What You Should Know About Choroidal Melanoma

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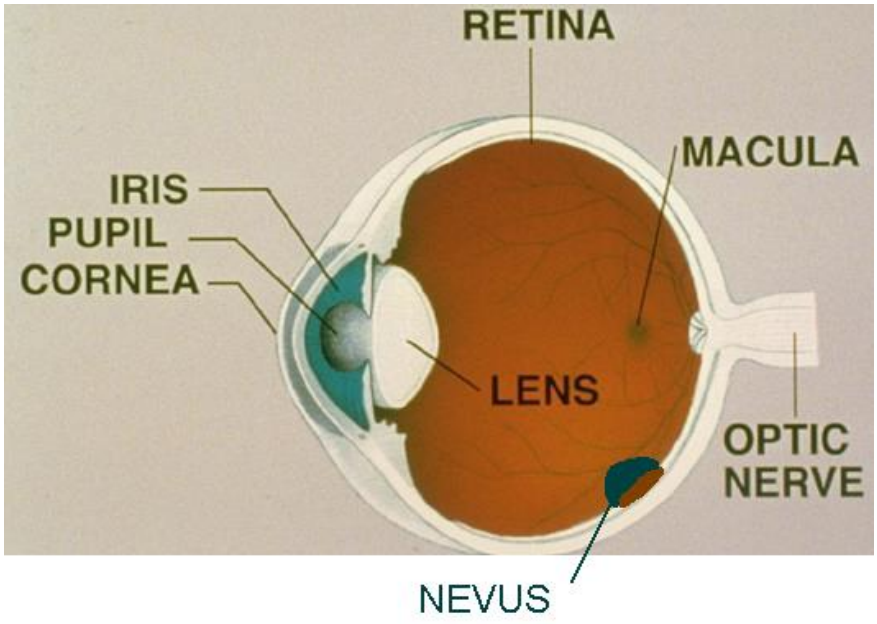
Choroidal melanoma is a cancer of the eye. It is the most common cancer that starts in the eye. In the United States, approximately 2000 new cases are discovered each year. All races can be affected, but whites are affected 10 times as frequently as blacks, and 3 times as frequently as people of Asian ancestry, thus pigmentation of the tissues in the back of the eye seems to be associated with some protection from the tumor. Women and men are equally affected. Family history of choroidal melanoma is rare. The cancer has no known predisposing environmental risks. Certain genetic conditions do increase the risk of choroidal melanoma, such as presence of multiple abnormal (dysplastic) moles, and an increased patch of pigment in the skin around the eye and in the white coating (the sclera) of the eye. Most cases occur late in life, but choroidal melanoma has a second smaller frequency peak in people in their 20's and 30's.

Where Do Choroidal Melanomas Come From?

Choroidal melanomas arise from moles present in the choroid, a thin layer of blood vessels, connective tissue, and pigmented cells found just inside the white sclera of the eye (see fig.1). Seven percent of people have moles in this layer, and they are almost always asymptomatic. Only 1 such mole in 5000 per year will undergo cancerous transformation to become a choroidal melanoma.

Figure 1.



How Serious Is Choroidal Melanoma?

Choroidal melanoma is a potentially lethal condition. As a cancer, it has the ability to spread (metastasize) to other organs, and if it does so, the patient usually will die from this process. For this reason, treatment of choroidal melanoma is recommended to kill the cancer before it spreads.

What Work-up Is Done?

The goal of the workup is to look for evidence of spread of the melanoma and to make sure the diagnosis is correct. To look for spread, the patient has a physical examination by the internist or family doctor. This picks up any skin or lymph node spread, found in 16% of cases where spread occurs. Blood is drawn for liver function tests since if choroidal melanoma spreads, it goes to the liver 90% of the time, and certain enzymes may be released into the bloodstream. A chest x-ray is done, since if spread occurs, it goes to the lung or its lining 24% of the time. An MRI scan of the eye may be obtained to look for any growth beyond the eyeball into the orbit. In only 2% of cases is any evidence of spread found at the time the eye cancer is diagnosed.

To confirm the diagnosis, a second opinion is obtained from another ophthalmologist. If there is any doubt, the two confer and perhaps ask other doctors to examine the patient.

How Is Choroidal Melanoma Treated?

Treatment of choroidal melanomas depends on several factors. The first factor is evidence of spread. If the cancer has spread, then treatment of the eye is usually not recommended, since the median time to death is just 9 months. The care in such cases is palliative - that is, directed toward comfort and quality of life. If no evidence of spread exists, then treatment to kill the melanoma in the eye depends on the size of the melanoma and its location in the eye.

If no cancer spread is evident, but the cancer is large - greater than 16 mm in greatest basal dimension or thicker than 10 mm, then enucleation is recommended. Enucleation is an operation to remove the entire eye and replace the lost volume with an artificial orbital implant. Over this implant, a prosthesis is fashioned and fit, which with modern techniques is cosmetically excellent and capable of some degree of normal motion. Enucleation is performed with the patient asleep. The surgery lasts about an hour and the patient can go home the same day. Approximately one month after surgery, the prosthetic shell is made by a specialist called an ocularist. Approximately one third of all patients who are diagnosed with a choroidal melanoma require an enucleation.[1]

If the melanoma is less than 16 mm in greatest basal dimension and less than 10 mm in thickness, radiation therapy (plaque therapy) is possible. An operation is performed sewing a gold applicator (plaque) about the size of a soft drink bottle cap to the wall of the eye over the

cancer. Seeds of radioactive Iodine^{125} are placed in the plaque and release radiation, which bombards the underlying cancer over several days after which the plaque is removed at a second operation. The first operation takes about an hour; the second about 15 minutes. Both are usually done under local anesthesia with sedation. The patient is hospitalized for 4 to 5 days.[2]

For smaller tumors, usually thinner than 4 mm, a type of laser treatment can be used called transpupillary thermotherapy. In this treatment, an infrared laser is used to heat the tumor tissue to approximately 65 degrees Celsius, inducing tumor death. This treatment is done in the office in stages. Usually three to four treatments are done at three-month intervals as the tumor shrinks.[3]

What Is The Follow-up?

One can never say that a patient with a choroidal melanoma is “cured”. Patients have died of metastasis 30 years after apparently successful treatment. Hence, lifelong follow-up is necessary. Initially, follow-up will be two to three visits per year, but later a yearly examination may be sufficient. Photographs and thickness measurements using ultrasonography are routinely performed to compare to baseline measurements.

How Can One Die of Choroidal Melanoma If The Initial Work-up Showed No Spread?

Our techniques for detecting spread of choroidal melanoma are imperfect. That is, we may be unable to pick up very small spots of tumor. For this reason, a negative workup for spread when the patient is initially evaluated does not mean that the cancer has not spread. It just means that no larger spots are detectable.

What Are The Odds For Survival With Choroidal Melanoma?

Statistics on survival odds frequently depress patients. They can sound sterile and bleak. For this reason, I would preface their presentation by reminding each patient that he or she is unique, and may well do much better than the odds predict. These statistics just give an average outcome for a large group of patients with a similar condition.

The odds for survival depend on the tumor size, the location of the tumor within the eye, the age of the patient, whether a certain genetic profile of mutations is present in the tumor cells, and the cell type of the melanoma.[4]

In general terms, for tumors with greatest basal dimension more than 16 mm or thickness greater than 10 mm, the chance of dying from the cancer at 5 years is 35%.

For tumors more than 3 mm thick but less than 10 mm thick and less than 16 mm in greatest basal dimension, the chance of dying from the cancer at 5 years is 20%.

For tumors less than 3 mm thick, the chance of dying from the cancer at 5 years is 2%.

Tumors in which cells have lost one copy of chromosome 3 have a higher rate of tumor related death. Gene expression profiling can further refine the risk of mortality according to a panel of specific gene mutations. Increasingly, samples of tumor tissue are taken by fine needle biopsy at the time of treatment to provide a more specific prognosis for the patient.[5]

If one takes all comers, approximately 50% of patients with choroidal melanoma eventually die of their cancer.[1]

What Is The Collaborative Ocular Melanoma Study?

This is a national study involving 43 centers in which treatments were compared to determine the best form of therapy. No new patients are entering the study. It showed that for large tumors, external radiation plus enucleation was no better than enucleation alone. For medium size tumors, the study showed that plaque therapy was as effective as enucleation with respect to mortality, with the added benefit of retention of the eye.

More recently research with genotyping has allowed ocular oncologists to provide a sharpened prognosis for patients based on the genetic characteristics of their tumors. Samples of tissue are sent to a genotyping laboratory and a profile is given that has predictive power regarding risk of mortality. Other research is directed toward more effective treatment of metastatic disease. Slow progress is being made.

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