OM SNAPSHOT

Ocular melanoma is an extremely rare cancer of the eye, occurring in 5 to 6 people per million. OM is different from skin (cutaneous) melanoma and there is little evidence it is caused by sun exposure. It is the second most common type of melanoma after skin melanoma, representing approximately 5% of all melanomas.

FAST FACTS

1. Around 2,000 new cases in the U.S. every year
2. Most common form of cancer in the adult eye
3. Very different from skin melanoma
4. Aggressive cancer often spreading to the liver
5. 50% chance of spreading (metastasizing)
6. No cure today for metastatic ocular melanoma

WHY OMF

A LIFELINE WHEN THERE WAS NONE

The Ocular Melanoma Foundation (OMF) was established in 2003 by Dr. Robert Allen, a renowned Virginia eye surgeon who was diagnosed with ocular melanoma and, in 2005, succumbed to the disease. Now the #1 online destination for OM information, OMF is dedicated to supporting patients, caregivers and cancer researchers.

Among its many educational and research initiatives, OMF created the world's only Patient Forum dedicated to OM, hosts the annual Eye Am Not Alone (EANA) patient retreat series, launched the Travel Assistance Grant (TAG) program and, in 2013, established a $50,000 per year OM research grant in collaboration with the AACR.

SEE A CURE

SUPPORTING PATIENTS & EYE CANCER RESEARCH UNTIL WE SEE A CURE.

"Let me applaud you and the entire team at OMF for the exemplary service and education you are providing patients and the community on ocular melanoma."
- Prithvi Mruthyunjaya, MD, Duke Eye Center

"The OMF [Eye Am Not Alone] retreat was inspiring and re-invigorating. I met a bunch of warriors who were creatively and self-assuredly taking their lives in their own hands and even having lots of fun at it, despite the serious subject. There was a lot of laughter there. The benevolent spirit was infectious."
- Peter (OM Patient)

EYE AM NOT ALONE
EDUCATING, SUPPORTING & BUILDING HOPE FOR THE OCULAR MELANOMA COMMUNITY

GET INVOLVED

volunteer@ocularmelanoma.org

OMF OCULAR MELANOMA FOUNDATION

Ocular Melanoma Foundation
A 501(c)3 Charitable Corporation EIN: 20-0355509
1717 K Street NW Suite 900
Washington, DC 20006
(202) 684.9380 | (800) 663-5930

www.ocularmelanoma.org

www.facebook.com/ocularmelanoma

@ocularmelanoma

OCULAR MELANOMA

AN OVERVIEW OF DIAGNOSIS, TREATMENT & PATIENT RESOURCES

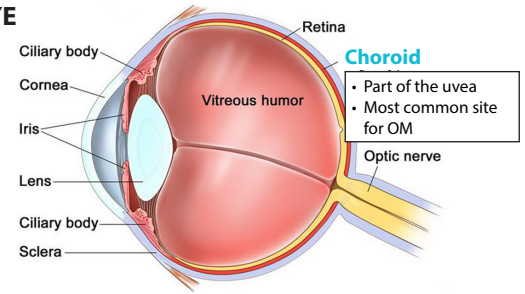
OMF OCULAR MELANOMA FOUNDATION

OM OVERVIEW

Ocular melanoma, or OM, is melanoma of the eye. Often called uveal melanoma or simply eye cancer, it is the most common form of cancer in the adult eye.

There are around 2,000 new cases of OM in the United States each year with an overall incidence of less than 6 people per million. Other eye tumors such as lymphomas and hemangiomas are even less common.

THE EYE



OM typically begins in the choroid, part of the uveal tract of the eye. As with skin cancer, OM is caused by mutations in the pigment-producing melanocytes. No one knows what causes OM, but the risks are higher in those with fair skin and blue eyes. Some studies have linked OM with sun exposure but more hard evidence is needed. Diagnosis rates increase with age, peaking at around 70 years of age.

OM represents 5% of all melanomas and while there are many similarities, OM is distinct from skin melanoma. While both are highly aggressive cancers, skin melanomas have different mutations (BRAF, NRAS, KIT), spread differently and have historically become more commonplace, a trend less evident with OM. The most common OM mutations are of the GNAQ and GNA11 genes; BAP1 is also common.

ONLINE RESOURCES

Ocular Melanoma Foundation www.ocularmelanoma.org
National Cancer Institute j.mp/NCI_OM
DecisionDx-UM Overview www.myuvealmelanoma.com
MRF CURE OM Initiative www.cureom.com
Kimmel Cancer Center Metastatic Uveal Melanoma Program www.kimmelcancercenter.org/kcc/kccnew/clinicalcare/eye/
OM Friends & Family Facebook Group www.facebook.com/groups/omffriends/

DIAGNOSIS

An eye tumor may go unnoticed and only present itself with blurred vision, flashing lights or floaters in the eye after it has grown large enough. Often, there is no vision irregularity at all and the tumor is only detected through a dilated eye exam by a trained ophthalmologist.

Beyond direct examination with an ophthalmoscope, the doctor may use ultrasound, CT scan, angiography or a needle biopsy in making a full diagnosis of an eye tumor. The doctor will also test to see if the disease has spread; ultrasound, MRI, CT and/or PET scans may be utilitized.

GENOMIC TESTING

Nearly half of all intraocular melanomas metastasize to the liver and, less often, to other parts of the body. Such metastatic disease occurs almost exclusively in patients whose tumor shows chromosome 3 loss or a class 2 gene expression profile, or both. These genetic abnormalities are associated with a high risk of metastasis, especially if the tumor is large.

Genetic analysis requires a tumor sample, which is most often collected from an unucleated eye or via biopsy. With conjunctival tumors, these methods can seed tumor cells to other parts of the ocular surface so excisional biopsy is preferred, removing the tumor intact.

There are several ways in which the tumor can be analyzed in the laboratory. Gene expression profiling (DecisionDx-UM from Castle Biosciences), categorizes intraocular melanomas into Class 1a (low risk), Class 1b (medium risk) and class 2 (high risk). Techniques such as multiplex ligation-dependent probe amplification (MLPA) (Impact Genetics), genomic hybridization and next generation sequencing provide information on dozens or hundreds of selected genes. This field is advancing rapidly so that in addition to predicting metastatic disease, genomic testing is increasingly being used to target treatment at the defective genes. Biopsy is best performed at the time of primary treatment, it may be possible to obtain a useful sample months or years later, although the chances of success are reduced.

Peter Hovland, MD PhD, OMF's Medical Director, recommends genomic testing as a way to adapt surveillance schedules to the level of risk. Also, many patients find living with uncertainty difficult and prefer to know their chances of future health so that they can prepare for any eventuality.

TREATMENT

Treatment of primary eye tumors is generally highly effective and aims to spare vision and ocular tissue while limiting the chances of the cancer spreading. The most common treatment for small- and medium-sized tumors is radiation, which 80-90% of OM patients receive. With plaque brachytherapy, a small disc-shaped shield encasing radioactive seeds is attached to the outside of the eye, over the tumor. This plaque is removed after several days and, according to the NCI, 85% of patients treated this way kept their eye for at least five years.

With large tumors, the eye may be removed via enucleation after which the patient receives an ophthalmic implant. An ocularist will fit a prosthesis over this implant and make it appear nearly identical to the remaining eye. Other common primary treatments include proton beam therapy (another form of radiation), transpupillary thermotherapy (TTT laser treatment) and, in some cases, surgical resection (removal of tumor tissue).

For treatment of metastatic disease, there is a wide range of liver-directed therapies including resection, ablation and radiation (e.g. CyberKnife). Treatments such as chemoembolization (TACE), radioembolization and hepatic perfusion (PHP/IHP) introduce cancer-destroying agents directly into the liver. There is, however, no approved systemic treatment for OM. Unlike with cutaneous melanoma, chemotherapy has not been shown to be effective. You can learn more about available treatment options and clinical trials at **ocularmelanoma.org**.

SURVEILLANCE

Nearly 50% of OM patients will go on to develop metastatic disease but, at the time of primary diagnosis, metastatic disease will only be seen in about 3% of patients due to the micrometastatic nature of OM.

The earlier metastatic disease can be detected, the more options are generally available. Surveillance (i.e. ongoing monitoring) may include liver function tests, chest x-rays, liver/abdominal imaging (ultrasound, MRI or CT scan) and/or comprehensive PET-CT. MRI with contrast and diffusion weighting every six months to a year is a common regimen. Genomic testing does inform surveillance plans but there is no clear consensus regarding ongoing monitoring so it is imperative to understand and evaluate all options with an oncologist specializing in OM.