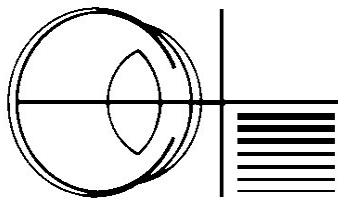


# **Ocular Melanoma**

## **Diagnosis and Treatment**

A Guide for Patients



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**Ophthalmic Oncology Center**

Tara A. Young, M.D.  
Bradley R. Straatsma, M.D.  
Temisan Amoruwa, Clinical Coordinator  
Lariza Sayas, Surgical Coordinator

Jules Stein Eye Institute  
100 Stein Plaza  
Los Angeles, CA 90095-7000  
(310) 206-7484  
[ophthalmiconcology@jsei.ucla.edu](mailto:ophthalmiconcology@jsei.ucla.edu)

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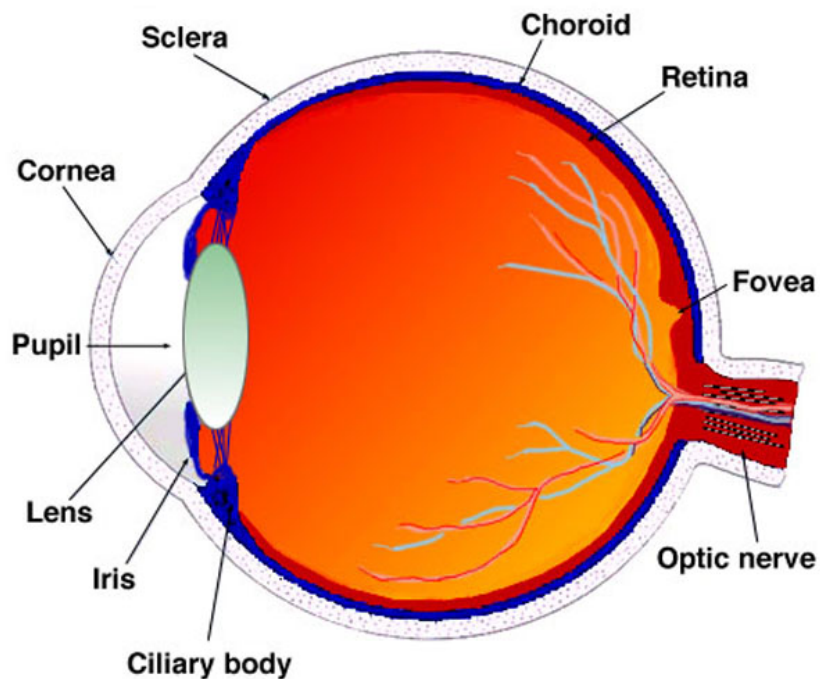
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## I. Anatomy of the Eye

The eye is a sensory organ connected to the brain by the optic nerve. The white outer coat of the eye is the sclera. The clear jelly filling the inside of the eye is the vitreous. The cornea and lens at the front of the eye focus images on the retina. The retina lines the inside of the back of the eye and functions as film does in a camera. It receives light through the pupil, the opening at the front of the eye, and sends messages to the brain via the optic nerve. The brain interprets these messages as sight. The choroid is pigmented tissue underneath the retina and it is full of blood vessels which nourish the retina. The iris and ciliary body are extensions of the choroid in the front of the eye. Uvea is the term used when speaking of the iris, ciliary body, and choroid as a unit.



## **II. Malignant Melanoma of the Eye**

### **Definition**

A melanoma is a primary tumor of the skin and also the eye. In the eye it arises from the pigmented cells of the uvea (choroid, ciliary body, or iris) of the eye and is not a tumor that started somewhere else and spread to the eye. Malignancy means that the tumor is a cancer which may spread to other parts of the body. When a tumor spreads to another part of the body it is called a metastasis. Not all tumors metastasize. Although some choroidal melanomas are more life-threatening than others, all should be treated as if they were malignant.

Melanoma of the eye occurs in about 6 people per million population per year. It almost always occurs in one eye and your other eye is usually at no higher risk to develop this tumor. There is usually no inheritance so your family is at no higher risk to develop this tumor. Interestingly, eye melanoma is rarely seen in dark skinned individuals and is more common in fair skinned, blue-eyed people.

### **Diagnosis**

Melanoma within the eye is diagnosed during an examination of the eye by an ophthalmologist. Your eye doctors are able to recognize a melanoma by the typical color, shape, and location of the tumor and by other features observed in an eye examination.

The diagnosis is suspected in our office as we examine your eye with the slit lamp machine and the headlight. A detailed drawing of the melanoma will be done as we map out the characteristics of the tumor. Sometimes transillumination

is used in the office. Transillumination allows us to see the tumor through the eye wall by directing a beam of light to the outside of the eye.

Some special tests enable us to be more certain of the size of the tumor and support the diagnosis of melanoma. These tests will be done by us at your first visit and possibly at some later visits. These tests, which usually cause no discomfort, include eye photography, ultrasonography (sound waves) and fluorescein angiography (intravenous dye studies of the back of the eye). There is no radiation involved in these tests.

In the photography tests, a bright flash will be used to photograph the back of your eye. In some cases, a special lens is necessary to place on your numbed eye to get high quality angled pictures of the tumor. In the ultrasonography test, soundwaves are directed towards the tumor by a small probe placed over jelly on the surface of the closed eye. The pattern made by reflection of the sound waves helps your doctor judge the thickness of the tumor. This is important in planning treatment. The fluorescein angiography test is useful in studying the blood vessels of the melanoma. In this test, a tiny amount of dye is injected into a vein in your arm. As the dye passes through the blood vessels in the back of the eye, a rapid sequence of photographs is taken through your pupil. For several hours after the test, your urine will be a yellow-green color as your body rids itself of the dye. The dye is harmless to the body, but some people may be allergic to it.

## **Treatment**

In the past it was almost always necessary to surgically remove the eye which contained an eye melanoma. In recent years, new methods of treatment have been developed which may be used to save the eye. There are now several ways to treat melanoma of the eye depending on several findings – the age and

health of the patient and the size, location, thickness, and growth of the tumor. The goal of treatment is to stop the growth of the tumor in the eye.

### **Observation**

In very small lesions, which are not highly suspicious, we may advise you that it is best to just do photographs and recheck the tumor in the office every few months. If the tumor does not grow in these months, then photographs, ultrasound, and repeat examinations may be continued because the tumor is inactive and may be a benign mole of the eye. If the tumor grows, then some other form of treatment is generally necessary.

### **Laser**

Laser treatment is rarely used for melanoma of the eye, but may be used for other small tumors in the back of the eye. It may take many sessions of laser to completely treat the tumor.

### **Radiation (Plaque Radiotherapy)**

Radiation treatment is a common form of treatment of the eye melanoma. It is used for small or medium sized tumors. The method of giving the radiation at the Jules Stein Eye Institute is as follows: a small radioactive plaque (about the size of a coin) is applied in the operating room to the eye. The plaque is made of gold and it looks like a button with radioactive seeds on one side. The gold plaque provides a platform for attaching the radioactive seeds and also functions as a radiation shield to protect the back of the eye. The ophthalmologist surgically secures the radioactive plaque to the eye over the area of the tumor. The side of the plaque with the radiation is placed directly over the tumor. The plaque radiation remains directly over the involved eye and does not affect the opposite eye, the brain, or the rest of your body. It is left in place for an exact

number of days to provide adequate radiation to the entire tumor. Usually, the patient receives 4 to 6 days of treatment. The radiation plaque is then removed in the operating room and you may be discharged the same day. Once the plaque is removed, you are not radioactive and none of your belongings are radioactive.

The radiation plaque causes gradual shrinkage of the tumor but the tumor may never disappear entirely. It will always remain as a scar in the back of the eye and will be seen on our examination. The goal of radiation is to sterilize the tumor of its malignant cells and the remaining cells are slowly eliminated by the body over many months. The effects of radiation may not be seen for 4 to 6 months or so. In a small percentage of patients (< 2%) the tumor regrows after radiation treatment and in these cases, a second plaque is necessary or the eye may need to be removed (enucleation).

Radiation may cause a decline of vision as a side effect of the treatment, but the majority of patients do well with fair vision in the affected eye and excellent vision in the unaffected eye. Radiation may damage some healthy parts of the eye. Damage to the blood vessels of the retina (radiation retinopathy) or to the optic nerve often causes a gradual loss of vision. In some cases, hemorrhage into the inner part of the eye (vitreous cavity) may occur and cause loss of vision. Radiation damage to the lens may cause a cataract, which may require removal by surgery sometime later. After radioactive plaque treatment, some patients note some dryness or irritation of the eye which usually can be relieved by the use of the eye drops called "artificial tears." Initially, it is not uncommon for the patient to have double vision because the muscles are moved aside during the surgery to allow placement of the plaque. In the great majority of cases, single vision returns gradually over several months as the stretched muscles regain their normal length.

Radiation can also be delivered by Proton Beam Therapy. In most cases the results of tumor treatment are no different than with plaque radiotherapy.

### **Biopsy of the Tumor**

At the time of plaque surgery a biopsy of your tumor will be performed. This not only confirms the diagnosis before proceeding with radioactive plaque treatment, but may also provide prognostic information about the tumor itself. In tumors that have a loss of chromosome 3 (monosomy 3) the risk of developing metastasis is higher than a tumor with a normal chromosome 3. This genetic abnormality is only found in the ocular melanoma, and not in the rest of your body. Therefore, this abnormality can not be transmitted to your offspring. You will be informed of this result after your treatment.

### **Enucleation**

Prior to the 1960's, the usual treatment for choroidal melanoma was enucleation (removal of the eye). Enucleation is still used to treat large melanomas where other treatments will not work.

The surgery for enucleation is done in the operating room. The eye is removed and a hard plastic nonreactive ball implant (about the size of the eye) is placed in the gap where the eye was removed. The patient is discharged from the hospital with a patch over the operated eye. In 2 to 4 weeks, a prosthetic eye (glass or plastic eye) is fitted. These eyes are artistically designed to appear exactly as your remaining eye. It is sometimes difficult to tell which eye is the plastic eye and which eye is real! However, the artificial eye does not move as fully as the real eye. There is no surgery at present to transplant an entire eye and this futuristic surgery will not be available in our lifetime.

After enucleation, there is a reduced field of vision on that side of the body when looking straight ahead and there is loss of depth perception as well. You can imagine what enucleation would be like by closing or patching one eye. Many of the skills of depth perception may be relearned with time and most patients continue with their same jobs and activities without any problems

whatsoever. Protective glasses or goggles are needed to protect the remaining eye during dangerous activities or sports. We have treated hundreds of people who have lost one eye and who continued to live normal, productive lives. You may wish to read the book *A Singular View: The Art of Seeing with One Eye* by Frank B. Brady.

### **Resection**

Resection of the eye melanoma means to cut out the tumor from the eye and leave the rest of the eye intact. This is used for only certain types of tumors, and may be combined with radioactive plaque placement.

The surgery for resection is done in the operating room and may require at least 2 to 4 hours to complete the surgery. It is quite tedious and is done only in a few places in the world because of the high level of skill required to perform the surgery. Our doctors will need to see you every few weeks and if all goes fine, the appointments are made for every few months after that.

Although the surgery removes the tumor, the side effects of the surgery may lead to blood in the eye because as the eye is cut open, it bleeds. The blood will blur your vision, but in most cases it clears on its own in a few weeks or months. Also, the eye is at risk for retinal detachment which can be repaired with additional surgery.

### **Your Team of Specialists**

If you are diagnosed with ocular melanoma, a team of specialists will help you get the best treatment, follow-up and resources necessary to facilitate your quality of life. Education, information and reassurance are important throughout this process. We want you to be aware of the team members that may be helping you and their role in your care.

### **Ocular Oncologist/Ophthalmologist**

Your ophthalmologist is a physician with special training in diagnosis and treatment of problems within the eye. Some ophthalmologists have a specialty in working with different types of problems of the eye and here at UCLA we have ophthalmologists with a special interest in ocular melanoma. Your ocular oncologist/ophthalmologist will diagnosis your melanoma, will make recommendations for treatment and will involve other specialists in your care. If surgery is needed -- for enucleation, placement of the radiation plaque or removal of the tumor -- the ophthalmologist will perform these operations. The ocular oncologist/ophthalmologist is the primary doctor taking care of your eyes.

### **Radiation Oncologist and Physicist**

The radiation oncologist is a physician who is specially trained in using radiation to treat cancers. If radiation is used to treat your melanoma, a radiation physicist will create your individualized plaque. You will be scheduled to have an appointment with the radiation oncologist and physicist. They are involved in designing the plaque to be the right size and shape and containing the right amount of radioactive material that will destroy the tumor. They work with the ophthalmologist to develop just the right type of plaque for you which will then be implanted by the ophthalmologist.

### **Optometrist**

An optometrist is often involved to assess the vision in your eyes. The optometrist plays an important role if corrective lens are needed to improve your vision.

### **Medical Oncologist**

A medical oncologist is a physician who is trained first in internal medicine and then develops a specialty in malignant tumors. At UCLA we have medical oncologists with specialties in melanoma and we work directly with these physicians. Because cancers have the ability to spread to other organs, the medical oncologists will play an important role in your initial evaluation and future follow-up. The medical oncologist may perform blood work and send you for special scans of your liver and other organs. He or she will answer your questions related to cancers and what kind of follow-up is needed. If your cancer does spread (you develop metastasis), the medical oncologist would be the physician who becomes your primary physician for the treatment of the metastatic melanoma. Because UCLA is a research institution dedicated to advancing research toward the eradication of cancer, many new treatments are being developed and research studies conducted in the area of metastatic cancer. Our patients may have the opportunity to participate in these studies if appropriate.

### **Ted Mann Family Resource Center**

The diagnosis of any type of malignancy often creates feelings of anxiety, sadness and worry on the part of patients and family members. These are normal feelings and reactions. Here at UCLA, we are committed to helping patients live fully and with the best quality of life after any diagnosis of a malignancy.

As part of your care, you will be scheduled for an appointment with a Health Psychologist or Oncology Social Worker at the *Ted Mann Family Resource Center* a place designed to help patients and family members face the challenges brought about by cancer and its treatments. It is located at UCLA across the street from the Jules Stein Eye Center. The staff at the Center is made up of specialists who understand the medical and psychosocial concerns of

patients and their families. In this meeting, you will have an opportunity to discuss how this diagnosis is affecting you and your family, clarify concerns or questions and develop effective coping strategies. For example, if you desire you will be able to learn proven techniques for managing stress and anxiety related to medical visits, such as upcoming surgeries or scans. There is no charge for this appointment. You are welcome to come alone or bring someone with you.

The Resource Center has many additional resources that may be helpful to you. We strongly urge patients and family members to go to the initial appointment and determine what services may be beneficial to them. You may call them directly at (310) 794-6644 to set-up or change your appointment. There is additional information on their web site at [www.CancerResources.mednet.ucla.edu](http://www.CancerResources.mednet.ucla.edu). The Resource Center is located in the 200 UCLA Medical Plaza Building, Suite 502 and is open from 8:30 am – 5:30 pm, Monday through Friday.

### **Other Team Members**

As in all medical situations, there are a variety of people including nurses, technicians, anesthesiologists, and surgery coordinators and other administrative people who will work together to provide you with access to the best information, resources and care.

### **Follow Up Appointments**

Your follow up appointments will be scheduled depending on the type of treatment that you were given. In general, an enucleation requires the fewest examinations after the surgery and resection requires the most examinations after the surgery. Despite the form of treatment, a good eye examination will be necessary at least 2 or 3 times a year for the rest of your life. A good physical

examination with liver blood tests and imaging is required two times a year for the rest of your life. This will be performed by your oncologist. The results of your medical examination and tests should be reported to us at each visit.

### **Prognosis**

After developing a melanoma of the eye, your life prognosis depends on several factors. These factors include the location and size of the tumor, the type of cells within the tumor as seen under a microscope, and the presence or absence of monosomy 3 (a loss of one pair of chromosome 3) in the tumor tissue. Your prognosis is dependent on whether the cancer spreads (metastasizes) to other important organs. The risk of spreading of the tumor does not seem to depend on the type of treatment. Even if the eye is enucleated, you are still at risk to develop spread of the tumor. In general, patients with eye melanoma do better than patients with many other malignancies. However, the tumor does have the capacity to spread. Once metastases are detected, survival worsens. When eye melanoma spreads, it usually goes to the liver first and lung second. Treatment by a medical oncologist is then necessary.

### **III. Research**

There are many research studies being performed on eye melanoma here at the Jules Stein Eye Institute. Research on better understanding the biology of this cancer to approaches toward systemic therapies to prevent spread of the tumor are presently being studied. Methods of detecting and treating early spread of the malignancy are being tested.

#### **IV. Funding**

Contributions to the Ophthalmic Oncology Center are graciously accepted. Please speak directly to our staff if you wish to make a donation. The funds provide support for research in eye melanoma, for treating patients with eye melanoma who have no insurance and cannot afford the treatment, and for teaching doctors all over the world how to recognize and treat eye melanoma.