# Ocular and Orbital Tumors

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## Introduction and Objectives

Intraocular and orbital tumors are relatively rare disease entities. Approximately 2100 new cases of primary orbital or ocular tumors are diagnosed annually in the United States. While cancer metastases to the eye or the orbit comprise the majority of ocular and/or orbital tumors, the most common primary tumor of the eye in adults is intraocular melanoma.

The treatment of ocular and orbital tumors depends on the tumor types as well as the extent of the diseases. Radiation plays a major role in the treatment of ocular or orbital tumors. This chapter focuses on the management of the more commonly diagnosed primary tumors of the eye and orbit and examines:

- Recommendations for diagnoses and staging procedures of various diagnoses of intraocular and orbital tumors
- The staging systems and prognostic factors for malignancies of the eye and orbit
- Treatment recommendations for these tumors as well as their supporting scientific evidence
- Follow-up care and surveillance of survivors of ocular and orbital malignancies

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1.1 Diagnosis, Staging, and Prognoses of Intraocular Melanoma

1.1.1 Diagnosis

Initial Evaluation

- Intraocular melanoma is usually diagnosed on ophthalmologic examination. Patients may or may not present with symptoms at the time of diagnosis, depending on tumor size, location in the eye, and production of secondary retinal detachment.

- Frequently, a patient is seen by an optometrist or ophthalmologist for an unrelated condition, then sent to a retinal specialist and ultimately to the ocular oncologist for an incidental finding. The overall delay for small tumors was up to 129 days as compared to 50 days for medium tumors and 34 days for large tumors (Level IV) (Damato 2001). The diagnosis is only made when seen by the ocular oncologist since almost 30% of patients referred to an ocular oncology service with a diagnosis of choroidal melanoma have an incorrect diagnosis (Level IV) (Khan and Damato 2007).

- The essential diagnostic procedures include ocular examination, especially indirect ophthalmoscopy, fundus photograph, ultrasonography and fluorescein angiography.

- With the exception of iris melanomas, which are amenable to biopsy, diagnosis is usually on a clinical basis and is in excess of 99% accurate (COMS, Level II) (Collaborative Ocular Melanoma Study Group 1990).

- The disease incidence in men is $6.8 \times 10^6$ and in women $5.3 \times 10^6$ (McLaughlin et al. 2005) in the United States. In Europe, incidence rates increase from south to north from < $2 \times 10^6$ in Spain and southern Italy up to > $8 \times 10^6$ in Norway and Denmark (Virgili et al. 2007). This variation of incidence with altitude also holds for the United States (Yu et al. 2006) for internal ocular melanoma, but is the reverse for external ocular melanoma (eyelid and conjunctival melanoma).

Laboratory Tests

- Initial blood tests should include a complete blood count with differential, basic blood chemistry, liver function tests, and renal function tests.

Imaging Studies

- Intraocular melanoma is commonly a localized disease and imaging studies for distant metastasis is usually not indicated for asymptomatic patients, except for pre-treatment chest X-ray.

- Ultrasound or CT scan of the abdomen is indicated to rule out hepatic metastasis in patients with abnormal liver function test or presenting with suggestive symptoms.

Pathology

- Intraocular melanoma is commonly diagnosed clinically and biopsy of the intraocular lesion should be avoided.

- Diagnosis is usually on a clinical basis and is in excess of 99% accurate (COMS, Level II) (Collaborative Ocular Melanoma Study Group 1990).

1.1.2 Staging

- Intraocular melanoma is staged clinically by location of tumor in the eye and by tumor dimension and presence or absence of extrascleral extension.

- The AJCC staging system (Table 1.1) gives the staging for uveal melanoma and melanoma of the ciliary body and choroid.