

UNDERSTANDING

Ocular Melanoma



University of Michigan
Kellogg Eye Center

**UNDERSTANDING
OCULAR
MELANOMA**

Kellogg physicians have cared for patients with ocular melanoma and other retinal diseases for many years. Our team of specialists is dedicated to delivering the most current treatments and the most compassionate care to patients who must deal, often suddenly, with a diagnosis of ocular cancer.

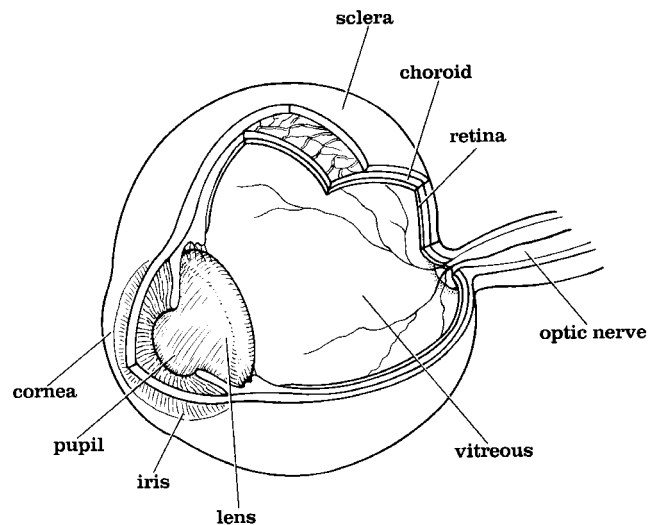
Our patients say that access to clear information is extremely helpful as they review decisions about their care. We hope this brief overview of ocular melanoma will be useful as you proceed through your treatment.

WHAT IS OCULAR MELANOMA?

Ocular melanoma is the most common primary cancer of the eye. Its incidence is about 3 per 500,000 individuals per year. It occurs most often in lightly pigmented individuals with a median age of 55 years. However, it can occur in all races and at any age. In the majority of cases, ocular melanoma slowly develops from the pigmented cells of the choroid (*choroidal melanoma*), but it also can develop from the pigmented cells of the iris (*iris melanoma*).

Ocular melanoma is a malignant tumor that can grow and spread (or *metastasize*) to other parts of the body which can result in death. There are, however, effective treatments, as described later in this booklet.

Ocular melanoma is completely different from skin or cutaneous melanoma. Ocular melanoma is not related to sun exposure; and its biology and routes of spread (*metastasis*) are very different from that of skin melanoma.



*Drawing courtesy of
the Collaborative Ocular
Melanoma Study Group.*

**HOW IS
OCULAR
MELANOMA
DIAGNOSED?**

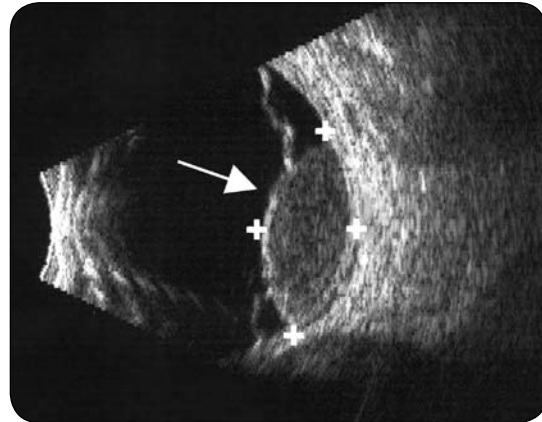
Small choroidal melanomas typically do not cause any symptoms. However, as the tumor enlarges, it can cause a variety of symptoms including light flashes, floaters, and loss of vision.

Ophthalmologists can diagnose most choroidal melanomas with a technique called *ophthalmoscopy*. Using specialized equipment, the ophthalmologist can examine the tumor through the cornea, which provides a window into the eye.



Dr. Grant Comer, Director of Ocular Oncology, examining one of his patients.

Certain tests help confirm the diagnosis. Ultrasound or echography, which uses sound waves to visualize the tumor, is very useful in assessing the tumor size, composition, and vascularity (blood flow). The Kellogg Eye Center is fortunate to have Ms. Kathleen Meyer as its ultrasonographer. Ms. Meyer is nationally recognized for her experience in ocular ultrasound.



The arrow in this ultrasound image points to the tumor in the eye. The mass is located between the “plus” signs.

Fluorescein angiography is occasionally helpful. In this test, sequential photographs of the tumor are taken after a yellow dye, fluorescein, has been injected into a vein in the arm.

In specialized Ocular Oncology Centers like the Kellogg Eye Center, the accuracy of the clinical diagnosis using these tests is 99.7 percent. Thus, unlike other cancers, biopsy is rarely necessary to confirm the diagnosis of a choroidal melanoma. Biopsy is necessary only when the diagnosis is uncertain.

UNDERSTANDING STAGING AND CHOROIDAL MELANOMA

Once the diagnosis of choroidal melanoma is established, additional procedures are required to ensure that the tumor has not spread (*metastasized*) through the blood to other organs. These procedures include a chest x-ray and a specialized x-ray called a computerized tomogram (CT) of the abdomen.

With the clinical examination and additional tests, it is possible to “stage” choroidal melanoma. In other words, to identify its size and the extent to which it exists in the body. There is no universally accepted staging system, but a common system is the TNM system used by the American Joint Committee on Cancer.

The following page includes a detailed description of these stages.

Tumor Staging System

T = size of tumor

N = spread to lymph nodes. Choroidal melanoma rarely spreads to the lymph nodes, as there are no lymphatic channels in the eye.

M = metastasis to distant organs

Definition of Ocular Melanoma Stages

Stage 1A	The tumor is 7 mm or less in diameter and is less than 2 mm in height. The tumor has not spread to distant organs.
Stage 1B	The tumor is 7-10 mm in diameter and is 2-3 mm in height. The tumor has not spread to distant organs.
Stage 2	The tumor is 10-15 mm in diameter and is 3-5 mm in height. The tumor has not spread to distant organs.
Stage 3	The tumor is more than 15 mm in diameter or greater than 5 mm in height. The tumor has not spread to distant organs.
Stage 4A	The tumor extends outside the eyeball but it has not spread to distant organs.
Stage 4B	The tumor may be any size, and has spread to distant organs, typically the liver.

Note: One millimeter is approximately the thickness of a dime.

TREATMENT OPTIONS

While it is unsettling to hear the words “ocular melanoma” from your ophthalmologist, this diagnosis is not a reason to despair. Ocular tumors, especially small ones, are very treatable and there is a high likelihood that your tumor can be completely destroyed. There are times, however, when a tumor is resistant to treatment or is too large to be treated conservatively. In these cases, in order to lessen the possibility of metastasis, the eye must be removed (*enucleated*).

Your ophthalmologist will explain all the options that are feasible for treating your specific ocular melanoma so that you understand the course of your treatment. In the following pages is information on four treatment options:

- Plaque therapy
- Enucleation
- Thermotherapy
- Additive therapy

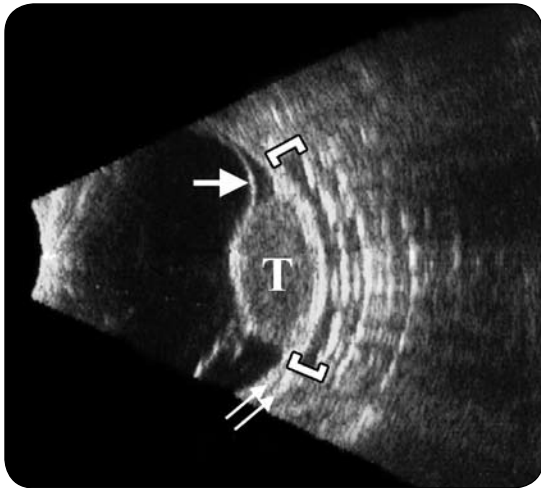
PLAQUE THERAPY

Most choroidal melanomas can be destroyed by special localized radiation therapy called *plaque therapy* or *brachytherapy*. The plaque consists of a small gold cover or shield with radioactive pellets inserted inside a plastic carrier. The plaque is sutured to the wall of the eye (*sclera*) beneath the base of the tumor while the patient is “asleep” under general anesthesia. The plaque therapy itself causes only minimal discomfort while the plaque is in place. The plaque is removed surgically once the tumor has received sufficient radiation to destroy it. This typically takes four days.

Plaque therapy is generally safe and effective. The gold cover prevents radiation from escaping and affecting the tissues surrounding the eye (the orbit) or brain. Its effectiveness was demonstrated by the Collaborative Ocular Melanoma Study. This clinical trial was funded by the National Eye Institute, part of the National Institutes of Health. It began in 1986 and was carried out at specific health centers across the country. The University of Michigan Kellogg Eye Center was a major participant. The Study showed that plaque therapy, in medium-sized tumors, was as effective as removal of the eye in preventing the spread of the tumor to other parts of the body.



This is the plaque that is placed on your eye. The radioactive “seeds” inside the plaque will act directly on the tumor to destroy it. It is a little thicker than a dime, and its diameter is the size of a nickel to a quarter.



This ultrasound shows the tumor and the plaque.

- T = tumor
- ➔ = retinal detachment caused by the tumor
- ⇔ = sclera
- [] = the plaque

Plaque therapy can be used for small and medium-sized melanomas. After plaque therapy, the melanoma cells are unable to multiply or grow and the tumor gradually shrinks to an inactive, scarred nodule. Very large melanomas (diameter greater than 16 mm or height greater than 10 mm) cannot be successfully treated with plaque therapy.

Plaque therapy now can be done on an outpatient basis. Patients typically have the plaque inserted on Monday morning and return on Friday to have the plaque removed.

Plaque therapy, however, can cause damage to sensitive ocular tissues, such as the optic nerve (the cable from the eye to the brain) and the macula (the central part of the retina). The closer the tumor is to the optic nerve or macula, the greater the risk of visual loss following plaque therapy.

Radiation therapy can also be carried out with high energy particles (helium ion or proton beam radiation) from a cyclotron, but this therapy results in a much higher radiation dose to the whole eye.

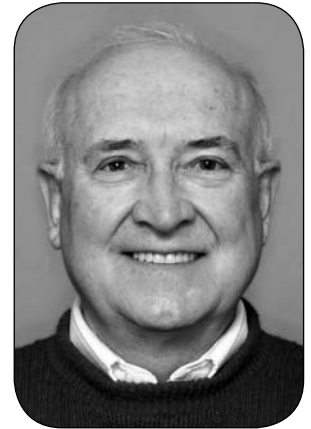
ENUCLEATION

Unfortunately, sometimes there are large melanoma tumors that cannot be treated by plaque therapy, and the only way to manage them is to remove the eye. This process is called enucleation. Kellogg's Oculoplastics Service performs all enucleations for choroidal melanomas.

Enucleation is performed with the patient "asleep" under general anesthesia. During the operation, an implant (a silicone ball) is used to replace the volume of the eye. The muscles that move the eye are sutured to the implant to provide some limited movement. The implant is not visible because normal tissue is sutured over it.

After healing occurs, an artificial eye (*prosthesis*) can be placed. The prosthesis is actually a plastic shell painted to resemble the normal eye. It fits over the implant. Although movement usually is less than that of the normal eye, the excellent appearance of the prosthesis makes it very unlikely that people will know the patient does not have two normal eyes.

The person who creates the prosthesis is known as an ocularist. The Kellogg Eye Center has one of the best in the country, Mr. Gregory Dootz. Mr. Dootz has been at the Eye Center for over 25 years and has developed close relationships with his patients. He understands their needs and fears and works with them to create beautiful prosthetic eyes that fit well and look real. He crafts these prosthetic eyes for adults and for children and infants, who require new eyes as they grow.



This photograph shows a gentleman with a prosthetic eye after his diseased eye was enucleated.

THERMOTHERAPY

Though rarely used, thermotherapy is one way to treat small (less than 3 mm in height), pigmented melanomas that are located in the back of the eye. Thermotherapy uses heat (hyperthermia) to destroy the tumor cells. The heat is provided by a diode laser using high energy levels and long duration of laser “spots” or “burns.”

This therapy requires only local anesthesia (called *retrobulbar anesthesia*). In retrobulbar anesthesia, a needle is passed below the eye to freeze the eye. After the anesthetic has taken effect the laser treatment is performed. The tumor may require 1 to 3 treatments which are typically spaced 3 months apart.

There are disadvantages to thermotherapy: it is limited to small tumors, and there is a high rate of local recurrence (the tumor starts growing again). Thermotherapy is ineffective in destroying tumor cells within the sclera, the white wall of the eye. Depending on the location of the tumor, 22–29 percent of eyes that undergo thermotherapy will develop a local recurrence of tumor within three years of the initial treatment. Thermotherapy can also result in mild to severe visual loss in the treated eye due to the intensity of the laser burns. At present, there is very limited long-term experience with this therapy.

ADDITIVE THERAPY

Patients with large choroidal melanomas have an increased risk of developing metastatic disease in other parts of the body. This can develop months or years after the eye is removed (enucleated) or treated.

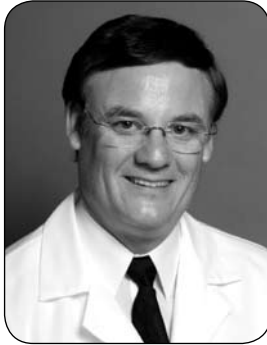
In many cancers, particularly breast cancer, additional therapy like chemotherapy or radiation after a lumpectomy or mastectomy has been shown to decrease metastases or local recurrences. Unfortunately, additional therapies have not been shown to be beneficial in ocular melanoma.

Although interferon therapy is partially beneficial in advanced skin melanoma, no studies have shown chemotherapy to be a benefit in ocular melanoma. The same is true for radiation.

MEET OUR OCULAR ONCOLOGY SPECIALISTS



Grant M. Comer, M.D.
*Director of Ocular
Oncology*



Gregory L. Dootz
Ocularist



Kathleen A. Meyer, R.D.M.S.
Ultrasonographer



Christine C. Nelson, M.D.
*Orbital and
Oculoplastic Surgeon*

Occasionally, the Ocular Oncology Service participates in clinical studies. If you are interested in participating in a trial, please ask your doctor whether any are available at Kellogg.

Patient Appointments: 734.763.5906

ADDITIONAL RESOURCES

The following organizations provide reputable and up-to-date information for patients

American Cancer Society

1.800.ACS.2345 1.800.227.2345

This toll-free number is staffed 24 hours a day

www.cancer.org

Cancer Research UK

www.cancerhelp.org.uk

Eye Cancer Network

www.eyecancer.com

National Cancer Institute of the U.S. National Institutes of Health

1.800.4.CANCER 1.800.422.6237

This toll-free number is staffed Monday–Friday, 9 a.m.–4:30 p.m.

www.nci.nih.gov/cancerinfo/pdq/treatment/intraocularmelanoma/patient

University of Michigan Cancer AnswerLine

1.800.865.1125

This toll-free number is staffed Monday–Friday, 9 a.m.–4:30 p.m.

www.cancer.med.umich.edu/support/cancer_answerline.shtml

University of Michigan Kellogg Eye Center

www.kellogg.umich.edu



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