma is less urgent than for other diseases. However, if undetected, these tumors can be deadly.

It is estimated that 2730 new cases of eye cancer, primarily melanoma, will occur in the United States in 2014, affecting 1290 women and 1440 men.¹ The International Society of Ocular Oncology estimates that ocular melanoma affects about 6500 individuals worldwide annually.²

Cancers of the eye and orbit will claim an estimated 310 lives in 2014, including 130 men and 180 women, the American Cancer Society estimates. Many more may experience vision loss. Metastasis can also occur, although research indicates that iris melanomas tend to be slow growing, small, and somewhat dormant, and thus they rarely metastasize.

Uveal melanoma can be treated successfully if caught early. Eye care professionals should keep this rare form of cancer in mind when conducting routine eye examinations and providing other eye care.

SYMPTOMS OF UVEAL MELANOMA

Cancers in or near the eye can include the following:4

- intraocular lymphoma, most often non-Hodgkin; many doctors believe this to be a type of central nervous system lymphoma
- · hemangioma, a benign vascular tumor of the choroid and retina

- · conjunctival melanoma, affecting the membrane that lines the eyelid and eyeball
- eyelid carcinoma (basal or squamous cell), a variation of skin cancer
 - · lacrimal gland tumor
 - · retinoblastoma, a form of childhood eye cancer

Uveal cancer tends to occur in white individuals with light eye color, fair skin, and the ability to tan. However, these conditions do not necessarily indicate a susceptibility to this disease.⁵ Patients with dysplastic nevus syndrome (many abnormal moles on the skin) may have a higher risk of developing uveal melanoma.⁶

People with uveal melanoma usually have no symptoms. Only in advanced forms will these cancers affect vision or cause appearance anomalies that would cause a patient or a doctor to suspect disease. Still, there are steps that can and should be taken in a routine eye examination to check for eye cancers. If doctors observe a feature in a patient's eye that could be cancerous, they should benchmark its size and be prepared to check it later to ensure that it is not changing.

MONITORING NEVI

Melanoma is typically thought of as a form of skin cancer, so patients may be surprised when it occurs in the eye. However, the routine checks used to monitor for melanoma on the skin are similar to those advised to monitor for melanoma in the eye. There is, however, this critical difference: The patient cannot personally check for ocular melanoma. Rather, these checks should be part of a routine eye examination.

Uveal melanoma begins when pigmented cells in the eye called *melanocytes* or *nevi* grow uncontrollably. Nevi in the iris and back of the eye are risk factors for iris or choroidal melanoma, types of uveal melanoma.

Not unlike birthmarks or beauty marks on the skin, nevi are seemingly harmless when labeled as moles. However, these lesions require surveillance to detect cancer, just as birthmarks, beauty marks, and moles on

the skin do. Doctors can advise patients to watch marks on the skin, but that is not possible for moles in the eye. Thus, the eye care physician has the obligation to monitor nevi carefully and to refer any condition that may be suspicious.

To conduct surveillance of nevi successfully, the examining doctor must check the eye before pupil dilation. The comprehensive eye examination may often begin with dilation, but iris nevi may be difficult or impossible to check after pupil dilation. Choroidal nevi are seen after dilation during evaluation of the retina.

Nevi conditions to observe include size, shape, color, and distortion of the pupil. Cataract may also suggest presence of a melanoma, including cataract in only 1 eye.

When nevi are observed, the patient should be advised. There need not be alarm. The patient should be reassured that continual monitoring by the patient's regular eye care professional and review by a referred professional are aimed at ensuring his or her continued good health, good vision, and well-being.

If nevi are observed, photography of the nevi and other documentation to establish a baseline for future comparison is recommended.

Iris nevi of 1 mm or greater and choroidal nevi of 3 mm or greater size warrant evaluation, further exploration, or referral. Slight changes in the nevi in size, color, or associated inflammation may be addressed with more frequent follow-up, such as at 3 or 6 months instead of 1 year. Dramatic changes merit referral to an ocular oncologist. Generally, anything that may be suspicious is fair game for another professional opinion.

Clinical differentiation between a nevus and a melanoma may be difficult. Investigations performed by ocular oncologists may include optical coherence tomography, transillumination, intravenous fluorescein angiography, ultrasonography, ultrasound biomicroscopy, and photography.²

CASE STUDY

Although intraocular melanoma tends to be a greater risk for older, white men with light-colored eyes, uveal melanoma can occur at any age. At Sabates Eye Centers, an 18-year-old woman presented with iris melanoma.

The patient had visited an office in our practice at age 11, but no melanoma was detected. She did not return for routine eye examination until 7 years later. When the patient received a routine eye examination prior to entering college, an iris melanoma was detected and treated.

Treatment for this form of cancer depends on the

location and the size of the tumor. Options can include surgery, radiation, resection, or a combination of these methods.^{2,7} Enucleation is another option.

In this case, the patient was treated with radiation therapy for 5 days in July 2014. Thereafter, 6-week checks were made to ensure that the tumor responded to the treatment. By mid-October 2014, the patient had a visual acuity of 20/20. The tumor was scarred down, and we considered the condition to be controlled. Re-checks will continue and will be reduced in frequency to quarterly for 1 year, then semiannually for 5 years.

CONCLUSION

The routine eye examination is the front line of uveal melanoma detection. Nevi are common, but they should not be routinely dismissed. A responsible approach is to explore any suspicious conditions. If nevi are present but not unusual-seeming, the best approach is to photograph the nevus and document its presence in the patient's record. The patient should be advised of the presence of the nevus. Upon subsequent exams, comparison should be made to detect change that warrants further exploration and referral.

Rather than waiting for something small to get out of control, doctors should monitor even the smallest growth or change in symptoms to act preventively. These cancers are rare and often undetected because they can be difficult to spot, and they are difficult to monitor due to their slow growth. Nevertheless, uveal melanoma is the most common primary intraocular cancer in adults. It can be treated successfully when treated early.

Komal B. Desai, BA, MD, is an assistant professor of ophthalmology and director of the Ocular Oncology Service at the University of Missouri-Kansas City School of Medicine and an ophthalmologist specializing in ocular oncology at Sabates Eye Centers in Kansas City, Missouri. Dr. Desai may be reached at (913) 261-2020; desaik@sabateseye.com.

- 1. What are the key statistics for eye cancer? American Cancer Society. http://www.cancer.org/cancer/eyecancer/detailedguide/eye-cancer-key-statistics. Accessed October 20, 2014.
- FAQs Melenoma [sic]. International Society of Ocular Oncology. http://www.isoo.org/melanoma. Accessed October 20, 2014.
- 3. Yap-Veloso MI, Simmons RB, Simmons RJ. Iris melanomas: diagnosis and management. *Int Ophthalmol Clin.* 1997;37(4):87-100.
- 4. Eye Cancer: Overview. American Society of Clinical Oncology. http://www.cancer.net/cancer-types/eye-cancer/overview. Accessed October 20, 2014.
- 5. Weis E, Shah CP, Lajous M, et al. The association between host susceptibility factors and uveal melanoma: a meta-analysis. *Arch Ophthalmol*. 2006;124(1):54-60.
- 6. What are the risk factors for eye cancer? American Cancer Society. http://www.cancer.org/cancer/eyecancer/detailedguide/eye-cancer-risk-factors. Accessed October 20, 2014.
- 7. How is eye cancer treated? American Cancer Society. http://www.cancer.org/cancer/eyecancer/detailedguide/eye-cancer-treating-general-info. Accessed October 20, 2014.