eyeInsights™

Uveal melanoma

- Risk factors
- Assessment & diagnosis
- Referral guidelines

Mass General Brigham
Mass Eye and Ear

Dear Colleagues,

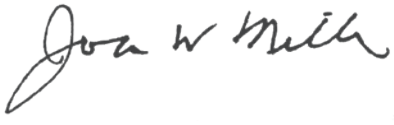
Over the last several decades, treatment for uveal melanoma—a rare cancer that can develop inside of the eye—has improved. Before 1975, the standard treatment was enucleation. Today, thanks to pioneering research at Mass Eye and Ear, many patients are able to preserve their sight with a precise therapy known as proton beam irradiation (PBI).

Building on over 50 years of service, the Ocular Melanoma Center at Mass Eye and Ear is a premier referral center for the diagnosis and treatment of ocular tumors and draws patients from around the world. Our uveal melanoma team has successfully treated thousands of patients using this very precise form of radiation.

Mass Eye and Ear researchers are continuing to pursue new and improved ways to treat tumors of the eye. The development and success of PBI is just one example of how our research efforts are rapidly advancing the care we provide.

Because patients with uveal melanoma are often asymptomatic, comprehensive ophthalmologists must be aware of the risk factors that make a patient more susceptible to developing these malignant tumors, including excessive exposure to ultraviolet light, race, and existing nevi located in the eye.

Inside this issue of *Eye Insights*, you will find helpful information in identifying the disease and current treatment options and outcomes.



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What is uveal melanoma?

Uveal melanoma is a rare malignant tumor that may develop in the iris, ciliary body, or choroid. It is the most common primary intraocular cancer in adults. Often discovered during a routine eye exam, uveal melanoma may arise from a pre-existing nevus. If left untreated, uveal melanoma can metastasize, most commonly to the liver.

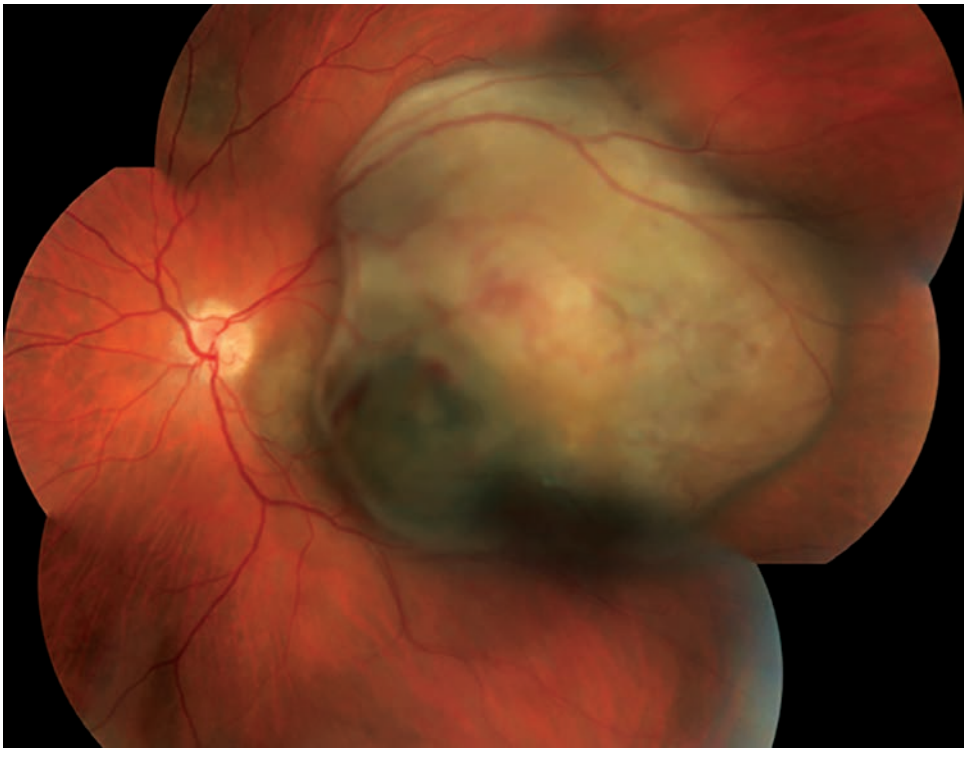
Symptoms

There are often no symptoms, but some patients experience:

- Flashing lights
- Floating particles
- Blurred vision
- Visual field loss

Prevalence

Uveal melanoma is very rare, with about 2,000 cases diagnosed each year in the United States. It is much more common in white individuals and occurs most often in adults over the age of 50.



ABOVE: *Uveal melanoma*

Ask the Experts

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Risk factors

Individuals with risk factors for uveal melanoma may have an increased likelihood of developing the disease. Risk factors include:

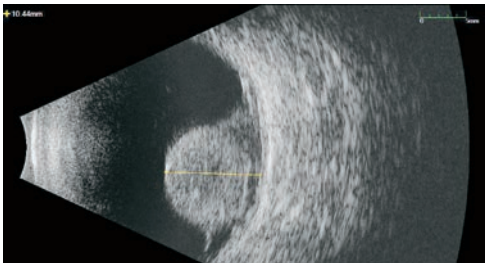
- White race
- Exposure to ultraviolet light
- Older age
- Blue or green eyes
- Inherited skin conditions
- Existing nevus in the eye

Assessment and diagnosis

Uveal melanoma is primarily discovered during a routine eye exam. Occasionally, symptoms such as floaters, flashes, or changes in vision may prompt the eye exam. Ancillary imaging and ultrasound are then used to determine the size and characteristics of the tumor.

Other imaging tests may include:

- Fluorescein angiography
- Fundus autofluorescence
- Optical coherence tomography



ABOVE: *Ultrasound of melanoma*

Treatment options

Radiation therapy with either brachytherapy or PBI is the most common treatment for uveal melanoma. These therapies allow the patient to retain the eye and functional vision in most cases.

Plaque brachytherapy

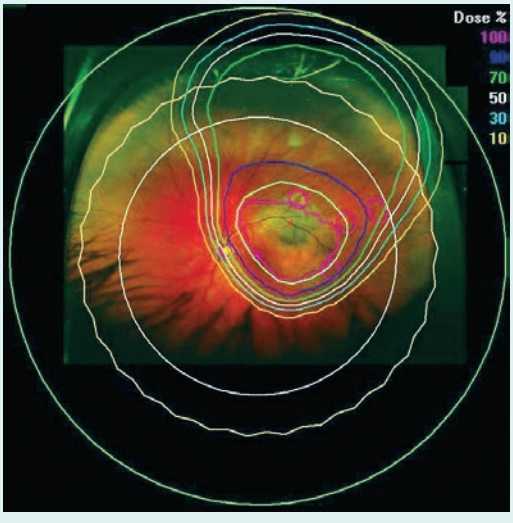
With this technique, a gold plaque embedded with radioactive seeds is sewn onto the eye near the tumor. It is left in place for a few days and then removed. Typically, patients can leave the hospital during treatment and return for plaque removal. The procedure to place the plaque is performed in an operating room while the patient is under anesthesia. The plaque is then removed after 3-7 days. A lead eye covering is used to protect the eye during treatment and to limit the spread of radiation.

Proton beam irradiation (PBI)

PBI provides a precise form of radiation to the tumor and is especially effective when treating tumors located near critical structures, specifically the optic nerve and the macula. The first step consists of surgery in which tantalum markers are sewn onto the eye to aid in tumor localization during radiation therapy. PBI treatments are painless and consist of five sessions that are scheduled over five consecutive days. Each radiation treatment lasts about one minute.

PBI developed at Mass Eye and Ear

PBI for uveal melanoma was developed by Dr. Evangelos Gragoudas and his colleagues at Mass Eye and Ear in collaboration with scientists at Harvard University's High Energy Physics Department and radiotherapists at Massachusetts General Hospital. It remains one of the most effective therapies for treating intraocular tumors.



LEFT: *Image of PBI treatment plan*

Uveal Melanoma Registry

Using outcomes research to advance disease management provides data for future research

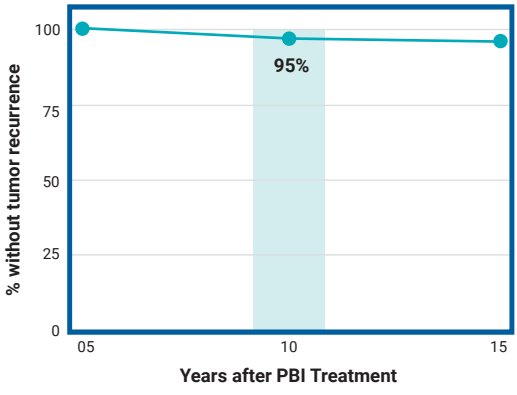
The Mass Eye and Ear Uveal Melanoma Registry provides an opportunity to analyze detailed demographic, clinical, and therapeutic information on more than 5,000 patients who have been treated with PBI at Mass Eye and Ear. A blood and tissue repository was added to the registry in the mid-1990s and contains samples from more than 2,500 patients with uveal melanoma. These data are used as a resource for biomarker and genetic research and enable Mass Eye and Ear to conduct several studies on cancer susceptibility and biomarkers. These data have been analyzed to advance our understanding of uveal melanoma and to learn more about the effectiveness of treatments offered to patients.

PBI outcomes

Forty years of follow-up studies show that PBI has the lowest local recurrence rate of any radiation therapy, and about 95% of patients are able to keep their eyes.

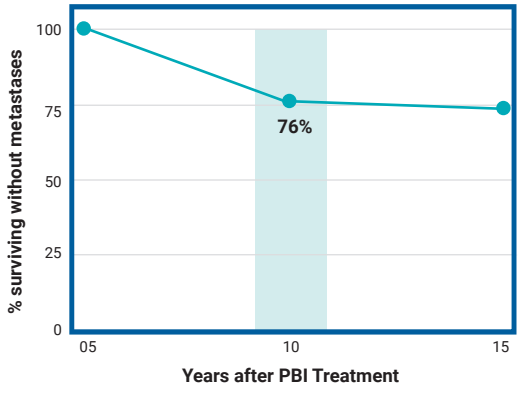
Most patients experience some loss of vision, which depends on the size and location of the tumor. Ten years after treatment, 30% of all patients still have vision better than 20/200.

Patients with smaller tumors located further away from the optic nerve and macula can have much better visual outcomes. Ten years after PBI, about 70% of these patients have vision better than or equal to 20/200.



Tumor control

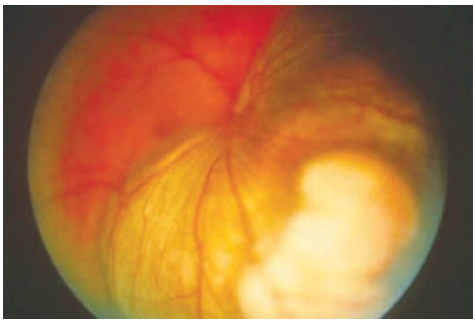
In patients who were treated with PBI, tumors remain controlled up to 15 years after treatment.



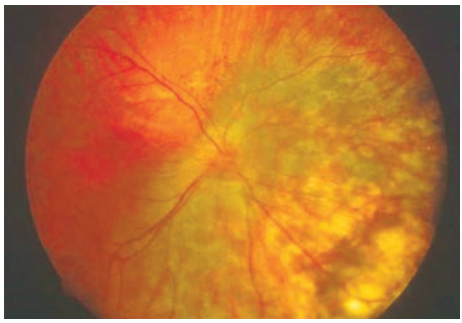
Metastasis-free survival

More than 80% of patients are alive without metastases five years after treatment and more than 75% are alive without metastases 10 years after treatment.

Ocular tumor: before and after treatment



Before treatment



After treatment

Referral guidelines

Consider referring a patient to a specialist as soon as uveal melanoma is suspected. Clinical features that increase suspicion for melanoma include lesion thickness > 2 mm identified on ultrasound, presence of subretinal fluid, orange pigment overlying the lesion, and proximity of the lesion to the optic disc (Jager et al, 2020). Working with a surgeon who specializes in ocular tumors will greatly improve patient outcomes.

Further reading

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eyeInsights™ MARCH 2024

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