

The Fundamentals of Uveal Melanoma

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Although rare, uveal melanoma is the most common primary intraocular malignant tumor in adults. Over the past 50 years, the incidence in North America has been stable, with 4.3 per million (or about 1,500 cases per year) being newly diagnosed (Sato, Han, & Yamamoto, 2008). Uveal melanoma differs from cutaneous melanoma in a variety of ways, such as primary treatment, metastatic pattern, and overall chemoresistance. Many of the classic chemotherapy regimens used in metastatic skin melanoma, such as dacarbazine or high-dose bolus interleukin-2, are ineffective when used in metastatic uveal melanoma. This article will review primary uveal melanoma.

Anatomy

The four tissues in the ocular region can give rise to primary melanoma, including the uveal tract, conjunctiva, eyelid, and orbit. The uvea is a vascular layer within the eye that is divided into the three anatomic compartments of the iris, ciliary body, and choroid (see Figure 1). Uveal melanoma is by far the most common of the ocular melanomas, and orbital melanoma is the least common (Shields & Shields, 2009). The iris is the most anterior portion of the uveal tract. It divides the anterior segment of the eye into the anterior and posterior chambers and is bathed by aqueous on both sides. The choroid, the most posterior part of the uveal tract, extends from the optic nerve to the ora serrata and is the pigmented vascular layer of the uveal tract that maintains the retina. Its primary function is to provide oxygen and nourishment to the outer retinal layers. The ciliary body is a ring-shaped structure when viewed from the front of the eye (Modi & Edward, 2008).

Epidemiology and Prognosis

Uveal melanoma represents 5%–6% of all melanoma diagnoses (Bedikian, 2006). The most important factor in the development of uveal melanoma is the presence of congenital ocular melanocytosis. This condition manifests as a gray-brown pigmentation in the periocular region, temporal fossa, palate, sclera, uvea, orbit, and meninges (see Figure 2). Caucasian individuals with this condition carry a 1 in 400 lifetime chance for the development of uveal melanoma. Other risk factors include the presence of a choroidal nevus, light eye color, fair skin, and inability to tan (Shields & Shields, 2009).

Survival rates for uveal melanoma remain poor. Intraocular location of a uveal melanoma also can affect a patient's prognosis for metastasis. Tumors confined to the iris carry the most favorable prognosis, followed by those in the choroids. Ciliary involvement carries the least favorable prognosis.

Tumor size (primarily largest tumor diameter) continues to be the dominant predictor for metastasis (American Joint Committee on Cancer, 2002). Choroidal melanomas are routinely classified by height and diameter as small, medium, or large (see Table 1). A small tumor at diagnosis carries a more favorable prognosis than a medium or large tumor (Bell & Wilson, 2004). Advances made in the treatment of the primary tumor have not resulted in any improvement in survival rates. As many as 50% of patients will de-

velop metastatic disease, even 10–15 years after diagnosis, which invariably leads to death (Triozzi, Eng, & Singh, 2008). At the time of diagnosis, 99% of patients have disease limited to the eye, but at least 30% will die of systemic metastases at five years and 45% at 15 years. The liver is involved in up to 95% of individuals who develop metastatic disease. Despite current therapy, the median survival of patients who develop liver metastases is reported to be about six months, and the one-year survival is estimated to be 10%–15% (Bedikian, 2006). Systemic monitoring twice yearly with physical examination and liver function tests and annual liver imaging and chest radiograph is advised (Sato et al., 2008).

Diagnosis

Most melanomas are dome-shaped, with the tumor thickness about 50% the basal dimension. Choroidal melanoma manifests as a brown or, less commonly, yellow mass deep in the retina and typically with secondary retinal detachment. Vitreous hemorrhage or secondary glaucoma can occasionally develop (Shields & Shields, 2009). Patients are frequently asymptomatic; however, they may complain of visual changes progressing to symptoms of retinal detachment.

The most reliable way to make the diagnosis of choroidal melanoma is with the use of indirect ophthalmoscopy by an experienced ophthalmologist (Modi & Edward, 2008). In cases that are atypical, ultrasonography, fluorescein angiography, indocyanine green angiography, optical

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